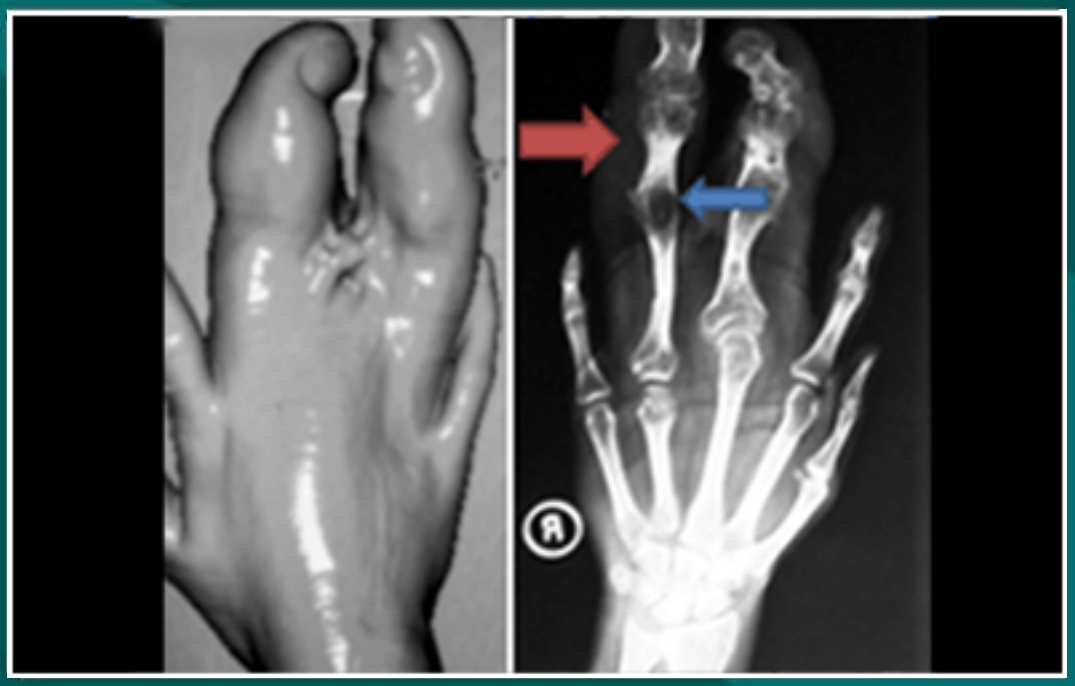
August 2017; Volume 8; Number 8

IJCRI

www.ijcasereportsandimages.com

International Journal of

CASE REPORTS AND IMAGES



Online submission at:
www.ijcasereportandimages.com/submit

ISSN: 0976-3198

International Journal of Case Reports and Images

Contact Details:

Editorial Office

Email: meditor@ijcasereportsandimages.com
Fax: +1-773-409-5040
Website: www.ijcasereportsandimages.com

Guidelines for Authors

Full instructions are available online at:
www.ijcasereportsandimages.com/submit/instructions-for-authors

Manuscript submission:
www.ijcasereportsandimages.com/submit

International Journal of Case Reports and Images

Contact Details:

Editorial Office

Email: meditor@ijcasereportsandimages.com
Fax: +1-773-409-5040
Website: www.ijcasereportsandimages.com

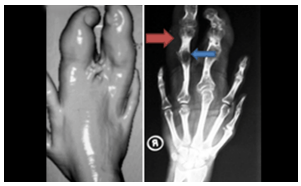
Guidelines for Authors

Full instructions are available online at:
www.ijcasereportsandimages.com/submit/instructions-for-authors

Manuscript submission:
www.ijcasereportsandimages.com/submit

International Journal of
Case Reports and Images

Cover Image



Volume rendering technique image showing gross enlargement of middle and ring figures of right hand, Plain radiographs right hand anteroposterior and lateral views showing enlargement of 3rd and 4th fingers with overgrowth of soft tissue as well as bony metacarpals and phalanges. There is predominantly increased lucency in soft tissue compartment of affected fingers suggestive of fat (red arrows) and secondary degenerative changes with partial ankylosis at interphalangeal joints (blue arrows)

Open Access

Contents

Vol. 8, No. 8 (August 2017)

SUBMISSION INSTRUCTIONS
All manuscripts, including illustrations, should be submitted at: www.ijcasereportsandimages.com/submit or email to: submit@ijcasereportsandimages.com
Author Guidelines: www.ijcasereportsandimages.com/submit/instructions-for-authors.php
For any questions contact the Editorial Office via e-mail at: info@ijcasereportsandimages.com or Fax: 1-773-409-5040

EDITORIAL OPEN ACCESS

Case reports and case series versus modern evidence-based medicine: Merit for an individual patient and public health

Ekamol Tantisattamo

Medicine is a science which is used by clinicians to heal patients. At the same time, the patients teach their doctors. Since directly contacting their patients, doctors may encounter with different unusual clinical presentations of common diseases or even common manifestations of rare diseases. These experiences have been expressed as case reports or case series as one of the teaching tools to assist young physicians and colleagues to apply the information for other patients who encounter with similar scenario. Today's practicing medicine requires evidence-based medicine to be cooperated into medical practices and the majority of the evidences are resulted from more complex research study designs including analytic and experimental studies. However, it is common that the original idea or motivation to conduct these types of research studies are based on case reports or case series. Some may believe in hierarchical value of evidence with the lowest one are case reports or case series. Several aspects of case reports or case series and evidence-based medicine are different, but the goal of these are to enhance medical knowledge and subsequently improve patient care. Therefore, case reports and case series remain contributing to medical knowledge and even originate new knowledge. This editorial aims to emphasize several important aspects of case reports and case series from general sense for managing individual patient through their impacts on public health aspect.

Ekamol Tantisattamo

Affiliation: Multi-Organ Transplant Center, Division of Nephrology, Department of Internal Medicine, William Beaumont Hospital, Oakland University William Beaumont School of Medicine, Royal Oak, Michigan, United States.

Corresponding Author: Ekamol Tantisattamo, MD, FACP, FASN, FNKF, FAST, American Society of Hypertension Specialist in Clinical Hypertension, Transplant nephrologist, Assistant Professor of Medicine, Multi-Organ Transplant Center, Division of Nephrology, Department of Internal Medicine, Oakland University William Beaumont School of Medicine, 3535 West 13 Mile Road, Suite 644, Royal Oak, Michigan, United States, 48073; Email: etantisattamo@oakland.edu

Received: 23 May 2017
Published: 01 August 2017

Several scientific knowledge arise from observation of people and scientists. Sir Isaac Newton studied and worked on the theory of gravity from observing a fall of an apple [1]. Many invention came from inspiration. Wright brothers dreamed to fly after having a toy helicopter [2]. In addition, the previously unsuccessful experiments make scientists to further observe and research. Thomas Alva Edison who was not the first light bulb inventor, but was the first inventor of the light bulb, which can be used in a practical way, successfully invented a light bulb after several people failed to do so at that time [3].

Similar to any scientific fields, medical knowledge started from an observation from a single patient who has unusual presentations of common diseases or even usual presentations of uncommon conditions. Documentation and sharing experiences about the things that are still unclear lead the young generation to extend that knowledge to become more evidence and innovation.

Among non-experimental studies, case reports and case series are the most basic type of the study, but they are often the fundamental knowledge of all other types of non-experimental studies including case series, cross sectional, case-control, or cohort studies and experimental study like clinical trial [4] (Figure 1).

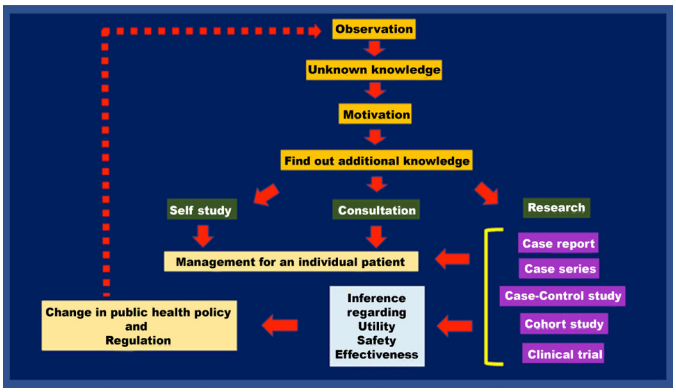


Figure 1: Conceptual framework demonstrates the potential relationship and a 'cycle of medical knowledge' generating from observation, different processes of obtaining information, and the impact of different types of research study designs on an individual patient through public health.

Nowadays, medical knowledge progresses continuously and rapidly. Medical research from different study designs including case reports and case series can bring different values to medical fields in different ways. The goal of these studies is to discover and add on to new medical knowledge. However, there are some different aspects that unique for each study design (Table 1).

All clinicians and healthcare providers practice and learn from their patients. Most clinicians consult their colleagues or specialists whenever they encounter with unclear clinical situations in terms of clinical manifestations, diagnosis, interpretation of laboratory data or investigation, and management particularly in the setting of rare conditions or diseases as well as rare manifestations in common diseases. Nevertheless, uncertainties sometimes remain. Previously published clinical case reports or case series are therefore other resource that are helpful to assist in their clinical decision making.

Not only the benefit of case reports to help in the management for a single patient, they also potentially extend the benefit to societies at large or even to public health. The recent example that emphasizes this benefit is Zika virus epidemic. A clinical case report [5, 6] can lead to subsequent analytic studies to characterize clinical manifestations, investigations, transmission, and complications of Zika virus infection in human and further studies during Zika virus outbreak [7–13] as well as discovery of management and prevention such as vaccine [14, 15].

Table 1: Comparison between case-based studies and modern evidence-based medicine

	Case-based studies	Modern Evidence-based Medicine
Examples	- Case reports - Case series	Observational studies - Cross sectional study - Case-control study - Cohort study Experimental studies - Clinical trial
Background or motivation	Typical presentation of rare diseases/ conditions or uncommon presentations in common diseases/ conditions	Determine relationship or causal inference
Aim	Improve medical knowledge [18].	
Population	Single or several cases	Groups of patients
Level of evidence	Lower [4]	Higher [4]
Application to regulation and policy	May lead to prompt changing e.g. drug safety and withdrawal	Takes time to collect data and conduct studies
Medical education	Common presentation as clinical vignette for house staff and physicians in almost all levels of scientific meetings.	Evidence-based medicine
Con-sequences	Lesson for healthcare providers Available resource for consultation	Extend evidence to become more analytic and experimental studies.

Several values of case reports and case series are not only contributing to the field of medicine, but also impacting or changing regulation or even public health policy. Particularly, the initial step of analytic research for drug safety often starts with case reports or case series [16]. This can be demonstrated by one of the new field of public health, pharmacoepidemiology involving in drug utilization, effectiveness, and safety. With so many innovative drugs, biological, biosimilars, or medical devices undergoing accelerated approval process, uncertainties and uncovered information particularly adverse drug experiences (ADE) or unintended side effects may arise. For instance, a report of new or unusual ADE can guide drug consumers, providers, manufacturers, and governmental organizations responsible to the regulation of drug utilization, effectiveness, and safety like Food and Drug Administration (FDA) to be alerted. This subsequently leads to further investigation, conducting research to prove the causal inference of the ADE and drug utilization. Ultimately, the action of these research leads to new regulation and management of the drug utilization for the public [17–21].

Therefore, case report and case series are actually the origin of several observational and experimental studies and can expand observation from characteristics of a single or multiple cases to more complex studies and eventually evidence-based knowledge, which can be applied for patient care through public health. The experiences from this application can subsequently lead to further observation of unknown knowledge requiring additional processes of obtaining knowledge from self-study, consultation, or research. This becomes a “cycle of medical knowledge” as demonstrated in the Figure 1.

Although several merits of case reports and case series, some disadvantages may take into the consideration when utilizing the information from case reports or case series mainly due to limitation of making causal inferences from a small number of studied patients (Table 2).

CONCLUSION

From the past, nowadays, and in the future, case reports and case series tell us the stories and clinical entities of rare conditions or diseases or rare presentations of the common diseases. They are also the foundation of many descriptive as well as analytic studies and eventually experimental trials. Case reports and case series can not only be a way to help and communicate other physicians and healthcare providers about unusual aspect of individual case or a small group of patients, but

Table 2: Common values of case reports or case series

	Main value	Comments	Examples
Values	Generating new hypothesis, knowledge and changing medical practices	History of medicine changing knowledge or medical practices	Alexander Fleming discovered penicillin [19].
	A tool to communicate among medical professionals	Published case report, case series, letter, or brief communication	
	Common way to teach medical trainees	Integration for teaching since pre-clinical years through all of medical professional careers as well as lesson to avoid repeated pitfalls	
	Promoting scholarly activity for medical trainees	Special case report sessions for trainees in several specialty medical scientific meetings	Case repot section in medical journals
	Foundation of more complex medical research	A motivating factor to conduct further complex observational and experimental medical research to determine causal inferences	Zika epidemic [5, 6]
	Alternative way for experimental studies	Unethical to conduct randomized clinical trial Study of rare diseases or condition	Re-challenge criteria of causal inference which may be unethical for clinical trials [20].
	Public regulation	Initial trigger for drug utilization, effectiveness, and safety in pharmacoepidemiology	
Disadvantages	Unable to make causal inferences	Lack of quantitative data, epidemiologic and statistic methods	Not representative and generalizability
	Anecdotal fallacy [21]	Over interpretation and personal emotional aspect of the readers [4].	
	Biases	Subjective Incomplete information from a retrospective in nature	

also a foundation of idea and motivation in conducting. In addition, case reports and case series are also another method to formally inform involved and responsible stakeholders to further examine and again conduct further more complex research studies to determine causal inferences and this subsequently leads to further action and regulation to ultimately improve patient care. Therefore, case reports and case series should remain one of the efficient tools to inform and educate our colleagues, extend further medical knowledge for patient care, and potentially involve in public health.

****

Keywords: Case reports, Case series, Modern evidence-based medicine, Pharmacoepidemiology, Public health

How to cite this article

Tantisattamo E. Case reports and case series versus modern evidence-based medicine: Merit for an individual patient and public health. Int J Case Rep Images 2017;8(8):487–490.

Article ID: Z01201708ED10010ET

****

doi:10.5348/ijcri-201710-ED-10010

****

Author Contribution

Ekamol Tantisattamo – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Fara P. That the apple fell and Newton invented the law of gravity, thus removing God from the cosmos. In: Numbers RL, Kampourakis K, editor. Newton's Apple and Other Myths about Science. USA: Harvard University Press; 2015. p. 48–56.
2. Crouch TD. The Bishop's Boys: A Life of Wilbur and Orville Wright. London: W. W. Norton & Company; 2003.
3. https://www.biography.com/people/thomas-edison-9284349
4. Nissen T, Wynn R. The clinical case report: A review of its merits and limitations. BMC Res Notes 2014 Apr 23;7:264.
5. Macesic N, Abbott IJ, Johnson DF. Photo quiz: Fever and rash in a husband and wife returning from the cook Islands. Clin Infect Dis 2015 Nov 1;61(9):1445, 1485–6.
6. Foy BD, Kobylinski KC, Chilson Foy JL, et al. Probable non-vector-borne transmission of Zika virus, Colorado, USA. Emerg Infect Dis 2011 May;17(5):880–2.
7. Gourinat AC, O'Connor O, Calvez E, Goarant C, Dupont-Rouzeyrol M. Detection of Zika virus in urine. Emerg Infect Dis 2015 Jan;21(1):84–6.
8. Schuler-Faccini L, Ribeiro EM, Feitosa IM, et al. Possible association between Zika Virus infection and microcephaly - Brazil, 2015. MMWR Morb Mortal Wkly Rep 2016 Jan 29;65(3):59–62.
9. Rasmussen SA, Jamieson DJ, Honein MA, Petersen LR. Zika Virus and birth defects–reviewing the evidence for causality. N Engl J Med 2016 May 19;374(20):1981–7.
10. Mlakar J, Korva M, Tul N, et al. Zika Virus associated with microcephaly. N Engl J Med 2016 Mar 10;374(10):951–8.
11. Brasil P, Pereira JP Jr, Moreira ME, et al. Zika Virus infection in pregnant women in Rio de Janeiro. N Engl J Med 2016 Dec 15;375(24):2321–34.
12. de Araújo TV, Rodrigues LC, de Alencar Ximenes RA, et al. Association between Zika virus infection and microcephaly in Brazil, January to May, 2016: Preliminary report of a case-control study. Lancet Infect Dis 2016 Dec;16(12):1356–63.
13. Bautista LE, Sethi AK. Association between guillain-barré syndrome and Zika virus infection. Lancet 2016 Jun 25;387(10038):2599–600.
14. Larocca RA, Abbink P, Peron JP, et al. Vaccine protection against Zika virus from Brazil. Nature 2016 Aug 25;536(7617):474–8.
15. Dowd KA, Ko SY, Morabito KM, et al. Rapid development of a DNA vaccine for Zika virus. Science 2016 Oct 14;354(6309):237–240.
16. Abou Chakra CN, Pariente A, Pinet M, Nkeng L, Moore N, Moride Y. Case series in drug safety: A review to determine characteristics and quality. Drug Saf 2010 Dec 1;33(12):1081–8.
17. http://www.fda.gov/downloads/RegulatoryInformation/Guidances/UCM126834.pdf
18. Vandenbroucke JP. Case reports in an evidence-based world. J R Soc Med 1999 Apr;92(4):159–63.
19. Haven KF. Marvels of Science: 50 Fascinating 5-Minute Reads. Littleton, Colorado: Libraries Unlimited. 1994. p. 182.
20. Hill AB. The Environment and Disease: Association or Causation? Proc R Soc Med 1965 May;58:295–300.
21. Charlton BG, Walston F. Individual case studies in clinical research. J Eval Clin Pract 1998 May;4(2):147–55.

EDITORIAL OPEN ACCESS

Patients with McCune-Albright syndrome are predisposed to pancreatic cancer

Ricardo Correa, Mary Esquivel

INTRODUCTION

McCune-Albright syndrome (MAS) is a rare disorder defined by the classical association of polycystic fibrous dysplasia, precocious puberty and café au lait spots [1]. McCune-Albright syndrome are also associated with an increased risk of various endocrine and non-endocrine neoplasms, such as thyroid adenoma, GH-secreting pituitary adenomas, adrenal Cushing's and hepatobiliary and pancreatic tumors. McCune-Albright syndrome is caused by and autosomal dominant activating mutations of the G-protein alpha subunit gene (GNAS) [2].

CASE REPORT

A 55-year-old male with McCune-Albright syndrome with multiple manifestations including polyostotic fibrous dysplasia, thyroid involvement, history of Leydig cell tumor, and cafe-au-lait spot presented with recurrent episodes of pancreatitis with a pancreatic cyst.

The patient's history is significant for recurrent episodes of pancreatitis. The first one dates back to 1998 and a subsequent episode was documented in 2003. He recalls undergoing upper endoscopy (probably EUS or an ERCP), that demonstrated pancreatic cysts. In this context, the patient denies a history of alcohol consumption or cholelithiasis. During the admission, liver function test and lipase and amylase were within normal limits. Magnetic resonance imaging (MRI) scan of abdomen plus magnetic resonance cholangiopancreatography (MRCP) showed dilatation of the extrahepatic common bile duct and pancreatic duct, with replacement of pancreatic parenchyma by dilated side branches or pancreatic cyst. These findings are concerning for intraductal papillary mucinous neoplasm (mixed type). An upper endoscopy with ultrasound of the pancreatic area was performed and multiple cysts were seen and biopsied. As per pathology report, there was mucin and CEA was 124, no malignancy identified on the cytology.

Ricardo Correa[1], Mary Esquivel[1]

Affiliation: [1]Endocrinology, Diabetes and Metabolism division of the Warren Alpert Medical School of Brown University.

Corresponding Author: Ricardo Correa, 1 Evie Dr. Warren, Rhode Island, 02885, USA; Email: riccorrea20@hotmail.com

Received: 27 May 2017
Published: 01 August 2017

DISCUSSION

Somatic activating G-protein alpha subunit gene mutations have been reported in various hepatobiliary and pancreatic neoplasm such as hepatocellular adenoma, hepatocellular carcinoma and pancreatic intraductal papillary mucinous neoplasm (IPMN). cAMP pathway is involved in the pathophysiology of this neoplasm. The prevalence of IPMN is poorly known but has been estimated to be only 25 per 100,000 [3]. The IPMN have been described in patient with Carney complex due to inactivating mutations of PKAR1A, one of the main regulators of the cAMP signaling pathway. Idiopathic pancreatitis has been described in patients with MAS [1, 4] and could possibly been explained by IPMN, since mild pancreatitis is a classic mode of discover these neoplasms. In our patient, the pancreatic cyst was found because he had 2 episodes of pancreatitis and imaging was performed. In 2013, small observational study was published where they describe the new association between MAS and pancreatic neoplasm (IPMN) and liver adenomas and choledochal cyst. In that study, 32% of the patients were found to have hepatic, pancreatic or biliary lesions. Three of six patients in the series had numerous branch-duct IPMN. That study strongly suggests that cAMP pathway is involved in IPMN tumorigenesis. Given the long-term malignant potential of IPMN, all MAS patients might be offered routine screening by MRI scan. All detected lesions, should benefit from a multidisciplinary counseling (surgeon and gastrointestinologist) and management with follow, biopsy and surgical indications should be advised. If no lesion is found, MRI scan might be performed every five years [5]. In our patient, biopsy

failed to demonstrate IPMN. We will follow-up him with imaging.

How will this affect clinical practice?

The patient presented with a pancreatic cyst and recurrent episodes of pancreatitis. This is a red flag due to the association between MAS and IPMN (presented). In this case, patient with evidence of pancreatic cyst since 2004 with a negative biopsy at that time. Screening is recommended every five years in patient without lesions. There is no recommendation on how frequent we should screen patient with lesions that are negative for malignancy. On this patient, it took eight years for re-screening. The new MRI scan showed a suggestive IPMN lesion that was not demonstrated with the biopsy. Continuous screening should be done, probably every two years. With this new information, MRI scan of abdomen in MAS patients with pancreatic/liver alteration should be done every 2–3 years because of the high risk of developing IPMN or hepatobiliary neoplasm.

****

Keywords: Endocrine, McCune-Albright syndrome, Pancreatic intraductal papillary mucinous neoplasm

How to cite this article

Correa R, Esquivel M. Patients with McCune-Albright syndrome are predisposed to pancreatic cancer. Int J Case Rep Images 2017;8(8):491–493.

Article ID: Z01201708ED10010RC

****

doi:10.5348/ijcri-201710-ED-10010

****

Author Contributions

Ricardo Correa – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Mary Esquivel – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Collins MT, Singer FR, Eugster E. McCune-Albright syndrome and the extraskeletal manifestations of fibrous dysplasia. Orphanet J Rare Dis 2012 May 24;7 Suppl 1:S4.
2. Chanson P, Salenave S, Orcel P. McCune-Albright syndrome in adulthood. Pediatr Endocrinol Rev 2007 Aug;4 Suppl 4:453–62.
3. Reid-Lombardo KM, St Sauver J, Li Z, Ahrens WA, Unni KK, Que FG. Incidence, prevalence, and management of intraductal papillary mucinous neoplasm in Olmsted County, Minnesota, 1984-2005: a population study. Pancreas 2008 Aug;37(2):139–44.
4. Kanda M, Knight S, Topazian M, Mutant GNAS detected in duodenal collections of secretin-stimulated pancreatic juice indicates the presence or emergence of pancreatic cysts. Gut 2013 Jul;62(7):1024–33.
5. Gaujoux S, Salenave S, Ronot M, et al. Hepatobiliary and Pancreatic neoplasms in patients with McCune-Albright syndrome. J Clin Endocrinol Metab 2014 Jan;99(1):E97–101.

CASE SERIES

PEER REVIEWED | OPEN ACCESS

A series of three cases of macrodystrophia lipomatosa: Rare cause of focal limb gigantism

Chiranjeev Kumar Gathwal, Kulvinder Singh, Saru Singh, Monika B. Gathwal, Santosh Munde, Gaurav Malik, Vikas Verma

ABSTRACT

Introduction: Macrodystrophia lipomatosa (MDL) is defined as rare congenital entity characterized by proliferation of mesenchymal elements predominantly the fatty component resulting in localized or regional enlargement of part or whole limb. It usually presents with cosmetic problems and variable functional deformity. Even though history and physical examination are usually diagnostic, radiological evaluation especially the cross-sectional modalities can reliably distinguish MDL from other forms of localized congenital gigantism. Case Series: Here we are presenting a series of three cases of macrodystrophia lipomatosa (MDL) presented to radiology department with history of disproportionate focal overgrowth of limb. Conclusion: Macrodystrophia lipomatosa is a rare form of congenital localized gigantism. As there are numerous etiologies of focal gigantism and clinical distinction may be difficult at times, radiological evaluation is very useful in confining the differential diagnosis. Imaging, with magnetic resonance imaging (MRI) in particular, provides vital clues to diagnose the condition which is confirmed by histopathology. Though surgical consultation is often offered for cosmetic reasons, it should be delayed till puberty or when functional problems exist as surgical results are not very rewarding with significant recurrence rate.

Keywords: Fatty overgrowth, Macrodactyly, Macrodystrophia lipomatosa, Soft tissue overgrowth

Chiranjeev Kumar Gathwal[1], Kulvinder Singh[2], Saru Singh[3], Monika B. Gathwal[4], Santosh Munde[5], Gaurav Malik[1], Vikas Verma[6]

Affiliations: [1]MD, Assistant Professor in Department of Radiodiagnosis, BPS GMC for Women Khanpur Kalan Sonepat Haryana; [2]MD, Senior Consultant in Health Map Imaging in Department of Radiodiagnosis, BPS GMC for Women Khanpur Kalan Sonepat Haryana; [3]MD, Associate Professor in Department of Anaesthesia, BPS GMC for Women Khanpur Kalan Sonepat Haryana; [4]MD, Assistant Professor in Department of Pathology, BPS GMC for Women Khanpur Kalan Sonepat Haryana; [5]MS, Associate Professor, Department of Orthopedics, BPS GMC for Women Khanpur Kalan Sonepat Haryana; [6]MBBS, Senior Resident in Department of Radiodiagnosis, BPS GMC for Women Khanpur Kalan Sonepat Haryana.

Corresponding Author: Chiranjeev Kumar Gathwal, MD, Department of Radiodiagnosis BPS GMC for women, Khanpur Kalan Sonepat, Haryana 131305; Email: drchiranjeev@gmail.com

Received: 06 December 2017
Accepted: 24 April 2017
Published: 01 August 2017

How to cite this article

Gathwal CK, Singh K, Singh S, Gathwal MB, Munde S, Malik G, Verma V. A series of three cases of macrodystrophia lipomatosa: Rare cause of focal limb gigantism. Int J Case Rep Images 2017;8(8):494–500.

Article ID: Z01201708CS10089CG

****

doi:10.5348/ijcri-201710-CS-10089

INTRODUCTION

Macrodystrophia lipomatosa (MDL) is rare congenital entity characterized by proliferation of mesenchymal elements predominantly the fatty component resulting in localized or regional limb overgrowth. This disease is

characterized by localized gigantism due to overgrowth of all mesenchymal elements predominantly the fibrofatty tissue and should be differentiated from macromelia or hemihypertrophy [1]. The term macrodystrophia lipomatosa was first coined by Feriz in 1925 for describing lower extremity gigantism cases, which were associated with massive overgrowth of the accompanying adipose tissue. Barsky later gave a more detailed description of the focal MDL causing local gigantism of digits [2]. Here we are presenting a series of three cases of macrodystrophia lipomatosa.

CASE SERIES

Case 1

An 18-year-old female was referred from orthopedic department with a history of progressive enlargement of middle and ring fingers of right hand since childhood and progressive decrease in movements since two years. Her parents initially noticed the abnormally enlarged fingers at age of 2–3 years which increased significantly over the years asymptomatically. However, later on she developed progressive pain and variable loss of motion for last two years. She was also operated for this condition for cosmetic reasons by local surgeon. However, details were not available with the patient. At presentation she had enlarged phalanges, pain and inability to completely flex the fingers. There was no evidence of pitting edema, bruit or other sensory changes. She also had kyphosis-scoliotic deformity of spine which was noticed at 7–8 years with significant progression over the years. She had taken treatment with frequent physiotherapy sessions. However, no significant improvement occurred. There is no history of past operative treatment for kyphosis-scoliotic deformity. There was no family history of similar illnesses.

The initial physical examination revealed the presence of local gigantism of middle and ring fingers of right hand as seen in volume rendered technique (VRT) image (Figure 1A). Variable restriction of movements of involved fingers at presentation was also noticed. There were no overlying skin changes or cutaneous stigmata seen. There was no neurological deficit. The other digits and contralateral foot were normal. Kyphosis-scoliotic deformity was also seen at the time of examination. Plain radiographs (Figure 1B) showed enlargement of 3rd and 4th fingers with overgrowth of soft tissue as well as bony metacarpals and phalanges. There was increased lucency in soft tissue compartment of affected fingers suggestive of fat. Secondary degenerative changes as joint space reduction, marginal osteophytes and variable partial ankylosis were also noticed at metacarpophalangeal and interphalangeal joints. Plain radiographs (Figure 1C) reveal significant kyphosis-scoliotic deformity with right sided convexity centered at D12 vertebra. Vertebral anomalies can also be appreciated in dorsolumbar spine. Ultrasonography and color Doppler using high-resolution probe revealed abundant increase in the adipose tissue without any increase vascularity or any vascular malformation. Non contrast computed tomography (NCCT) hand (Figure 1D–E) revealed overgrowth of bony as well as soft tissue components of affected fingers. Metacarpals and phalanges showed growth in length and width with significant disproportionately overgrowth of fat in increased soft tissue compartment. Degenerative changes with partial ankylosis at metacarpophalangeal and interphalangeal joints can also be appreciated. No other imaging of spine was available at time of presentation. The patient underwent surgery of fingers for cosmetic reasons. Histopathology confirmed the presence of significantly excess adipose tissue in the excised specimen scattered within thin mesh-like fibrous tissue extending up to periosteal surface. A complete diagnosis of recurrent macrodystrophia lipomatosa affecting middle and index fingers of right hand was made.

Case 2

A nine-year-old female presented to radiology department for radiographic evaluation with a long history of progressive disproportionate enlargement of the medial two toes of the left foot. The overgrowth of toes was noticed in childhood which increased progressively with age. Initially it was asymptomatic but later on she developed difficulty in wearing slippers, walking with recurrent injury to the foot. On physical examination, 1st and 2nd toes of left foot were disproportionately enlarged with excessive soft tissue component on the plantar aspect. There was also widening of the web space between 1st and 2nd toes. There were no overlying cutaneous changes, pitting edema or bruit. Rest of the toes of left foot as well as right foot appeared normal. Plain radiography (Figure 2) of the left foot revealed disproportionately enlarged 1st and 2nd toes involving both bony and soft tissue components. There was increase in length and diameter of the involved phalanges with preserved cortex and trabecular pattern. There is also splaying of bony phalangeal ends with dorsal angulation; involving the 2nd toe more markedly. The overgrown soft tissue component showed increased lucency suggesting increased adipose tissue. Biopsy revealed encapsulated lobules of well differentiated fatty tissue extending up to periosteum. Based on typical clinical history, characteristic radiographic findings and histopathology, diagnosis of macrodystrophia lipomatosa was made.

Case 3

A three-year-old girl was referred to our department for magnetic resonance imaging (MRI) evaluation of local gigantism of the right foot. Parents gave history of enlargement of right foot since birth, particularly the 2nd and 3rd toes for which the patient was operated at one and a half years of age in some remote hospital. There

was some cosmetic improvement after the operation. However, operative records were not available at time of presentation. She had been relatively asymptomatic with only mild pain over her sole from plantar keratosis. However, in last six months parents noticed further accelerated overgrowth of her right foot including the adjacent great toe. Physical examination revealed diffuse enlargement of her right foot with exceptionally large first and third toes. Also painless non-compressible soft tissue overgrowths were palpable at the lateral aspect of ankle and plantar aspect of foot. No dilated superficial vein or vascular mark was noticed. Contralateral foot was normal. No other skeletal abnormality was found. Previously, there was plain CT scan done from outside with the patient. Plain CT scan revealed significantly overgrown right foot with overgrowth of both bony and soft tissue compartments. The overgrown soft tissue compartment was predominantly lucent suggestive of fatty nature. Both CT scan and reconstructed 3D bony images revealed enlarged and thickened 1st, 2nd and 3rd metatarsals and residual phalanges with widening of inter-digital gap. Postoperative changes can be appreciated in 2nd and 3rd toes. Magnetic resonance imaging scan of the right lower leg, ankle and foot was performed. There was increased deposition of subcutaneous fat at the lateral and posterior aspects of the ankle, posterior and plantar aspects of the heel, plantar surface of foot with disproportionately enlarged first and residual third toes. The increased fatty overgrowth had the same signal intensity as rest of the normally visualized subcutaneous fat. Streaky T1-hypointense and T2-hypointense signal intensities suggestive of fibrous strands could be observed inside the overgrown subcutaneous fat. The right tibial nerve distal part was fusiform enlarged with abundant lipomatous tissue seen as T1 hyperintense component with suppression on FatSat sequences interspersed among the thickened nerve fascicles (Figure 3).

The imaging findings were consistent with macrodystrophia lipomatosa and which was further confirmed on biopsy. Based on clinical history, radiographic evaluation and histopathological examination diagnosis of recurrent macrodystrophia lipomatosa was made.

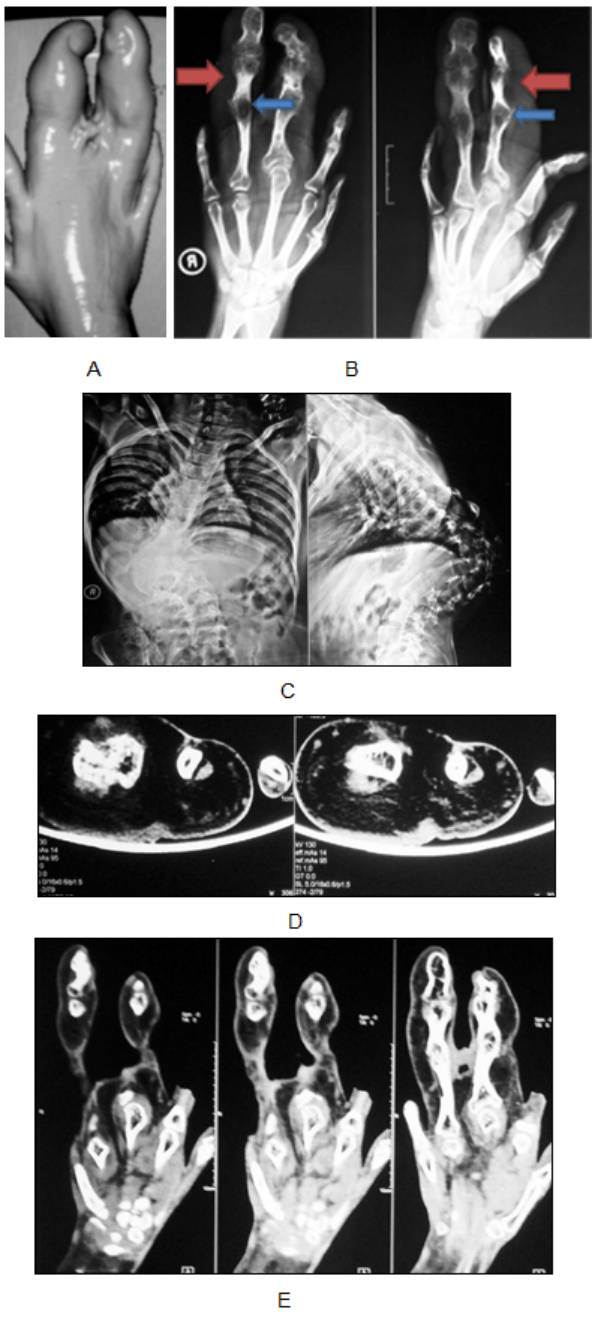
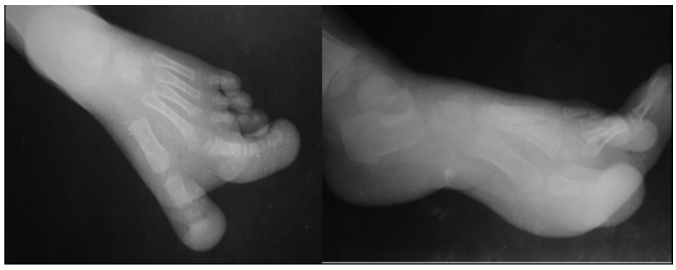


Figure 1 (A): Volume rendering technique image showing gross enlargement of middle and ring figures of right hand, (B) Plain radiographs right hand anteroposterior and lateral views showing enlargement of 3rd and 4th fingers with overgrowth of soft tissue as well as bony metacarpals and phalanges. There is predominantly increased lucency in soft tissue compartment of affected fingers suggestive of fat (red arrows) and secondary degenerative changes with partial ankylosis at interphalangeal joints (blue arrows), (C) Plain radiographs dorsolumbar spine anteroposterior and lateral views show kyphosis-scoliotic deformity with right sided convexity and centered at D12 vertebra. Vertebral anomalies can also be appreciated in dorsolumbar spine. (D, E) NCCT right hand (axial 1D and coronal 1E) showing overgrowth of middle and ring fingers involving both bony and soft tissue components with enlarged metacarpals and phalanges with disproportionately significant overgrowth of fat in increased soft tissue component. Degenerative changes with partial ankylosis seen at metacarpophalangeal and interphalangeal joints.

DISCUSSION

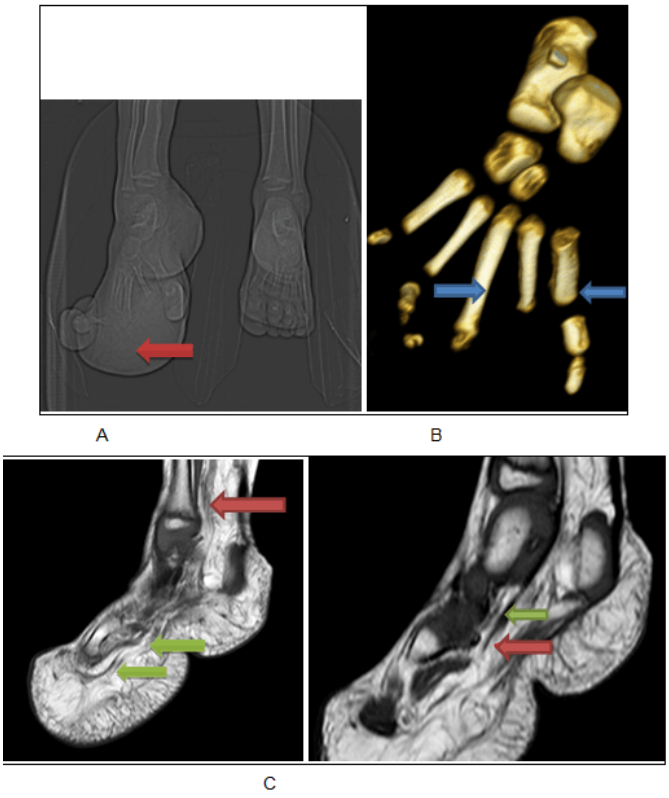
Macrodystrophia lipomatosa (MDL) is rare congenital entity characterized by proliferation of mesenchymal elements predominantly the fatty component resulting in localized or regional enlargement of part or whole limb.

This abnormality occurs most frequently in the distribution of median and plantar nerves in upper and lower extremities respectively. Usually, one or more digits of the unilateral limb are affected; there have been few reports of involvement of entire limb, bilateral limbs and abdominal wall as well [3, 4]. In most reported cases to date, the lesions are present at birth or develop



A B

Figure 2: (A, B) Plain radiography (anteroposterior and lateral views) of the left foot revealed disproportionate growth of 1st and 2nd toes involving both bony and soft tissue compartments. There is increase in length and diameter of the involved phalanges with splaying of ends and dorsal angulation; 2nd toe is being more affected. The overgrown soft tissue component shows increased lucency suggesting increased adipose tissue.



A B

C

Figure 3: (A) Plain computed tomography scan revealed significantly overgrown right foot with overgrowth of both bony and soft tissue compartments. The overgrown soft tissue compartment is predominantly lucent (red arrow) suggestive of fatty nature, (B) Reconstructed volume rendered 3D image revealed enlarged and thickened 1st, 2nd and 3rd metatarsals (blue arrows) and 1st toe phalanges with widening of inter-digital gap. Postoperative changes can be appreciated in 2nd and 3rd toes, (C) Sagittal T1-weighted image demonstrates the deposition of fatty tissue which infiltrates between the linear T1-hypointense nerve fascicles of the right tibial nerve (red arrow), with a characteristic spaghetti-like appearance. Fatty infiltration of the muscles over plantar aspect is also seen (green arrows), (D) Another sagittal T1W image demonstrating thickening and fatty infiltration of medial plantar nerve (red arrow) coursing posteromedial to flexor hallucis longus tendon (green arrow). Excessive fatty overgrowth seen in lower leg and foot especially affecting planter aspect and heel region.

within the first weeks of life, but generally only starts to cause problems as the child grows. Though Feriz in 1925 coined the term macrodystrophia lipomatosa, Barsky in 1967 described two forms of macrodactyly, static as enlargement of digits or limb in proportion of rest of the body and progressive form as disproportionate overgrowth of digits or limb and is usually associated with fatty overgrowth [5].

Exact etiopathogenesis of MDL being not well understood, the possible mechanisms given in literature include lipomatous degeneration, disturbed fetal circulation, disturbances of growth inhibiting factors, errors in segmentation, and trophic influence of tumefied nerve. The pathogenesis of bony enlargement is because of endosteal and periosteal deposition of bone [6].

The symptomatic problems associated with macrodystrophia lipomatosa are cosmetic and mechanical. Cosmetic problem is the usual presenting complaint in all ages but mechanical problems are encountered in adolescence due to secondary degenerative joint changes causing restricted movements and functions. Since overgrowth mainly involves the volar and plantar aspects, it can produce dorsal deviation of affected parts. This may lead to interference in normal day to day activities or make patient prone to repeated trauma. Osteophyte overgrowth can sometimes cause compression of adjacent nerves and vessels, most commonly seen in carpal tunnel syndrome [7, 8]. Other associations include lipomatous growths in intestines and other tissues, calvarial abnormalities, pigmented nevus, pulmonary cysts, syndactyly, polydactyly, clinodactyly, brachydactyly and symphalangism [9].

Imaging studies play an important role in characterization of nature of the hypertrophied tissue. Typical radiographic findings of macrodystrophia lipomatosa include excessive growth of soft tissue as well as osseous tissue. The presence of predominant radiolucent areas in overgrown soft tissue compartment suggests fatty nature of the soft tissue. Usually the volar aspect of the fingers is disproportionately involved [10, 11]. There is osseous hypertrophy and cortical thickening in the affected part of the body and this may lead to exostoses like bony outgrowths from the involved bone. Widening at the distal end of the bones gives the characteristic mushroom-like appearance [10, 11]. Ultrasonography and Doppler show large amounts of subcutaneous tissue, infiltration of the muscle and thickening of the affected nerves with absence of any increased vascularity. Excessive growth of the bone and fatty tissue proliferation are well appreciated findings detected on CT scan. The volar and plantar aspects of the fingers are disproportionately involved [10, 11].

The excessive fat seen in macrodystrophia lipomatosa is not encapsulated and MRI scan can easily demonstrate the fatty infiltration of the muscles. There may be linear T1/T2 hypointense fibrous bands noted within this abnormal fat. The fibro-adipose tissue appears hyperintense on both T1-weighted and T2-weighted MRI images, and is

identical to that of normal subcutaneous fat tissue. Soler et al. [10] proposed that MRI scan should be used as the diagnostic method of imaging for macrodystrophia lipomatosa to detect excess fibro-adipose tissue and enlargement of other mesenchymal tissues.

Histopathological findings show an abundant increase in adipose tissue scattered fibrous tissue [11]. Underlying subcutaneous tissues, nerve sheaths, muscles, periosteum, and even bone marrow involvement can also be seen. Differentials include neurofibromatosis type I, fibrolipomatosis hamartoma of nerve with macrodactyly, hemangiomatosis lymphangiomatosis, Klippel–Trenaunay–Weber syndrome and Proteus syndrome. Neurofibromatosis often has positive family history and certain characteristic cutaneous manifestations. Fibrolipomatosis of the nerve can also occur in isolation or with associated localized gigantism. Associated macrodactyly are seen in two-thirds cases and are usually difficult to distinguish from MDL.

It presents with typical nerve lesion associated with intramuscular fat deposition. In MDL, abnormal fat deposits are not limited within the nerve sheaths and muscles but also involve the subcutaneous tissues and tendons. Also there is periosteal involvement leading to the bony changes such as hypertrophy, exostoses, ankylosis of interphalangeal joints and fatty invasion of the medullary cavity which is quite specific for macrodystrophia lipomatosa [12].

Surgical intervention is the treatment of choice for macrodystrophia lipomatosa mainly to improve the cosmetic appearance while preserving the neurologic function as much as possible. Judicious and planned use of multiple debulking procedures, epiphysiodesis and various osteotomies are advisable to achieve the best results [13]. Surgery should be delayed till completion of growth if the deformity is not very serious and if no nervous system symptoms are present as there is localized recurrence rate of 33–60% after surgery [14].

CONCLUSION

To conclude, macrodystrophia lipomatosa is a rare form of congenital localized gigantism. As there are numerous etiologies of focal gigantism and clinical distinction may be difficult at times, radiological evaluation is very useful in confining the differential diagnosis. Imaging, with magnetic resonance imaging in particular, provides vital clues to diagnose the condition which is confirmed by histopathology. Though surgical consultation is often offered for cosmetic reasons, it should be delayed till puberty or when functional problems exist as surgical results are not very rewarding with significant recurrence rate.

****

Author Contributions

Chiranjeev Kumar Gathwal – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Kulvinder Singh – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Saru Singh – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Monika B. Gathwal – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Santosh Munde – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Gaurav Malik – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Vikas Verma – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Goldman AB, Kaye JJ. Macrodystrophia lipomatosa: Radiographic diagnosis. AJR Am J Roentgenol 1977 Jan;128(1):101–5.
2. Feriz H. Macrodystrophia lipomatosa progressive. Virchows Arch Pathol Anatomic Physiol Clin Med 1925;260:308–68.
3. Aydos SE, Fitoz S, Bökesoy I. Macrodystrophia lipomatosa of the feet and subcutaneous lipomas. Am J Med Genet A 2003 May 15;119A(1):63–5.
4. Jain R, Sawhney S, Berry M. CT diagnosis of macrodystrophia lipomatosa: A case report. Acta Radiol 1992 Nov;33(6):554–5.
5. Boren WL, Henry RE Jr, Wintch K. MR diagnosis of fibrolipomatous hamartoma of nerve: Association with nerve territory-oriented macrodactyly (macrodystrophia lipomatosa). Skeletal Radiol 1995 May;24(4):296–7.
6. Rohilla S, Jain N, Sharma R, Dhaulakhandi DB.

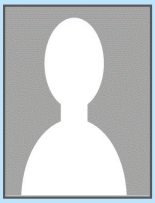
Macrodystrophia lipomatosa involving multiple nerves. J Orthop Traumatol 2012 Mar;13(1):41–5.
7. Meyer BU, Röricht S. Fibrolipomatous hamartoma of the proximal ulnar nerve associated with macrodactyly and macrodystrophia lipomatosa as an unusual cause of cubital tunnel syndrome. J Neurol Neurosurg Psychiatry 1997 Dec;63(6):808–10.
8. Kozanoglu E, Koc F, Goncu K. Macrodystrophia lipomatosa with multiple entrapment neuropathies: A case report. Int J Neurosci 2008 Apr;118(4):545–53.
9. Watt AJ, Chung KC. Macrodystrophia lipomatosa: A reconstructive approach to gigantism of the foot. J Foot Ankle Surg 2004 Jan–Feb;43(1):51–5.
10. Soler R, Rodríguez E, Bargiela A, Martinez C. MR findings of macrodystrophia lipomatosa. Clin Imaging 1997 Mar–Apr;21(2):135–7.
11. Gupta SK, Sharma OP, Sharma SV, Sood B, Gupta S. Macrodystrophia lipomatosa: Radiographic observations. Br J Radiol 1992 Sep;65(777):769–73.
12. De Maeseneer M, Jaovisidha S, Lenchik L, et al. Fibrolipomatous hamartoma: MR imaging findings. Skeletal Radiol 1997 Mar;26(3):155–60.
13. Wu KK. Macrodactylia fibrolipomatosis of the foot. J Foot Surg 1991 Jul–Aug;30(4):402–5.
14. Brodwater BK, Major NM, Goldner RD, Layfield LJ. Macrodystrophia lipomatosa with associated fibrolipomatous hamartoma of the median nerve. Pediatr Surg Int 2000;16(3):216–8.

ABOUT THE AUTHORS

Article citation: Gathwal CK, Singh K, Singh S, Gathwal MB, Munde S, Malik G, Verma V. A series of three cases of macrodystrophia lipomatosa: Rare cause of focal limb gigantism. Int J Case Rep Images 2017;8(8):494–500.

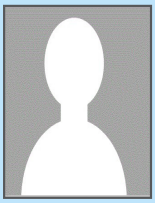
Chiranjeev Kumar Gathwal is Assistant Professor and Head at Department of Radiology BPS Government Medical College for Women, Khanpur Kalan, Sonepat, Haryana, India. He earned undergraduate degree (MBBS) from UCMS, New Delhi, India and postgraduate degree (MD Radiology) from Dr RML Hospital, New Delhi, India. His research interests include musculoskeletal, central nervous system, obstetrics and interventional radiology.
Email: drchiranjeev@gmail.com

Kulvinder Singh is Senior Consultant at HealthMap Imaging BPS Government Medical College for Women, Khanpur Kalan, Sonepat, Haryana, India. He earned MBBS and MD Radiology degrees. His research interests include musculoskeletal and interventional radiology.
Email: ksingh.rad@gmail.com

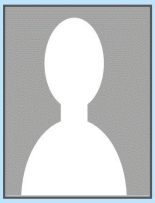


Saru Singh is Associate Professor in Department of Anesthesia, BPS Government Medical College for Women, Khanpur Kalan, Sonepat, Haryana, India.

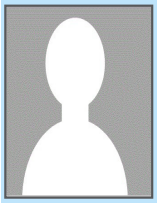
Monika B. Gathwal is Assistant Professor in Department of Pathology in BPS Government Medical College for Women, Khanpur Kalan, Sonepat, Haryana, India. She earned undergraduate degree (MBBS) from PGIMS Rohtak and MD Pathology from AIIMS, New Delhi. Her research interests include musculoskeletal, breast and central nervous system pathology.
Email: drmonikabreta@yahoo.com



Santosh Munde is Associate Professor, Department of Orthopedics, BPS Government Medical College for Women, Khanpur Kalan, Sonepat, Haryana, India.



Gaurav Malik is Assistant Professor in Department of Radiodiagnosis, BPS Government Medical College for Women, Khanpur Kalan, Sonepat, Haryana, India.



Vikas Verma is Senior Resident in Department of Radiodiagnosis, BPS Government Medical College for Women, Khanpur Kalan, Sonepat, Haryana, India.

CASE SERIES

PEER REVIEWED | OPEN ACCESS

Swing mandibulotomy access to deep lobe parotid tumors: A case series

Pradipta Das, Kavita Nivargi, S. Girish Rao, Sushma Mehta, Moumita Maity Rana, Pritam Chandrakant Chaudhary, Abhishek Singh Nayyar

ABSTRACT

Introduction: Pleomorphic adenoma is the most common salivary gland tumor. Most of these tumors originate in the superficial lobe with few involving the deeper lobe with medial extensions in to the adjacent para-pharyngeal space (PPS). The histological variation of the tumor along with its particular location poses difficulty in its accessibility for adequate removal of the tumor. Different approaches have been described with varying indications for different clinical situations and intricacies in gaining access to the tumor because of the difficulty involved in getting into the para-pharyngeal space (PPS). Thus, it becomes of utmost significance to select the right surgical approach for such kind of cases balancing maximum exposition and complete and safe removal of the tumor with minimal functional and esthetic morbidity. Case Series: In the present case series, two patients were selected on standard criterion based on histological type, site, age, sex, clinical features, radiological evaluation, recurrence of the parotid tumors and surgical access to these tumors. Patients selected on eligibility criteria were evaluated and underwent, further, radiological evaluation and were surgically managed. Thereafter, long term follow-up was done clinically as well as radiologically. Successful surgical outcome was determined on the basis of functional and clinical recovery. Conclusion: Selecting the best approach to maximize visibility, ensure complete removal of the tumor and to reduce morbidity is an important decision to be made. Swing mandibulotomy access provides an excellent mandible sparing surgical approach to gain access to deep lobe parotid tumors which are otherwise not accessible without procedures which leave significant morbidity.

Keywords: Adenoma Pleomorphic, Parotid tumors, Swing mandibulotomy

Pradipta Das[1], Kavita Nivargi[2], S. Girish Rao[3], Sushma Mehta[4], Moumita Maity Rana[5], Pritam Chandrakant Chaudhary[6], Abhishek Singh Nayyar[7]

Affiliations: [1]Assistant Professor, Department of Surgery, Kalinga Institute of Medical Sciences, Patia, Bhubaneswar, Odisha, India; [2]Reader, Department of Anaesthesia, Pandit Deendayal Upadhyay Dental College and Hospital, Solapur, Maharashtra, India; [3]Former Professor and Head, Department of Oral and Maxillofacial Surgery, D.A.P.M.R.V. Dental College and Hospital, J.P. Nagar, 1ST Phase, Bangalore, Karnataka, India; [4]Former Postgraduate Student, Department of Oral and Maxillofacial Surgery, D.A.P.M.R.V. Dental College and Hospital, J.P. Nagar, 1ST Phase, Bangalore, Karnataka, India; [5]Postgraduate Student, Department of Oral and Maxillofacial Surgery, Awadh Dental College and Hospital, Jamshedpur, Jharkhand, India; [6]Postgraduate Student, Department of Oral and Maxillofacial Surgery, ACPM Dental College and Hospital, Dhule, Maharashtra, India; [7]Reader cum PG Guide, Department of Oral and Maxillofacial Medicine and Radiology, Saraswati Dhanwantari Dental College and Hospital and Postgraduate Research Institute, Parbhani, Maharashtra, India.

Corresponding Author: Dr. Abhishek Singh Nayyar, 44, Behind Singla Nursing Home, New Friends' Colony, Model Town, Panipat-132 103, Haryana, India; Email: singhabhishek.rims@gmail.com

Received: 27 August 2016
Accepted: 22 September 2016
Published: 01 August 2017

How to cite this article

Das P, Nivargi K, Rao G, Mehta S, Rana MM, Chaudhary PC, Nayyar AS. Swing mandibulotomy access to deep lobe parotid tumors: A case series. Int J Case Rep Images 2017;8(8):501–509.

Article ID: Z01201708CS10090SR

****

doi:10.5348/ijcri-201711-CS-10090

INTRODUCTION

Pleomorphic adenoma is the most common salivary gland tumor [1]. Most of these tumors originate in the superficial lobe with few involving the deeper lobe with medial extensions in to the adjacent parapharyngeal space [2]. These tumors are generally considered to be benign even if the lesion presents with varied histological features due to different components with myxoid or, chondroid components. The main characteristics include its high recurrence rate and not infrequent malignant transformation. Most cases present as asymptomatic swellings without involving the facial nerve until there is substantial growth and impingement of the facial nerve. Often, onset of a facial nerve deficit, changes in consistency, rapid growth, and pain are the signs for the malignant transformation of the lesion. The histological variation of the tumor along with its particular location poses difficulty in its accessibility for adequate removal of the tumor. Because of the difficulty involved in getting into the parapharyngeal space, different approaches have been described including the trans-cervical, the first approach, described by Morfit HM [3], trans-cervical-trans-parotid, the most widely used, helpful in parapharyngeal space tumors originating in the parotid deep lobe, trans-palatal or, trans-oral, described by Ehrlich H [2] and limited to small non-vascular tumors, trans-mandibular with mandibular osteotomy, described as a complement to the other approaches in order to improve and increase access to the parapharyngeal space, with Ariel et al. [4] being the first to propose this approach to enter the parapharyngeal space with numerous variations in the procedure being described later [5, 6], and lastly, the orbito-zygomatic approach to the middle cranial fossa described by Fisch U [7] to give access to parapharyngeal space tumors affecting the temporal bone or, relatively larger tumors reaching the base of skull.

CASE SERIES

Case 1

A 59-year-old male was presented to the unit complaining of a gradually increasing, painless swelling in the right parotid region since four months. The swelling was associated with dysphagia and dysphonia. The patient gave history of pleomorphic adenoma of right parotid 15 years back following which he underwent superficial parotidectomy. Pleomorphic adenoma was diagnosed by Fine-needle aspiration cytology (FNAC) technique. Magnetic resonance imaging scan revealed a dense extensive, homogeneous mass, 5 cm in diameter, originating in the right deep parotid lobe and extensively involving the parapharyngeal space, base of skull and infra-temporal fossa pushing the soft palate to one side (Figure 1A–B). The patient was admitted for definitive surgical management. He underwent excision of the lesion via para-median mandibular osteotomy for access to the lateral pharyngeal space, base of skull and infra-temporal fossa to completely remove the tumor while preserving the facial nerve. Postoperatively, the patient recovered uneventfully with no facial and/or, inferior alveolar nerve deficits and no osteotomy related complications resuming normal mandibular movements. Postoperative computed tomography follow-up after one year, also, revealed no signs of tumor recurrence (Figure 2).

Case 2

A 63-year old female reported to the unit with a gradually increasing, painless swelling on the left side of neck from last four months. The patient did not have any signs and symptoms of dysphagia and dysphonia.

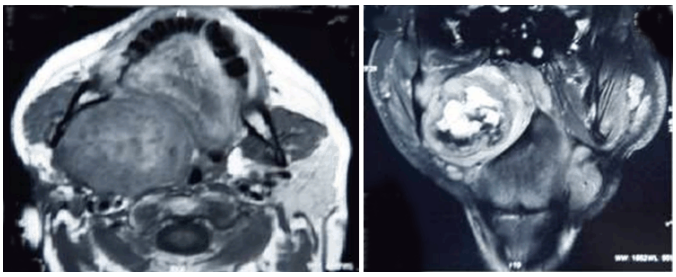


Figure 1: (A, B): Preoperative magnetic resonance imaging scan depicting a dense extensive homogeneous mass, 5cms in diameter, originating in the right deep parotid lobe and extensively involving the para-pharyngeal space, base of skull and infra-temporal fossa pushing the soft palate (Case 1).

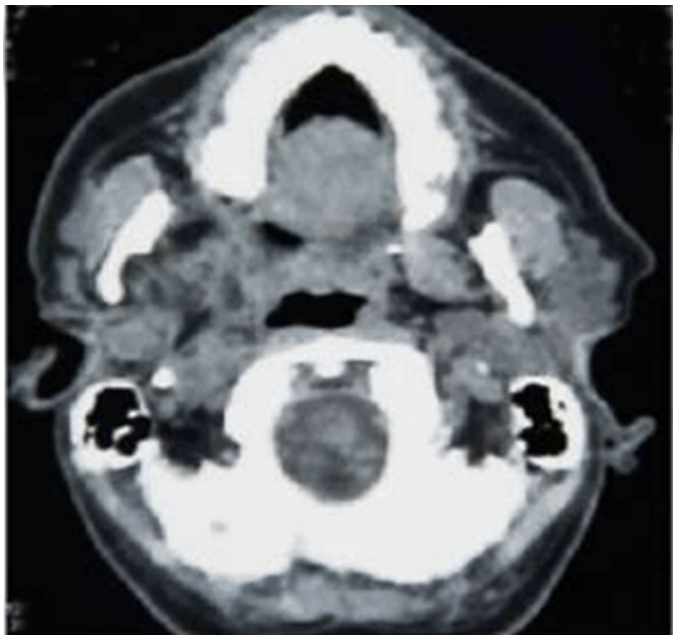


Figure 2: Postoperative computed tomography scan follow-up after one year with no signs of tumor recurrence (Case 1).

She had a history of superficial parotidectomy of left parotid gland 23 years back along with radiotherapy. On examination, a well-defined, 2 cm lesion was found in postauricular region which was fixed, non-tender, and firm in consistency. Another well-defined lesion of 2 cm dimension was present in the left submandibular region near angle of the mandible. Fine-needle aspiration cytology (FNAC) was suggestive of pleomorphic adenoma of deep lobe of parotid. Magnetic resonance imaging scan revealed a well-defined, large lobulated mass, extending from the left ramus of mandible with extension into the parapharyngeal space medially and another oval mass in left retro-auricular region superficial to sternocleidomastoid muscle with another lesion in the pre-auricular region (Figure 3A–B). Patient underwent surgical excision with para-median mandibular osteotomy to access the tumor of the deep lobe (Figure 4A–C). She withstood the procedure well with no residual functional and/or, esthetic deficits.

Technique of mandibular swing approach and excision of tumor: Under nasotracheal intubation, routine skin preparation and draping, the operation began with a pre-auricular incision which was carried-out anteriorly into the submandibular region reaching the mandibular symphysis. The trunk of the facial nerve was identified by using the insertion of the posterior belly of the digastric muscle on the mastoid process as a landmark and detaching the parotid lobe from the external auditory canal to identify the tragal pointer. The submandibular skin flap was dissected in sub-platysmal plane in the direction of the mandible and paying close attention to the preservation of the marginal mandibular branch of the facial nerve. Next, the periosteum was slit along the inferior mandibular border and in the premolar region, a subperiosteal dissection was done to allow a mandibular osteotomy to be made just anterior to the mental foramen preferably within the interproximal space between the canine and the first premolar. Titanium bone plates were fitted and screw holes were drilled before starting the osteotomy procedure to facilitate approximation of the mandibular fragments at the end of the procedure. The osteotomy was, then, started and the medial aspect of the mandible was freed up to the lingula region where the neurovascular bundle was isolated and preserved. This allowed lateral and superior dislocation of the body and ramus of the mandible exposing the parapharyngeal space tumor which was, then, easily mobilized and delivered. At this point, there was a wide exposure from below, laterally and anteriorly which helped to control areas of the tumor that might involve local structures such as the internal carotid artery, eustachian tube and the base of the skull. Tumor size and possible extensions were, also, assessed by finger dissection. Involvement of the mucosa is rare with neoplasms of the parotid parenchyma and hence, the mass was thoroughly enucleated from the adjacent connective tissue. Once the tumor was delivered, the osteotomized mandibular segments were re-approximated and stabilized with the adapted bone plates after ensuring the position of the condyle in the glenoid fossa. Hemostasis was achieved using bipolar cautery and incisions were reconstructed layer by layer and a suction drain was placed. Postoperative management included a short course of a broad-spectrum antibiotic which was prolonged in one of the patients wherein the integrity of the mucosa was compromised.

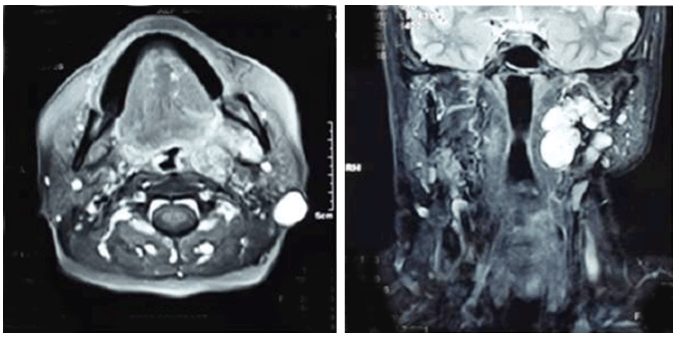


Figure 3: (A, B): Preoperative magnetic resonance imaging scan revealing well-defined large lobulated mass from left ramus of mandible with parapharyngeal space extension (Case 2).

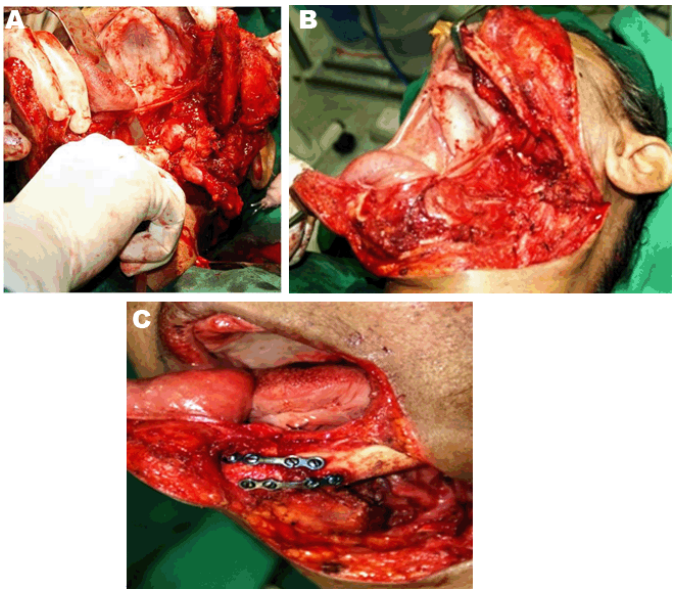


Figure 4 (A–C): Enucleation and excision of the entire mass in-toto with swing mandibulotomy with the mandible dislocated superiorly; fixation of the mandibular segments was carried-out with mini plates and screws (Case 2).

DISCUSSION

Pleomorphic adenomas are slow growing, well-demarcated tumors constituting around 80% of benign parotid tumors [1, 2]. The mainstay of parotid surgical procedures involves the cleavage of the gland along the two planes divided by the facial nerve [3]. Around 90% of the parotid tumors arise in the superficial lobe while approximately, 80–85% of the glandular tissue lies lateral to the facial nerve [1, 4, 5]. Approximately 10–12% of the parotid tumors arise from the deep lobe with only a small proportion (~1%) with a tendency to

extend medially towards the parapharyngeal space [1, 5, 6]. The complex anatomic relations of the deep lobe with adjacent structures often makes tumors involving this lobe and peripheral areas a definite surgical challenge for adequate approach [7, 8]. Although recurrent parotid pleomorphic adenomas have been the subject of much attention in the clinical literature, especially, pertaining to managerial issues, there is a relative paucity of information defining the imaging characteristics of these lesions in the radiologic literature [9–12]. Multiplicity of lesions has been described as a reliable indicator of recurrence in the parotidectomy bed of patients with prior pleomorphic adenoma resections [10]. Imaging is used to predict the origin, exact location and the size of parapharyngeal tumors. Magnetic resonance imaging scan with gadolinium enhancement is considered better than CT scan and is the examination of choice. It can reliably distinguish a deep lobe parotid tumor from a primary parapharyngeal tumor of neurovascular origin or, of extra-parotid minor salivary glands, from evidence, in T2-weighted slices, of the fatty layer between the tumor and the pharyngeal wall. USG-guided or, CT-guided fine-needle aspiration cytology (FNAC) is usually performed to determine the nature of the mass. According to data in literature, FNAC is accurate in 90–95% of cases. It is performed transorally, transcervically or, guided by USG or, CT scan and predicts the nature of the lesion which assists surgeon-patient planning. Management recommendations for recurrent tumors include observation, local or, extensive surgery, radiation therapy, or, a combination of these [9, 13–16]. Recurrences are often extensive and multifocal making the risk of facial nerve damage and chances of re-recurrence high [8, 13, 17]. Malignant transformation to carcinoma is a well-known entity and literature reports a rate of between 1.4% and 6.3% in the larger series of patients [18]. The risk of malignant transformation seems to be higher in patients older than 40 years of age, men, and patients with solitary nodules over 2 cm in diameter [8]. The risk of malignancy has, also, been linked to the frequency of recurrence [14, 19]. An accurate diagnosis is essential for planning the best surgical approach to safely and radically remove parapharyngeal space tumors.

Papadogeorgakis et al. [20] considered five main parameters in selecting the best approach in treating tumors of the PPS including the proximity and the projection of the tumor to the oropharyngeal wall or, the neck, the size of the tumor, the suspicion of malignancy, the vascularity and relation of the tumor to the neck neurovascular bundle. Different surgical techniques and mandibular osteotomy designs have been described to improve surgical access for the removal of parapharyngeal space tumors since Roux's first description of division of the lower lip and mandible in 1829 [9].

In 1929, Trotter W15 reported a median translingual pharyngotomy in which he extended Roux's midline section of the mandible by splitting the tongue through the midline for exposure of the base of the tongue and the mid portion of the pharynx. In 1984, Attia et al. [21] introduced the double mandibular osteotomy technique to significantly improve access to the para-pharyngeal space (PPS), while still protecting the inferior alveolar and lingual neurovascular bundles. Approaches described by Attia et al. [21] and Biedlingmaier et al. [22] involve splitting of the lip with attendant esthetic consequences. Approaches involving lateral neck dissection are indicated for large, diffuse tumors, but full surgical exposure is impeded by the presence of the mandible. Such a problem can be overcome by involving mandibular osteotomies. Simple mandibular distraction was proposed by Martin HE [23] in 1957. It sometimes entails resecting the styloid process and the stylomandibular ligament [24, 25] or, the posterior margin of the ramus of the mandible. This approach makes isolation of the superior portion of large neoplastic growths particularly difficult. It, also, increases the intraoperative risks of rupturing the tumor capsule and of injury to the larger vessels. Mandibulotomy, however, allows wider and safer access to the parapharyngeal space, with better control for adequate tumor excision. Various techniques involving osteotomy of the ramus [26, 27], the angle [28, 29], or, the body [5, 30] of the mandible have been described. All, however, involve resection of the inferior alveolar nerve within the bone except para-median mandibulotomy. To avoid sacrificing the mandibular neurovascular bundle, Pogrel and Kaplan MJ27 proposed a horizontal osteotomy of the ramus performed just above the lingula region. The inferior mandibular fragment can, then, be distracted downwards and a few centimeters of access space can, thus, be gained. Flood and Hislop [1] inverted-L osteotomy, which does preserve the inferior alveolar nerve, only partially improves on the intraoperative visibility as provided by Pogrel and Kaplan's [27] technique.

Increasing surgical exposure (while preserving mandibular nerve function) through a superficial parotid lobectomy followed by a mandibulotomy incision anterior to the mental foramen to access the parapharyngeal space has been proposed by Attia et al. [21]. Spiro et al. [10] describe an approach which provides adequate control both of the neoplasm and of the cervical vessels but compromises esthetic outcome because these authors deem a labiotomy to be necessary. Biedlingmaier et al. [22] stated that it is needed to allow full outward rotation of the mandibular body. Seward GR [5] in 1985, used approach consisting of a para-symphysis mandibulotomy, anterior to the mental foramen, with no cheilotomy or, opening of the floor of the mouth, and preserving the inferior alveolar nerve. Another possible variant is osteotomy at the level of the condyle or, more recently described, vertical osteotomy of the mandible to facilitate greater mobilization of the corresponding segment of the mandible. He, also, described the possibility of approaching the parapharyngeal space through a lateral neck dissection and a mandibulotomy anterior to the mental nerve without labiotomy (the technique chosen

in the present case series). Seward's approach provides a two-fold advantage over other techniques including an adequate field exposure from below, laterally, and anteriorly for radical tumor surgery, and an alternative osteotomy without labiotomy and sacrifice of the inferior alveolar nerve. In planning the surgical treatment of a benign neoplasm, these esthetic as well as functional considerations, should be foremost. The fitting of bone plates before making the mandibular osteotomy limits the need of maxillo-mandibular fixation to a few days of elastic guidance of the occlusion. This access allows the parapharyngeal space to be reached with relative ease, and provides excellent visibility with wide surgical exposure to secure local neurovascular structures and thereby, preserves both sensory and motor functions, without appreciable esthetic compromise. The median labiomandibulotomy was popularized by Martin et al. [16] in 1961. Since the first osteotomies were described by Ariel et al. [4], several variants [31] have been described. McGregor and MacDonald [17] described modifications of the osteotomy with the use of power saws and dental drills to avoid dental extractions. Mandibulotomy can be performed anterior to mental foramen-medial mandibulotomy or, posterior to mental foramen-lateral mandibulotomy. Lateral mandibulotomy is, nowadays, seldom used because the combination of radiation effect and loss of blood supply by dividing the inferior alveolar artery may result in the non-union or, osteoradionecrosis of the mandible [18]. Median mandibulotomy can be, further, classified into midline (median) mandibulotomy, between the two central incisors and para-midline (para-median) mandibulotomy, between the lateral incisor and canine [19].

In a classical midline (median) mandibulotomy with a mandibular swing approach, the genioglossus, geniohyoid and mylohyoid muscles have to be transected [10], however, in a para-midline (para-median) mandibulotomy with lateral mandibular swing, the centrally located genioglossus and geniohyoid muscles can be preserved [11]. According to Dubner Sanford et al. [32], median mandibulotomy (mandibular swing) supplanted mandibular resection for access to oral and oropharyngeal tumors when there is intervening grossly normal tissue between the tumor and the bone. It has, also, proved useful for exposure in selected patients with deep lobe parotid or, parapharyngeal space (PPS) tumors. The authors had reviewed 313 mandibulotomies performed between 1959 and 1988 with emphasis on indications, complications, and modifications in technique. In a comparative study between these two types, Tsung et al. [14] did not find any differences that were statistically significant. Amin MR et al. [12], however, concluded straight midline osteotomy to be advantageous with no significant complications seen within one-year follow-up. The midline, stair-step osteotomy was designed to provide better stability to the osteotomy site [33]. Yiwen et al. [34] resected five pharyngeal and parapharyngeal space tumors and showed that mandibular swing approach is an optimal technique for thoroughly removing retro-mandibular tumors of the parotid gland. The excisions did not injure the facial nerve nor led to any significant complications.

In all 5 cases, tumors were located in the parapharyngeal space while in one of the cases, the tumor protruded into the palate. During their five-year analysis, Jungehuelsing et al. [35] evaluated nine patients with extensive, deep lobe, benign, parotid gland tumors, who underwent lateral parotidectomy followed by a modified mandibular swing procedure without lip split. In all the patients, adequate exposure was obtained for total resection of the parapharyngeal space lesions with mandibular swing technique. Hye-Young Na et al. [13], reviewed retrospectively, 30 patients who had a uniform surgical technique consisting of a lower lip-splitting, modified stair-step osteotomy with at least 14 months postoperative follow-up during a five-year period. According to them, an ideal mandibulotomy technique required no intermaxillary fixation (IMF), preserved the occlusion in a precise way, enabled early function, entailed no secondary procedures, and was free of complications [36].

The main problem with the original mandibulotomy technique concerned the postoperative fragment mobility due to a bony gap on the osteotomy line. In spite of rigid flxation, the bony gap remains, and mobility of unstable fragments increases the probability of postoperative complications such as malocclusion, non-union, and osteoradionecrosis following postoperative irradiation. Mandibulotomy is an important surgical approach for tumors in the oral cavity, oro-pharynx, para-pharyngeal space, nasopharynx and skull base [14]. Based on these factors, mandibular swing approach has been considered to be an ideal technique to approach tumors in the pharyngeal and parapharyngeal space region avoiding blind and unnecessary injuries (Han et al. 2002 and Sun et al. 2006). Although, the significant concerns regarding the mandibular swing approach include incomplete concrescence of mandible, inarticulacy, deglutition obstacles, middle ear effusion and bleeding, and cranial nerve palsy (Wang et al., 1998). Abdel-Haleem et al. [8] has reported 11 patients diagnosed to have parapharyngeal space occupying lesions that were subjected to surgical excision by the trans-mandibular approach. In 2007 and 2009, Kolokythas et al. [37] reported the use of a double mandibular osteotomy without lip split to remove tumors of the parapharyngeal space (PPS). Double mandibular osteotomy facilitates the resection of even large and malignant neoplasms of the parapharyngeal space but the double osteotomy seems to create stability problems of the mandible and thus, postoperative occlusal derangement which has to be treated later, then, by lingual splints and arch bars. When considering a mandibular osteotomy procedure, complete removal of the tumor, maintenance of occlusion with acceptable temporomandibular joint function, and functional preservation of facial and inferior alveolar

nerves are the major considerations to be kept in mind. The decision to perform a mandibular osteotomy is based on several factors including tumor type, location, size, and inability to access the space adequately via the trans-cervical-trans-parotid, trans-cervical, orbito-zygomatic middle fossa and trans-mandibular and infra-temporal approaches and combinations thereof [38].

The final decision to perform a mandibulotomy is made once the trans-cervical or, trans-cervical-trans-parotid approach to the parapharyngeal space is performed and access for total tumor removal assessed. If the tumor can be removed with the trans-cervical or, trans-cervical-trans-parotid approach, then, a mandibulotomy is not performed. If the tumor cannot be adequately exposed via the trans-cervical or, trans-cervical-trans-parotid approach because of the involvement of the pre-styloid compartment of the parapharyngeal space such as with tumors that originate in the deep lobe of the parotid or, parapharyngeal space (PPS), or, ectopic salivary gland tissue, a single, anterior, symphyseal osteotomy is sufficient for parapharyngeal space access and tumor exposure. Removal of a mandibular incisor to allow for an anterior mandibular osteotomy is, also, recommended. A midline mandibular osteotomy has been described with a lip and soft tissue incision across the midline to provide access for exposure and placement of the rigid fixation. This is unnecessary for the mandibular swing technique. It is sufficient to expose the mandible, as described above, and not to perform lip and soft tissue split. The mandibular processes turn around an imaginary axis through the symphysis and mandible articulation has enough latitude to allow wide lateral deflection. Multiple techniques and combinations of osteotomies have been employed to facilitate access to the parapharyngeal space. In the present cases, the use of a single midline mandibular osteotomy technique was employed for exposure of deep lobe parotid gland tumors allowing mandibular swing and improved access to the para-pharyngeal space.

CONCLUSION

Swing mandibulotomy access (median mandibulotomy approach) has supplanted mandibular resection for access to oral and oro-pharyngeal tumors when there is intervening grossly normal tissue between the tumor and the underlying bone. It has, also, proved useful for exposure in selected patients with deep lobe parotid or, parapharyngeal space tumors. Swing mandibulotomy technique provides excellent access up to the base of skull with no significant morbidities. While employing swing mandibulotomy access to resect a deep lobe parotid or, parapharyngeal space tumors, the important goals considered should include preservation of facial nerve function, complete removal of the tumor without rupture, maintenance of the preoperative occlusion, functional temporomandibular joint movement, and preservation of the inferior alveolar nerve sensation. Furthermore, long-term follow-up of patients with pleomorphic adenoma, even confined to superficial lobe, plays a very important role as it has a high chance to recur in the deep lobe, even after 15–20 years, requiring follow-up for the lifetime.

****

Author Contributions

Pradipta Das – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Kavita Nivargi – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
S. Girish Rao – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Sushma Mehta – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Moumita Maity Rana – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Pritam Chandrakant Chaudhary – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Abhishek Singh Nayyar – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

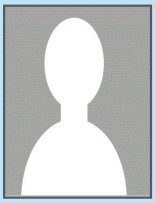
REFERENCES

1. Flood TR, Hislop WS. A modified surgical approach for parapharyngeal space tumours: Use of the inverted 'L' osteotomy. Br J Oral Maxillofac Surg 1991 Apr;29(2):82–6.
2. Ehrlich H. Mixed tumors of the pterygomaxillary

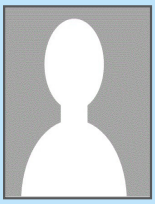
space; operative removal; oral approach. Oral Surg Oral Med Oral Pathol 1950 Nov;3(11):1366–71.
3. Morfit HM. Retromandibular parotid tumors: Their surgical treatment and mode of origin. AMA Arch Surg 1955 Jun;70(6):906–13.
4. Ariel IM, Jerome AP, Pack GT. Treatment of tumors of the parotid salivary gland. Surgery 1954 Jan;35(1):124–58.
5. Seward GR. Nodular enlargements of the salivary glands. In: Moore JR, Editor. Surgery of the Mouth and Jaws. Oxford: Blackwell Scientific Publications; 1985. p. 694.
6. Seward GR. Tumours of the parapharyngeal space. J R Coll Surg Edinb 1989 Apr;34(2):111–2.
7. Fisch U. Infratemporal fossa approach to tumours of the temporal bone and base of the skull. J Laryngol Otol 1978 Nov;92(11):949–67.
8. Abdel-Haleem A, El Sayed A, Hakeem HA. Trans-mandibular approach in para-pharyngeal tumors: When to do it? Egyptian Journal of Ear, Nose, Throat and Allied Sciences 2011;12:25–31.
9. Butlin HT. Diseases of the tongue: Clinical manuals for practitioners and students of medicine. London: Cassell and Company; 1885. p. 331–2.
10. Spiro RH, Gerold FP, Shah JP, Sessions RB, Strong EW. Mandibulotomy approach to oropharyngeal tumors. Am J Surg 1985 Oct;150(4):466–9.
11. Krespi YP, Sisson GA. Transmandibular exposure of the skull base. Am J Surg 1984 Oct;148(4):534–8.
12. Amin MR, Deschler DG, Hayden RE. Straight midline mandibulotomy revisited. Laryngoscope 1999 Sep;109(9):1402–5.
13. Na HY, Choi EJ, Choi EC, Kim HJ, Cha IH, Nam W. Modified mandibulotomy technique to reduce postoperative complications: 5-year results. Yonsei Med J 2013 Sep;54(5):1248–52.
14. Dai TS, Hao SP, Chang KP, Pan WL, Yeh HC, Tsang NM. Complications of mandibulotomy: Midline versus paramidline. Otolaryngol Head Neck Surg 2003 Jan;128(1):137–41.
15. Trotter W. Operations for malignant disease of the pharynx. Br J Surg 1929;16:485–95.
16. Martin H, Tollefsen HR, Gerold FP. Median labiomandibular glossotomy: Trotter's median (anterior) translingual pharyngotomy. Am J Surg 1961 Dec;102:753–9.
17. McGregor IA, MacDonald DG. Mandibular osteotomy in the surgical approach to the oral cavity. Head Neck Surg 1983 May-Jun;5(5):457–62.
18. Pan WL, Hao SP, Lin YS, Chang KP, Su JL. The anatomical basis for mandibulotomy: Midline versus paramidline. Laryngoscope 2003 Feb;113(2):377–80.
19. Dubner S, Spiro RH. Median mandibulotomy: A critical assessment. Head Neck 1991 Sep–Oct;13(5):389–93.
20. Papadogeorgakis N, Petsinis V, Goutzanis L, Kostakis G, Alexandridis C. Parapharyngeal space tumors: surgical approaches in a series of 13 cases3 Int J Oral Maxillofac Surg 2010 Mar;39(3):243-50.
21. Attia EL, Bentley KC, Head T, Mulder D. A new external approach to the pterygomaxillary fossa and parapharyngeal space. Head Neck Surg 1984 Mar–Apr;6(4):884–91.
22. Biedlingmaier JF, Ord R. Modified double mandibular osteotomy for tumors of the parapharyngeal space. J Oral Maxillofac Surg 1994 Apr;52(4):348–52.
23. Martin HE. Surgery of Head and Neck Tumors. New York: P.B. Hoeber; 1957.
24. Warrington G, Emery PJ, Gregory MM, Harrison DF. Pleomorphic salivary gland adenomas of the parapharyngeal space: Review of nine cases. J Laryngol Otol 1981 Feb;95(2):205–18.
25. Morita N, Miyata K, Sakamoto T, Wada T. Pleomorphic adenoma in the parapharyngeal space: Report of three cases. J Oral Maxillofac Surg 1995 May;53(5):605–10.
26. Bass RM. Approaches to the diagnosis and treatment of tumors of the parapharyngeal space. Head Neck Surg 1982 Mar–Apr;4(4):281–9.
27. Pogrel MA, Kaplan MJ. Surgical approach to the pterygomaxillary region. J Oral Maxillofac Surg 1986 Mar;44(3):183–7.
28. Berdal P, Hall JG. Para-pharyngeal growth of parotid tumors. Acta Otolaryngol 1970;263:164–6.
29. Carr RJ, Bowerman JE. A review of tumours of the deep lobe of the parotid salivary gland. Br J Oral Maxillofac Surg 1986 Jun;24(3):155–68.
30. Shaheen OH. Problems in Head and Neck Surgery. London: Bailliere Tindall; 1984. p. 70–91.
31. Fernández Ferro M, Fernández Sanromán J, Costas López A, Sandoval Gutiérrez J, López de Sánchez A. Surgical treatment of benign parapharyngeal space tumours. Presentation of two clinical cases and revision of the literature. Med Oral Patol Oral Cir Bucal 2008 Jan 1;13(1):E61–4.
32. Dubner S, Spiro RH. Median mandibulotomy: A critical assessment. Head Neck 1991 Sep–Oct;13(5):389–93.
33. Cohen JI, Marentette LJ, Maisel RH. The mandibular swing stabilization of the midline mandibular osteotomy. Laryngoscope 1988 Oct;98(10):1139–42.
34. Yiwen Ma, Liying Li, Chuanliang Du. Mandible Swing Approach for Excision of Tumors from Parapharyngeal Space. Modern Applied Science 2008; 2:167–70.
35. Jungehuelsing M, Guntinas-Lichius O, Klussmann JP, Eckel HE, Stennert E. Modifications of the midline mandibulotomy for access to the parapharyngeal space. Laryngoscope 2010 Aug;120(8):1557–62.
36. Shinghal T, Bissada E, Chan HB, et al. Medial mandibulotomies: Is there sufficient space in the midline to allow a mandibulotomy without compromising the dentition? J Otolaryngol Head Neck Surg 2013 May 2;42:32.
37. Kolokythas A, Eisele DW, El-Sayed I, Schmidt BL. Mandibular osteotomies for access to select parapharyngeal space neoplasms. Head Neck 2009 Jan;31(1):102–10.
38. Raveh E, Segal K, Chaimoff M, Feinmesser R. Surgical approaches to the para-pharyngeal space. Oper Tech Otolaryngol Head Neck Surg 1996;7:327–32.

ABOUT THE AUTHORS

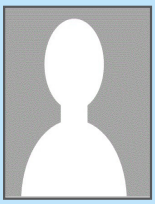
Article citation: Das P, Nivargi K, Rao G, Mehta S, Rana MM, Chaudhary PC, Nayyar AS. Swing mandibulotomy access to deep lobe parotid tumors: A case series. Int J Case Rep Images 2017;8(8):501–509.



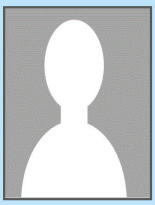
Pradipta Das is Assistant Professor, Department of Surgery, Kalinga Institute of Medical Sciences, Patia, Bhubaneswar, Odisha, India.



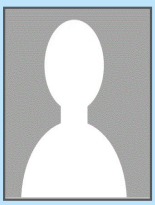
Kavita Nivargi is Reader, Department of Anaesthesia, Pandit Deendayal Upadhyay Dental College and Hospital, Solapur, Maharashtra, India.



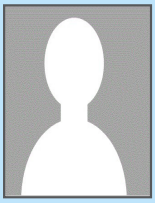
S. Girish Rao is Former Professor and Head, Department of Oral and Maxillofacial Surgery, D.A.P.M.R.V. Dental College and Hospital, J.P. Nagar, 1st Phase, Bangalore, Karnataka, India.



Sushma Mehta is Former postgraduate student, Department of Oral and Maxillofacial Surgery, D.A.P.M.R.V. Dental College and Hospital, J.P. Nagar, 1st Phase, Bangalore, Karnataka, India.



Moumita Maity Rana is postgraduate Student, Department of Oral and Maxillofacial Surgery, Awadh Dental College and Hospital, Dhule, Maharashtra, India



Pritam Chandrakant Chaudhary is postgraduate Student, Department of Oral and Maxillofacial Surgery, ACPM Dental College and Hospital, Jamshedpur, Jharkhand, India

Abhishek Singh Nayyar is Reader cum PG Guide, Department of Oral Medicine and Radiology, Saraswati Dhanwantari Dental College and Hospital and Post-Graduate Research Institute, Parbhani, Maharashtra, India.

CASE SERIES

PEER REVIEWED | OPEN ACCESS

Throat discomfort: A harbinger of a lethal diagnosis

Kevin Rajakariar, Anoop Koshy, Jithin Sajeev, George Proimos

ABSTRACT

Atypical presentations of acute aortic dissection are associated with delayed diagnosis and increased mortality rates. We describe two cases of isolated throat discomfort as the only symptom of a Stanford type A aortic dissection. In the first case, a 77-year-old male presented with isolated throat discomfort. After delayed recognition of aortic dissection, computed tomography (CT) aortography confirmed type A aortic dissection extending into the great vessels. He suffered a cardiac arrest and died. In second case, a 57-year-old male who described chest pain and a coronary angiogram was unremarkable. Post-procedure, he complained of severe throat pain and due to early suspicion of aortic dissection, an urgent CT aortography confirmed the diagnosis. He subsequently underwent successful graft replacement. The contrasting case series illustrates the importance of considering throat discomfort as an early manifestation of aortic dissection.

Keywords: Aortic dissection, Sore throat, Throat discomfort, Type A

Kevin Rajakariar[1], Anoop Koshy[1], Jithin Sajeev[1], George Proimos[1]

Affiliation: Department of Cardiology, Box Hill Hospital, Eastern Health, Victoria.

Corresponding Author: Dr. Kevin Rajakariar, 8 Arnold St, Box Hill, 3128, VIC; Email: kevin.rajakariar2@easternhealth.org.au

Received: 20 May 2017
Accepted: 05 June 2017
Published: 01 August 2017

How to cite this article

Rajakariar K, Koshy A, Sajeev J, Proimos G. Throat discomfort: A harbinger of a lethal diagnosis. Int J Case Rep Images 2017;8(8):510–513.

Article ID: Z01201708CS10091KR

****

doi:10.5348/ijcri-201712-CS-10091

INTRODUCTION

Clinical diagnosis of acute aortic dissection (AAD) is often challenging in the emergency room. Classical symptoms are uncommon and atypical presentations are associated with delayed diagnosis and an incremental rise in mortality approaching 2% for every hour following the onset of symptoms [1]. Acute aortic syndromes typically present with sudden onset of 'tearing' chest, abdominal or back pain [2]. In a real-world setting, however, absence of these pathognomonic symptoms can make AAD a 'clinical chameleon'. Awareness of atypical symptoms and early diagnosis correlated with improved clinical outcomes [2, 3]. A review by the European Society of Cardiology found 30% of patients to be misdiagnosed prior to the discovery of aortic dissection [4], and almost half of these were labeled and treated as an acute coronary syndrome [5]. Numerous atypical symptoms have been described, including trapezius ridge pain, fever, paresthesia and hiccups [6–9]. In this case series, we describe two reports of Stanford type A aortic dissection presenting with isolated throat discomfort.

CASE SERIES

Case 1

A previously healthy 77-year-old male, presenting with isolated sudden onset throat discomfort. He had no history of hypertension, relevant family history or other cardiovascular risk factors. Examination demonstrated bradycardia at a rate of 38 beats per minute with no radio-radial delay and although mildly hypertensive, there was no clinically significant brachial blood pressure difference. An ECG showed transient high grade atrioventricular block and serial troponins were within normal limits (Figure 1A). A transthoracic echocardiogram (TTE) was performed later that day which demonstrated a dilated 5.9 cm ascending aorta, subsequently an urgent computed tomography (CT) aortography revealed an extensive type a aortic dissection into the brachiocephalic, left common carotid and subclavian arteries in addition to bilateral external iliac, common iliac, and common femoral arteries. He progressed to acute aortic rupture and died as a result of hemorrhagic shock following the CT scan (Figure 1B).

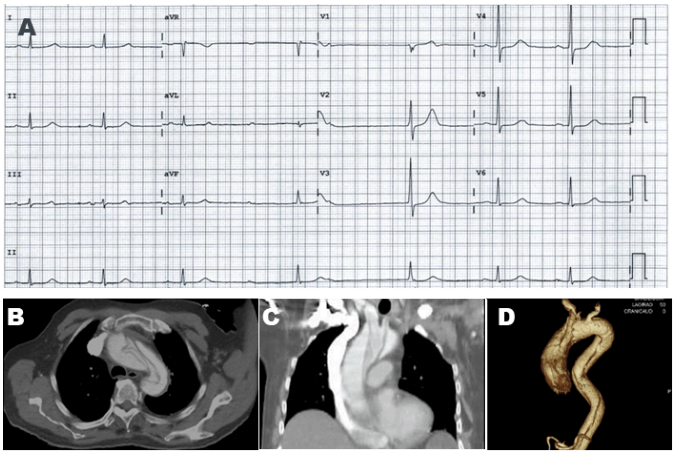


Figure 1: (A) Electrocardiogram demonstrating high grade atrioventricular block, (B) Contrast-enhanced computed tomography demonstrating intimal flap, (C) Diagnostic of Stanford type A aortic dissection from the aortic root with extension into the supra-aortic vessels, and (D) 3D reconstructed images show the spiral false lumen in the descending aorta.

Case 2

A 57-year-old male presented with bilateral arm paresthesia on a background of known hypertension and hypercholesterolemia, treated with irbesartan, hydrochlorothiazide and simvastatin. He had no family history of aortic syndromes or vasculitis. Blood pressure reading was elevated at 150/90, but was equal bilaterally, and an ECG demonstrated ST elevation in aVR with reciprocal ST depression in the anterior, inferior, and lateral leads (Figure 2A). High sensitivity troponin was mildly elevated at 20 ng/L (normal range <14 ng/L). He underwent urgent transradial coronary angiography which showed no flow limiting coronary artery stenosis, but was commenced on dual anti-platelets without heparinization. Following the procedure, his symptoms and ECG changes resolved, however he developed intense throat discomfort. The possibility of aortic dissection was raised and CT aortography confirmed type A aortic dissection from the aortic cusps extending into the brachiocephalic, left common carotid and subclavian arteries (Figure 2B). He subsequently underwent a successful aortic graft replacement.

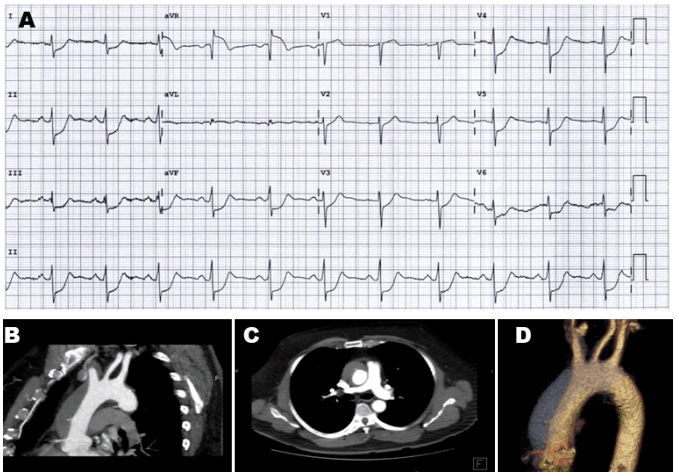


Figure 2: (A) ECG demonstrating aVR ST elevation with reciprocal ST depression in leads I, II, III, aVF, V3-V6, (B, C) Contrast-enhanced CT demonstrating type A dissection extending from the aortic valve to the origin of the descending aorta, (D) A small amount of arterial contrast can be seen in the false lumen which may represent a fenestration.

DISCUSSION

Branch vessel occlusion ischemia typically causes chest and back pain. However, up to 10% of patients with aortic dissection lack these cardinal features [5]. An incremental mortality rate following the onset of symptoms highlights the importance of making an early diagnosis in patients with acute aortic syndromes [3]. In a subset of the patients, throat pain may be the sole feature to prompt consideration of AAD. This has only been reported twice in literature [8, 9]. The throat discomfort is likely attributable to propagation of the dissection plane to the great vessels, as was the case in both patients. Other possible pathophysiological mechanisms may include lower cranial nerve irritation of the cardiac branch of the left vagus nerve secondary to supra-aortic vessel compression [10].

Throat discomfort is a clinically significant symptom that could aid in early diagnosis of acute aortic dissection in a patient where other typical symptoms are absent. This is particularly significant in cases where patients are misdiagnosed as an acute coronary syndrome,

as a combination of antiplatelet pre-loading and heparinization in the emergency department, followed by a coronary angiogram can often be fatal due to the risk of guidewire induced extension of the dissection plane. While not all throat pain equates to an aortic dissection, in the clinical situation where the symptom does not match the overall clinical state warrants consideration of the diagnosis.

CONCLUSION

In the clinical situation where the symptom does not match the overall clinical state warrants consideration of the diagnosis, while not all throat pain equates to aortic dissection.

****

Acknowledgements
We would like to thank the patients for allowing us to use these cases to help widen our knowledge. No financial assistance was provided for the study.

Author Contributions
Kevin Rajakariar – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Anoop Koshy – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Jithin Sajeev – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
George Proimos – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

REFERENCES

1. Tsai TT, Nienaber CA, Eagle KA. Acute aortic syndromes. Circulation 2005 Dec 13;112(24):3802–13.
2. Harris KM, Strauss CE, Eagle KA, et al. Correlates of delayed recognition and treatment of acute type A aortic dissection: The international registry of acute aortic dissection (IRAD). Circulation 2011 Nov 1;124(18):1911–8.
3. Nienaber CA, Eagle KA. Aortic dissection: New frontiers in diagnosis and management: Part I: From etiology to diagnostic strategies. Circulation 2003 Aug 5;108(5):628–35.
4. Erbel R, Alfonso F, Boileau C, et al. Diagnosis and management of aortic dissection. Eur Heart J 2001 Sep;22(18):1642–81.
5. Hansen MS, Nogareda GJ, Hutchison SJ. Frequency of and inappropriate treatment of misdiagnosis of acute aortic dissection. Am J Cardiol 2007 Mar 15;99(6):852–6.
6. Holicek S. Hiccup as a major sign of aortic dissection. Int J Cardiol 2014 Dec 20;177(3):e105–6.
7. Kondo T, Uehara T, Ikegami A, Hirota Y, Ikusaka M. Bilateral trapezius ridge pain with sustained low-grade fever caused by aortic dissection. Can J Cardiol 2015 Sep;31(9):1204.e17–8.
8. Liu WP, Ng KC. Acute thoracic aortic dissection presenting as sore throat: Report of a case. Yale J Biol Med 2004 May;77(3–4):53–8.
9. Anguille S, Derweduwen AM, Lenz J, Vanuytsel L, Cools FJ. Sore throat: A trivial complaint masking a life-threatening condition. Med J Aust 2009 Apr 20;190(8):454–6.
10. Myers DE. Vagus nerve pain referred to the craniofacial region: A case report and literature review with implications for referred cardiac pain. Br Dent J 2008 Feb 23;204(4):187–9.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Respiratory papillomatosis in a case of Carmi syndrome: An unusual presentation

Sandipta Mitra, Sayan Hazra, Arunabha Sengupta

ABSTRACT

Introduction: Congenital pyloric atresia with junctional epidermolysis bullosa and congenital cutis aplasia is known as Carmi syndrome. Case Report: We report a case of squamous papilloma of the respiratory tract with posterior urethral valve in a case of Carmi syndrome. Conclusion: Such presentation is extremely unusual and has not been documented yet.

Keywords: Carmi syndrome, Epidermolysis bullosa, Posterior urethral valve, Pyloric atresia, Respiratory papilloma

How to cite this article

Mitra S, Hazra S, Sengupta A. Respiratory papillomatosis in a case of Carmi syndrome: An unusual presentation. Int J Case Rep Images 2017;8(8):514–518.

Article ID: Z01201708CR10812MS

****

Sandipta Mitra[1], Sayan Hazra[1], Arunabha Sengupta[2]

Affiliations: [1]Junior Resident, Department of Otorhinolaryngology and Head Neck Surgery, Institute of Post Graduate Medical Education and Research, Kolkata, West Bengal, India; [2]Professor and Head of the Department, Department of Otorhinolaryngology and Head Neck Surgery, Institute of Post Graduate Medical Education and Research, Kolkata, West Bengal, India.

Corresponding Author: Dr. Sandipta Mitra, 618, 'O' Block, New Alipore, Kolkata, West Bengal, India, 700053; Email: onlysandipta@gmail.com

Received: 12 March 2017
Accepted: 02 May 2017
Published: 01 August 2017

doi:10.5348/ijcri-201773-CR-10812

INTRODUCTION

Carmi syndrome is an extremely rare disease manifesting with congenital pyloric atresia and junctional epidermolysis bullosa, with the inheritance pattern being autosomal recessive [1, 2].

CASE REPORT

A preterm male infant was delivered by lower uterine cesarean section, with a history of polyhydramnios in the mother. The newborn infant developed repeated episodes of vomiting with regurgitation of feeds since birth. A diagnosis of congenital pyloric atresia was made for which he underwent gastroduodenostomy (Figure 1) on the fifth day of life. He also developed bullous lesions over the trunk and extremities, with new lesions developing with trivial trauma (Figure 2, 3). At six months of age, the child developed poor stream of urine with discharge of whitish flecks with fever. Urine culture was positive for *Pseudomonas aeruginosa*, sensitive to gentamicin. Ultrasound showed hydronephrotic changes in both kidneys with dilatation of bilateral ureters. He was diagnosed to have posterior urethral valve (Figure 4) for which vesicostomy (Figure 5) had to be done after attempts of cystoscopy guided valve fulguration were deemed ineffective in a setting of recurrent urinary tract infection and hydronephrosis. The child presented with respiratory distress and hoarseness at four and a half years of age. Thereafter, direct laryngoscopy was done which revealed multiple masses occupying bilateral false cords, left aryepiglottic fold and anterior commissure (Figure 6). The masses were firm, fleshy, pedunculated and did not bleed on touch. Biopsy was taken and surgical excision of the masses was done. The histopathology examination revealed squamous papilloma (Figure 7). The child again presented with acute severe respiratory distress with cyanosis two months following discharge,

for which emergency tracheostomy had to be done. Attempts of weaning were tried but the child was unable to tolerate it. At present, the child is 9.5-year-old, tracheostomized, school-going with age-appropriate neurodevelopment. His 6.5-year-old sister underwent gastroduodenostomy for congenital pyloric atresia on 21st day of life and vesicostomy for posterior urethral valve at two years of age. She was also diagnosed with junctional epidermolysis bullosa at four years of age. The mother of the patient also suffered from polyhydramnios during the birth of her second child. There is a history of sibling death within first week of life in his father's generation following a blistering disorder.

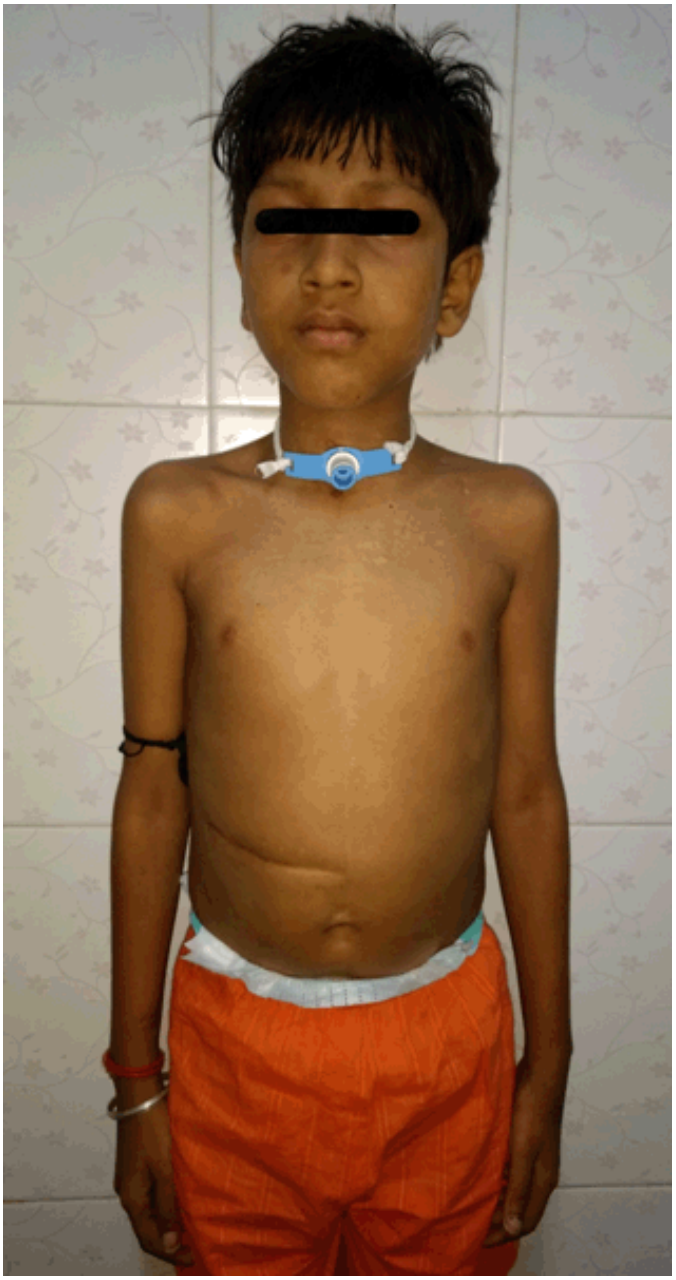


Figure 1: Child with tracheostomy tube in situ with scar of gastroduodenostomy incision.

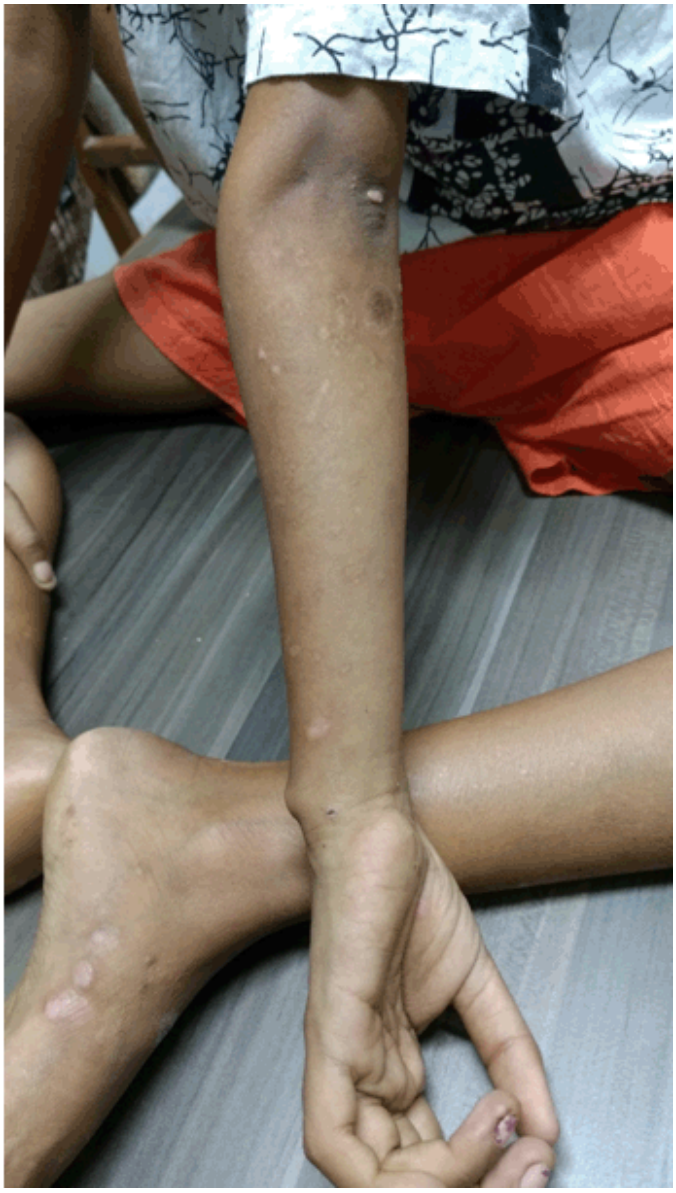


Figure 2: Healed scars of ruptured bullosa over extremities.

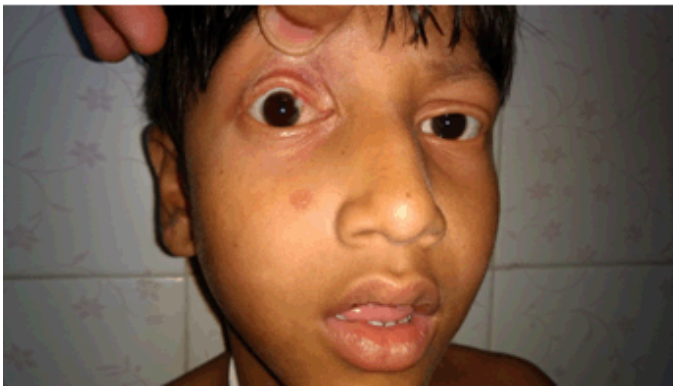


Figure 3: Bullosa involving right limbal conjunctiva.

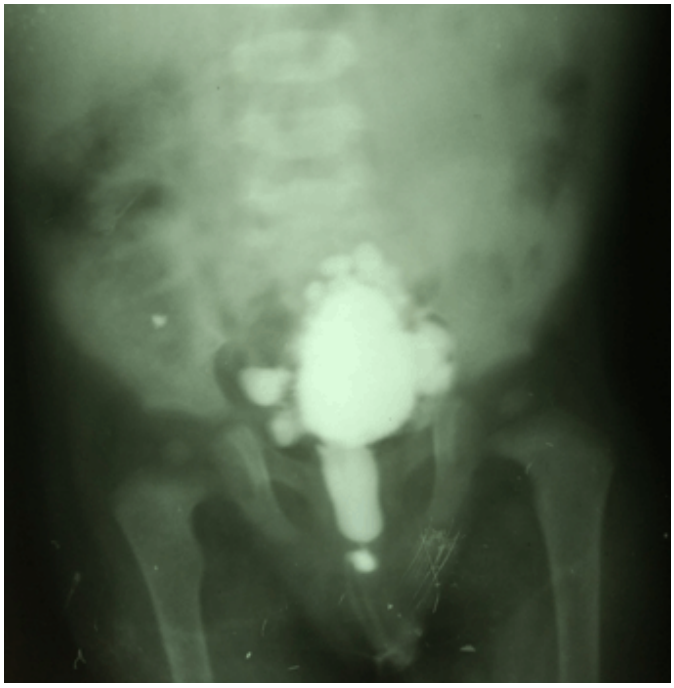


Figure 4: Voiding cystourethrogram showing dilatation and elongation of posterior urethra and bladder trabeculation.

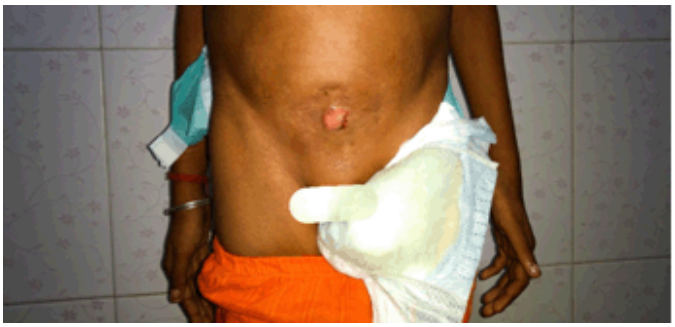


Figure 5: Scar of vesicostomy.

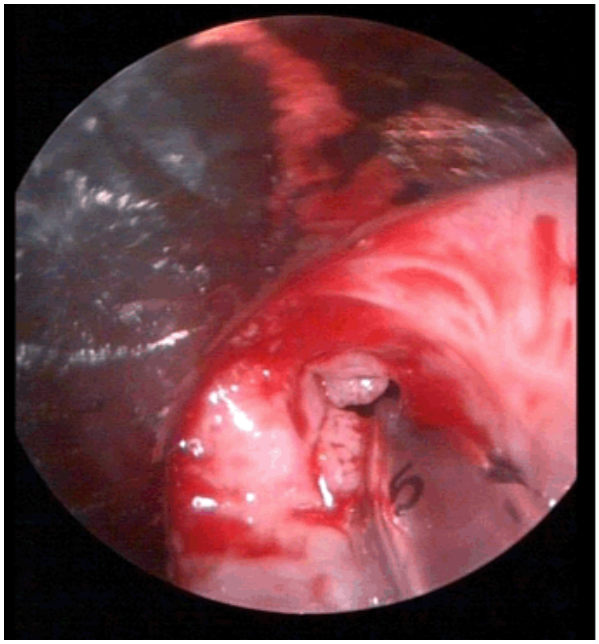


Figure 6: Fleshy masses in bilateral false cords, left aryepiglottic fold and anterior commissure.

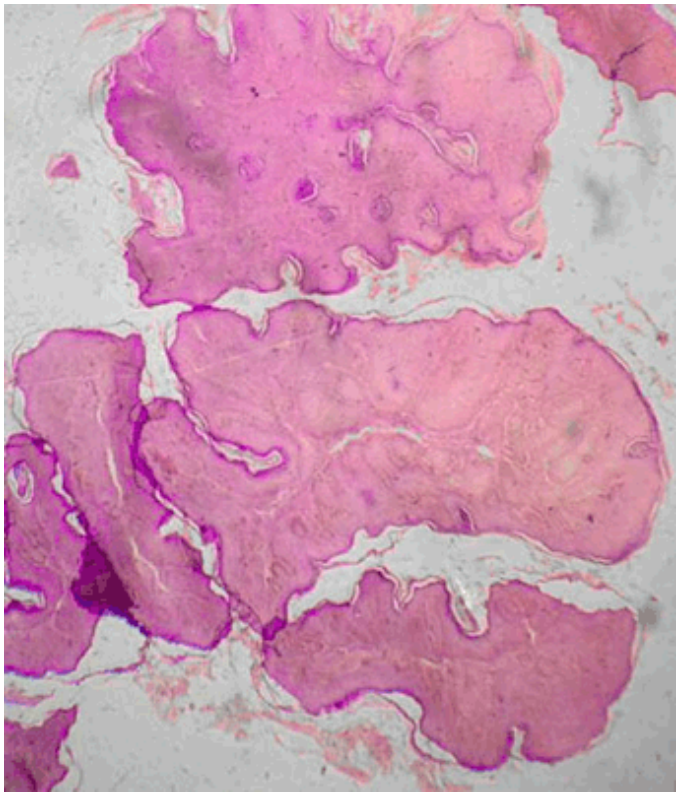


Figure 7: Photomicrograph showing multiple squamous papillae (H&E stain, x400).

DISCUSSION

Respiratory papillomatosis is a benign tumor of the respiratory tract, usually caused by human papilloma virus 6 or 11 infection, often presenting with hoarseness of voice, stridor and acute airway obstruction requiring emergency tracheostomy. The reported incidence of recurrent respiratory papillomatosis in a population based Danish study is 3.62 per 100,000 [3]. Such population based data in the Indian subcontinent is limited. Hence, to strengthen the database, a national registry [4] has been introduced. Surgical excision of the papillomas remains the mainstay of treatment, though medical therapy including intralesional antivirals [5] and HPV vaccines [6] have also been tried. Here we report an unusual case of respiratory papillomatosis and posterior urethral valve in addition to junctional epidermolysis bullosa with congenital pyloric atresia, which constitute Carmi syndrome. Junctional epidermolysis bullosa with congenital pyloric atresia has been associated with mutation in the α6β4 integrin genes (ITGA6, ITGB4) [2], leading to the formation of blisters which rupture on mechanical insults. Congenital heart disease [7] has also been reported in Carmi syndrome. Peptic perforation [8] and enterocolitis [9] are known complications of Carmi syndrome. The prenatal diagnosis of Carmi syndrome is made by ultrasonographic findings like polyhydramnios, gastric dilatation, snowflake sign in amniotic fluid and complete separation of chorioamniotic

membrane [10]. Preimplantation genetic detection [11] and immunofluorescence assisted villous trophoblast analysis [12] have also been as diagnostic tools. Although management of this rare disorder is early diagnosis and timely symptomatic intervention, gene therapy may yield promising results and creates ample scopes of research.

CONCLUSION

Respiratory papillomatosis is a common disease in the pediatric population that may have syndromic association as highlighted in the case report above. Hence formulation of a multidisciplinary approach to the diagnosis of respiratory papillomatosis with thorough systemic examination, keeping such multi-system affection in mind is imperative to rule out any syndromic association which may be often missed. Moreover, further studies may help to expand the spectrum of symptoms that define this rare disease Carmi syndrome.

****

Author Contributions

Mitra Sandipta – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Hazra Sayan – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Sengupta Arunabha – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

REFERENCES

1. Carmi R, Sofer S, Karplus M, et al. Aplasia cutis congenita in two sibs discordant for pyloric atresia. Am J Med Genet 1982 Mar;11(3):319–28.
2. Mutlu M, Kalay E, Dilber B, et al. Pyloric atresia-junctional epidermolysis bullosa syndrome showing novel c.4505-4508insACTC mutations in integrin b4 gene (ITGB4). Turk J Pediatr 2015 Jul–Aug;57(4):385–7.
3. Larson DA, Derkay CS. Epidemiology of recurrent respiratory papillomatosis. APMIS 2010 Jun;118(6–7):450–4.
4. Mishra A, Singh DB, Verma V. Recurrent respiratory papillomatosis: The need for an Indian national registry. J Laryngol Voice 2011;1(5):36–7.
5. Murono S, Nakanishi Y, Tsuji A, et al. Intralesional cidofovir injection for recurrent respiratory papillomatosis in Japan. Auris Nasus Larynx 2016 Oct;43(5):541–5.
6. Hermann JS, Weckx LY, Monteiro Nürmberger J, et al. Effectiveness of the human papillomavirus (types 6, 11, 16, and 18) vaccine in the treatment of children with recurrent respiratory papillomatosis. Int J Pediatr Otorhinolaryngol 2016 Apr;83:94–8.
7. Aydin M, Zenciroglu A, Yaman A, et al. Carmi syndrome with congenital heart defects. Am J Med Genet A 2010 Aug;152A(8):2120–2.
8. Joshi M, Krishnan L, Kuruvila S. Large gastric perforation in carmi syndrome: A morbid complication in a rare association. J Neonatal Surg 2012 Oct 1;1(4):57.
9. Prabhu V, Sankar J, Srinivasan A, Sathiyasekaran M. Desquamative enterocolitis: An intestinal variant of Carmi syndrome presenting as protein-losing enteropathy. Indian J Gastroenterol 2008 Sep–Oct;27(5):215–6.
10. Dural O, Acar DK, Ekiz A, et al. Prenatal ultrasound findings and a new ultrasonographic sign of epidermolysis bullosa with congenital pyloric atresia: A report of three cases. J Med Ultrason (2001) 2014 Oct;41(4):495–8.
11. Ozge A, Safak H, Ebru H, et al. First successful preimplantation genetic diagnosis of epidermolysis bullosa with pyloric atresia: Case study of a novel c.4505-4508insACTC mutation. J Assist Reprod Genet 2012 Apr;29(4):347–52.
12. D'Alessio M, Zambruno G, Charlesworth A, Lacour JP, Meneguzzi G. Immunofluorescence analysis of villous trophoblasts: A tool for prenatal diagnosis of inherited epidermolysis bullosa with pyloric atresia. J Invest Dermatol 2008 Dec;128(12):2815–9.

ABOUT THE AUTHORS

Article citation: Mitra S, Hazra S, Sengupta A. Respiratory papillomatosis in a case of Carmi syndrome: An unusual presentation. Int J Case Rep Images 2017;8(8):514–518.

Sandipta Mitra is pursuing residency at the Department of Otorhinolaryngology and Head Neck Surgery at IPGMER, Kolkata. Her areas of interest include head neck oncology and anterior skull base surgery.
Email: onlysandipta@gmail.com

Sayan Hazra is pursuing residency at the Department of Otorhinolaryngology and Head Neck Surgery at IPGMER, Kolkata. His areas of interest include minimally invasive otologic intervention techniques and phonosurgery.
Email: sayanhazra@yahoo.co.in

Arunabha Sengupta is the Head of the Department, Department of Otorhinolaryngology and Head Neck Surgery, IPGMER, Kolkata. His areas of interest include neurotology and head neck oncology. He has published several papers in national and international journals.

CASE REPORT PEER REVIEWED | OPEN ACCESS

A case of isolated ST segment elevation in augmented vector right secondary to occluded or under perfused STOUP-collateral circulation

Edward Rojas, Molly Malone, George Stoupakis

ABSTRACT

Introduction: Lead augmented vector right (aVR) has been historically underestimated in the evaluation of acute coronary syndromes in the emergency department and other clinical settings. ST-elevation in aVR has shown to be a predictive marker of critical stenosis of the left main coronary and is associated with increased 30 day mortality. Case Report: We report an unusual case of acute ST-elevation myocardial infarction (STEMI) consistent with left main disease presenting with aVR ST-elevation in a patient with ischemic stroke. During percutaneous coronary intervention (PCI), the patient was found to have a total occlusion of the distal left main coronary artery (LMCA) which was collateralized from a large, dominant right coronary artery (RCA) which had no obstructive lesions. We hypothesize that the electrocardiography (ECG) are secondary to hypotension-related hypoperfusion of RCA collateral circulation supplying the anterior wall, in the setting of a chronically occluded LMCA. The patient succumbed to his disease eleven days after presentation. Conclusion: Our purpose is to raise awareness of the high mortality associated with this ECG abnormality. Prompt recognition of isolated aVR ST changes would allow for identification of high risk patients leading to an early invasive approach with appropriate activation of the catheterization laboratory.

Keywords: Augmented vector right (aVR) lead, Collateral circulation, Left main coronary artery, ST-elevation myocardial infarction (STEMI)

How to cite this article

Rojas E, Malone M, Stoupakis G. A case of isolated ST segment elevation in augmented vector right secondary to occluded or under perfused STOUP-collateral circulation. Int J Case Rep Images 2017;8(8):519–522.

Article ID: Z01201708CR10813ER

****

doi:10.5348/ijcri-201774-CR-10813

Edward Rojas[1], Molly Malone[2], George Stoupakis[3]

Affiliations: [1]MD, Department of Medicine, Rutgers, New Jersey Medical School, Newark, New Jersey; [2]MD, Department of Emergency Medicine, Hackensack University Medical Center, Hackensack, New Jersey; [3]MD, FACC, Department of Cardiology, Hackensack University Medical Center, Hackensack, New Jersey.

Corresponding Author: George Stoupakis, MD, Department of Cardiology, Hackensack University Medical Center, Hackensack, New Jersey; Email: George.stoupakis@hackensackmeridian.org

Received: 20 March 2017
Accepted: 02 May 2017
Published: 01 August 2017

INTRODUCTION

The utility of lead augmented vector right (aVR) in the evaluation of acute coronary syndrome has been historically underestimated in clinical practice [1]. Nonetheless, ST elevation isolated to lead aVR and especially in combination with other ischemic changes, has been shown to be a predictive marker of critical stenosis of the left main coronary artery. Augmented vector right ST-elevation has been associated with a higher 30-day mortality that was independent of concomitant ST segment changes in other leads [2]. The increased mortality of this condition is demonstrated in this case

report; our patient succumbed to his disease eleven days after presentation to the emergency department.

We report an unusual case of acute ST-elevation myocardial infarction consistent with left main disease presenting with aVR ST elevation in the setting of hypotensive collateral insufficiency in a patient with ischemic stroke. This case highlights the importance of recognizing an unusual pattern on electrocardiography (ECG) for early detection of left main disease. Also demonstrates, a case of ST elevations due an occluded or under perfused collateral circulation (STOUP- collateral circulation).

CASE REPORT

A 75-year-old Asian male with prior history of cerebrovascular accident, type 2 diabetes, hypertension and dementia was brought to the emergency department with a chief complaint of aphasia, left sided facial droop and right sided hemiparesis. Symptoms began acutely at 09:00 a.m. on the day of admission when returning home from church. In the emergency department, blood pressure was initially 137/72 mmHg, heart rate of 86bpm, and electrocardiogram (ECG) showed 0.1 mV ST elevations in aVR and V1, and ST-depression in lead I and II (Figure 1A). Computed tomography scan of head without contrast showed no acute intracranial hemorrhage and the patient was deemed an appropriate candidate for t-PA. After initial t-PA bolus, patient was started on a t-PA infusion. Due to worsening hypertension reaching 200/88 mmHg, he was started on a titratable nicardipine infusion. Transiently, the patient experienced an episode of nausea and vomiting and became hypotensive to 70/45 mmHg leading to the discontinuation of nicardipine. Shortly thereafter, the patient was noted to have new ST changes on telemetry monitor. A second ECG (Figure 1B) confirmed a 0.3 mV ST-elevation in aVR and V1 with reciprocal changes in lead II, III and aVF. Normal ECG shown in Figure 1C. Bedside point of care ultrasound (POCUS) showed anterior and apical hypokinesis. The patient subsequently developed impending respiratory failure secondary to cardiogenic shock and pulmonary edema.

The patient was taken directly to the catheterization laboratory where he was found to have a total occlusion of the distal left main coronary artery (LMCA) which was collateralized from a large, dominant right coronary artery (RCA) that had no obstructive lesions. To confirm that left circulation was chronically occluded, we attempted to wire into the left anterior descending (LAD) artery, but the wire could not cross through the left main stenosis, confirming a chronic occlusion (Figure 2A).

Treatment, outcome and follow-up

This patient developed a STEMI due to a totally occluded LMCA and hypotensive collateral insufficiency of the RCA in the setting of treatment of an acute ischemic stroke. His coronary angiogram showed a chronically occluded distal LMCA with a dominant RCA and significant collateral circulation (Figure 2B). The patient did not require any immediate revascularization in the catheterization laboratory due to improved mean

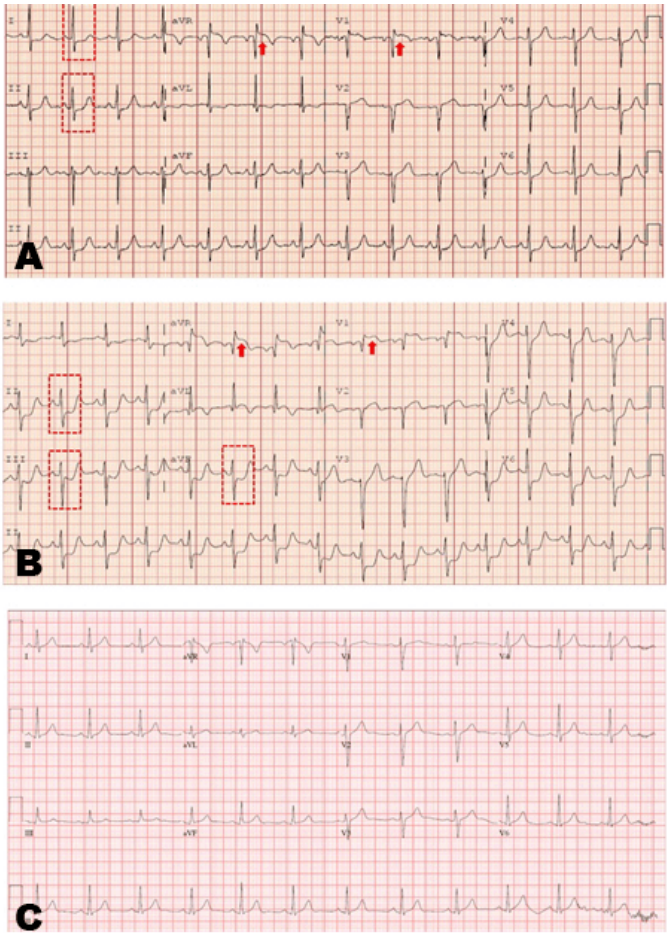


Figure 1: (A) Initial electrocardiogram (ECG) showing 0.1 mV ST-elevations in aVR and V1 (red arrows) and ST depression in lead I and II (red boxes), (B) Repeat ECG four hours after presentation showing 0.3 mV ST-elevation in aVR and V1 (red arrows) with reciprocal changes in lead II, III and aVF (red boxes), and (C) Normal ECG.

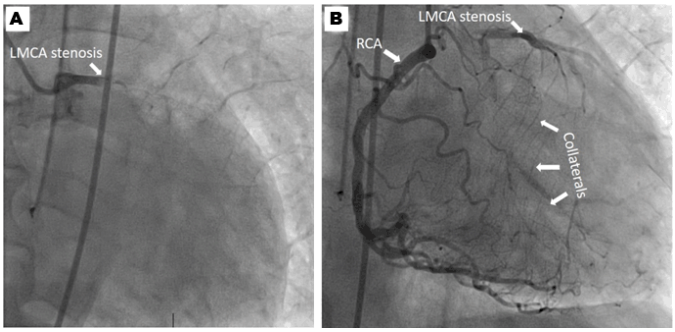


Figure 2: Coronary angiogram. Panel A shows a patent and dominant right coronary artery with extensive collateral system to the left ventricle. Panel B demonstrates a totally occluded left main coronary artery.

arterial pressure and improved collateral perfusion. Given recent administration of t-PA, intravenous heparin was not recommended due to increased risk of bleeding. Post-cardiac catheterization and after clearance by neurosurgery and neurology, the patient was started on aspirin and metoprolol succinate. He was managed in the intensive care unit for 48 hours, extubated and transferred to stroke unit.

Echocardiogram was performed showing a left ventricular ejection fraction of 40% correlating with moderate systolic dysfunction, and mild concentric left ventricular hypertrophy. Also, the entire anterior, basal and mid anterolateral wall, mid anteroseptal segment, apical lateral segment, and apex were hypokinetic. Grade II diastolic dysfunction was also seen. During his hospital study, the patient was found to have multiple episodes of atrial fibrillation that was rate controlled with digoxin and nicardipine hydrochloride with appropriate response. The patient was eventually planned for staged coronary bypass surgery. However, one week after presentation developed septic shock secondary to aspiration pneumonia and ultimately expired.

DISCUSSION

Interpretation of the ECG in the appropriate clinical setting remains the cornerstone in diagnosis of STEMI [3–5] and helps in localizing stenotic lesions [6]. Nonetheless, lead aVR has been historically ignored in clinical practice partially because of its relative isolation (aVR has no other adjacent lead) [1, 2, 7]. Analysis of this particular lead is of great importance mainly due to its anatomical position.

Yamaji et al. [8] were able to illustrate that ST-elevation in aVR especially when greater than that in V1, predicts with high sensitivity and specificity LMCA occlusion. Acute occlusion of the LMCA is generally associated with high mortality. Without collateral circulation, patients are at risk of cardiogenic shock. ST-segment elevation in lead aVR was also strongly associated with increased rates of in-hospital recurrent ischemic events and heart failure.

Wong et al. [2] demonstrated a higher mortality in patients with aVR ST-elevation compared to those with ST elevation in any other leads. This might be partially explained by its association with severe LMCA disease [9]. Our patient initially presented with a 0.1 mV ST-elevation in aVR that progressed into frank STEMI. We hypothesize that hypotension-related hypoperfusion of RCA collateral circulation, in the setting of a chronically occluded LMCA was the cause of STEMI. Similar cases have been described in literature associating aVR ST-elevations with significant left main coronary artery or ostial LAD stenosis [10, 11]. The role of collateral circulation in coronary artery disease is a growing and promising field [12] and our case is an example of the enormous therapeutic potential of inducing arteriogenesis and angiogenesis to establish collateral circulations able supply ischemic areas of the myocardium.

Prompt recognition of isolated aVR ST changes due to critical LMCA and LAD occlusions would allow for identification of high risk patients and appropriate activation of the catheterization laboratory [13]. An early invasive approach might be especially beneficial in patients presenting with this finding [14].

CONCLUSION

This case of isolated ST segment elevation in augmented vector right (aVR) secondary to an occluded or under perfused (STOUP) collateral circulation demonstrates the importance of early recognition of changes in aVR, as well as the therapeutic potential of an efficient collateral circulation.

****

Author Contributions

Edward Rojas – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Molly Malone – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
George Stoupakis – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

REFERENCES

1. Gorgels AP, Engelen DJ, Wellens HJ. Lead aVR, a mostly ignored but very valuable lead in clinical electrocardiography. J Am Coll Cardiol 2001 Nov 1;38(5):1355–6.
2. Wong CK, Gao W, Stewart RA, et al. aVR ST elevation: An important but neglected sign in ST elevation

acute myocardial infarction. Eur Heart J 2010 Aug;31(15):1845–53.
3. Barold SS. Willem Einthoven and the birth of clinical electrocardiography a hundred years ago. Card Electrophysiol Rev 2003 Jan;7(1):99–104.
4. Fisch C. Centennial of the string galvanometer and the electrocardiogram. J Am Coll Cardiol 2000 Nov 15;36(6):1737–45.
5. Thygesen K, Alpert JS, Jaffe AS, et al. Third universal definition of myocardial infarction. Circulation 2012 Oct 16;126(16):2020–35.
6. Engelen DJ, Gorgels AP, Cheriex EC, et al. Value of the electrocardiogram in localizing the occlusion site in the left anterior descending coronary artery in acute anterior myocardial infarction. J Am Coll Cardiol 1999 Aug;34(2):389–95.
7. George A, Arumugham PS, Figueredo VM. aVR – the forgotten lead. Exp Clin Cardiol 2010;15(2):e36–44.
8. Yamaji H, Iwasaki K, Kusachi S, et al. Prediction of acute left main coronary artery obstruction by 12-lead electrocardiography: ST segment elevation in lead aVR with less ST segment elevation in lead V(1). J Am Coll Cardiol 2001 Nov 1;38(5):1348–54.
9. de Winter RJ, Verouden NJ, Wellens HJ, et al. A new ECG sign of proximal LAD occlusion. N Engl J Med 2008 Nov 6;359(19):2071–3.
10. Hirapur I, Mantgol RV, Agrawal N. Isolated ST elevation in lead aVR during TMT at high workload as a marker of ostioproximal LAD occlusion. BMJ Case Rep 2014 Mar 18;2014.
11. Kossaify A. ST Segment Elevation in aVR: Clinical Significance in Acute Coronary Syndrome. Clin Med Insights Case Rep 2013 Mar 21;6:41–5.
12. Freedman SB, Isner JM. Therapeutic angiogenesis for coronary artery disease. Ann Intern Med 2002 Jan 1;136(1):54–71.
13. Rokos IC, French WJ, Mattu A, et al. Appropriate cardiac cath lab activation: optimizing electrocardiogram interpretation and clinical decision-making for acute ST-elevation myocardial infarction. Am Heart J 2010 Dec;160(6):995–1003, 1003.e1–8.
14. Barrabés JA, Figueras J, Moure C, Cortadellas J, Soler-Soler J. Prognostic value of lead aVR in patients with a first non-ST-segment elevation acute myocardial infarction. Circulation 2003 Aug 19;108(7):814–9.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Hyperleucocytic chronic myeloid leukemia with facial nerve palsy at presentation

Adama Isah Ladu, Aisha Mohammed Abba

ABSTRACT

Introduction: Chronic myeloid leukemia (CML) is a clonal malignancy arising from the hemopoeitic stem cell (HSC). The disease can be complicated by hyperleucocytosis which is a medical emergency associated with high mortality of about 20–40%, and can results in various complication including tumour lysis syndrome, disseminated intravascular coagulation and leucostasis. The common sites for leucostasis are the lungs and central nervous system. Case Report: We report a 15-year-old girl presented with one month history of progressive headache, fever, night sweat and one week history of right facial deviation and drooping of the left eyelid. Examination revealed hepatosplenomegaly and right infra nuclear seventh palsy. Complete blood count revealed hyperleucocytosis, with total white cell count (WCC) of > 275 109/l. Peripheral blood film and bone marrow cytology were in keeping with CML in chronic phase. Real time polymerase chain reaction analysis of whole blood was positive for Philadelphia chromosome. Cytoreductive treatment with hydroxyurea was started immediately following initial supportive treatment to prevent tumor lysis syndrome. Progressive improvement in both clinical and laboratory parameters was achieved, however, the patient still had residual facial palsy at third month follow-up. Conclusion: This is a rare case of hyperleucocytic chronic myeloid leukemia complicated by infra nuclear seventh palsy at the initial presentation. There was partial response to cytoreduction using hydroxyurea.

Keywords: Chronic myeloid leukemia, Hyperleukocytosis, Facial nerve palsy

How to cite this article

Ladu AI, Abba AM. Hyperleucocytic chronic myeloid leukemia with facial nerve palsy at presentation. Int J Case Rep Images 2017;8(8):523–527.

Article ID: Z01201708CR10814AL

****

doi:10.5348/ijcri-201775-CR-10814

Adama Isah Ladu[1], Aisha Mohammed Abba[2]

Affiliations: [1]MBBS, MSc, FMCPath, Lecturer 1 and Consultant Hematologist, Department of Hematology and Blood Transfusion, University of Maiduguri Teaching Hospital, Borno State, Nigeria; [2]MBBS, FMCPath: Lecturer 1 and Consultant Hematologist, Department of Hematology and Blood Transfusion, University of Maiduguri Teaching Hospital, Borno State, Nigeria.

Corresponding Author: Dr. Adama Isah Ladu, Department of Hematology and Blood Transfusion, University of Maiduguri Teaching Hospital, Borno State, Nigeria; Email: adamaisahladu@gmail.com

Received: 04 February 2017
Accepted: 22 May 2017
Published: 01 August 2017

INTRODUCTION

Chronic myeloid leukemia (CML) is a clonal malignancy arising from the hematopoietic stem cell. The hallmark of the disease is characterized by the presence of Philadelphia chromosome. This arises from the reciprocal translocation between the Abelson gene (ABL) on the long-arm of chromosome 9 and the breakpoint cluster region (BCR) on the long-arm of 22 [t(9,22) [1].

Hyperleukocytosis is defined as a white blood cell level of >100x10^9/L, and can complicate any form of

leukemia. This is a medical emergency associated with high mortality of about 20–40%, and can results in various complication including tumor lysis syndrome, disseminated intravascular coagulation and leukostasis [2, 3]. The common sites for leukostasis are the lungs and central nervous system (CNS). There are previous reports of adolescent patient with CML presenting with severe sensory impairment as a result of hyperleukocytosis complicated by CNS leukostasis [4]. Patients with overt CNS leukemia may manifest with cranial nerve palsy, commonly with unilateral seventh nerve palsy and less often the third and sixth cranial nerve [5]. In this report, we present a case of CML in an adolescent girl with hyperleukocytosis and seventh cranial nerve palsy at diagnosis.

CASE REPORT

A previously healthy 15-year-old girl was admitted with one-month history of moderate to severe headache, weight loss, fever; one week prior to presentation she developed abnormal sensation in both ears, deviation of angle of the mouth to the left, with inability to close the right eye, and partial drooling of the left eye. Five days before admission, she developed a progressive scalp swelling which was initially the size of a pea, and said to have ruptured and bled spontaneously into the surrounding area, there was no other bleeding episodes. Her past medical history was unremarkable. General examination revealed an irritable and lethargic girl, sweaty, febrile (37.8°C), mildly pale, with a respiratory rate of 26 cycles per minute and pulse rate of 110 beats per minute, there was no peripheral lymphadenopathy. Local examination revealed no purpura or ecchymosis, however, there was a discreet, firm, moderately tender hemorrhagic mass on the scalp, measuring 10x10 cm (Figure 1). Neurological examination revealed no evidence of meningism; there was a right infra nuclear facial nerve palsy. There was partial ptosis of the left eye with otherwise normal ocular movement in all directions (Figure 2), both pupils were reactive to light and accommodation. No other focal neurology was evident. The spleen was palpable 16 cm and the liver 6 cm below their respective costal margins.

Complete blood count revealed hematocrit of 28%, total white cell count of $275x10^9$/L and platelets count of $980x10^9$/L. Peripheral blood film (PBF) revealed all spectrum of white blood cells (Figure 3) –myeloblast 3%, promyelocytes 6%, myelocytes 40%, metamyelocytes 12%, band form 19% and neutrophil 20%. Bone marrow aspiration (BMA) cytology showed markedly increased myelopoiesis, the myelogram showed evidence of sequential maturation along the cell line with a peak at the myelocyte stage and myeloblast count of less than 5%. The PBF and BMA were in keeping with CML in chronic phase; rtPCR analysis of whole blood was positive for Philadelphia chromosome and the b3a2 transcript type (classical CML) was detected; quantification revealed BCR-ABL quantity of 4,578 copies per 2.5 micro liter of cDNA, ABL copies of 931,750 per 2.5 µl of cDNA and BCR-ABL/ABL ratio of 0.49%. The serum biochemistry was unremarkable. A skull X-ray revealed no bony involvement by the scalp swelling (Figure 4). Cerebrospinal fluid analysis was negative for malignant cells. Chest X-ray revealed cardiomegaly with right upper lobe opacity. Two-dimensional echocardiogram was performed and showed severe left ventricular dysfunction and dilated left ventricular wall, with increased pulmonary vascular resistance.

An assessment of chronic myeloid leukemia presenting with hyperleukocytosis complicated by CNS leukostasis was made. Supportive treatment was started immediately with hydration using normal saline (150 ml/kg), allopurinol 100 mg daily and low dose aspirin (in view of the markedly raised platelets count) for 24 hr, before commencement of cytoreduction with hydroxyurea (40 mg/kg/day QDS). This was to expand the plasma volume and reduce the attendant risk of tumor lysis syndrome that may accompany cytoreduction. The patient showed progressive improvement in both clinical and laboratory parameters 72 hours after admission (Table 1). Her dose of hydroxyurea was titrated based on her total white blood cell count, and by two weeks following cytoreduction with hydroxyurea her WBC was stable at $<10x10^9$/L (Table 1). The scalp mass was managed conservatively with daily saline dressing, and resolved completely by day-10 of admission. The patient was referred to another tertiary centre in South west Nigeria where imatinib treatment

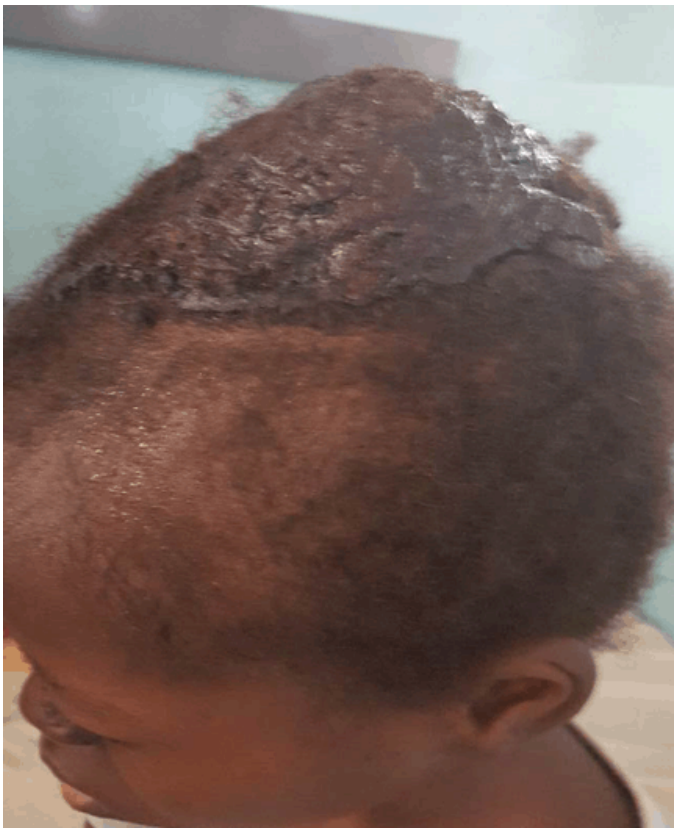


Figure 1: Scalp mass at presentation.

Figure 2: Right infra nuclear facial palsy with loss of nasolabial fold and inability to close the right eye (not shown in picture). Partial left eye drooling present.

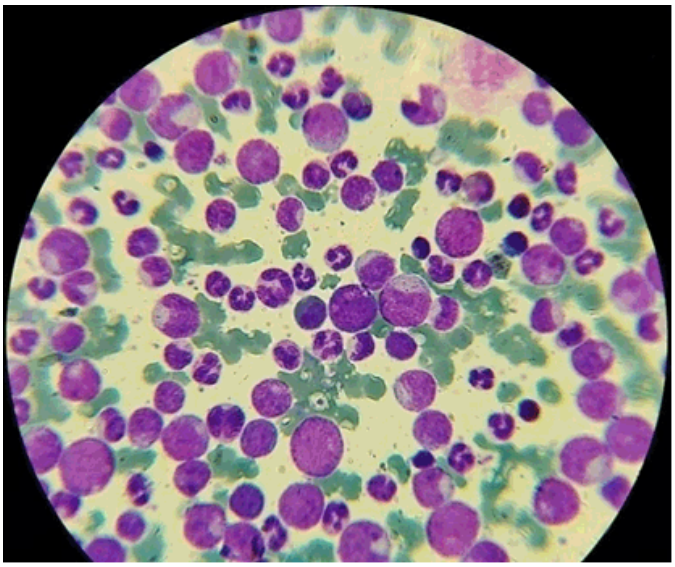


Figure 3: Peripheral blood showing all spectrum of myeloid cells present.

Table 1: Clinical and laboratory parameters following supportive treatment and hydroxyurea

Days	Total WBC	Platelets	Spleen size	Scalp mass
Day-1	>275x10^9/ml	>900x10^9/ml	18 cm	10x9 cm
Day-3	215x10^9/ml	750x10^9/ml	14 cm	8x9 cm
Day-5	120x10^9/ml	305x10^9/ml	12 cm	5x4 cm
Day-14	8x10^9/ml	280x10^9/ml	6 cm	Resolved

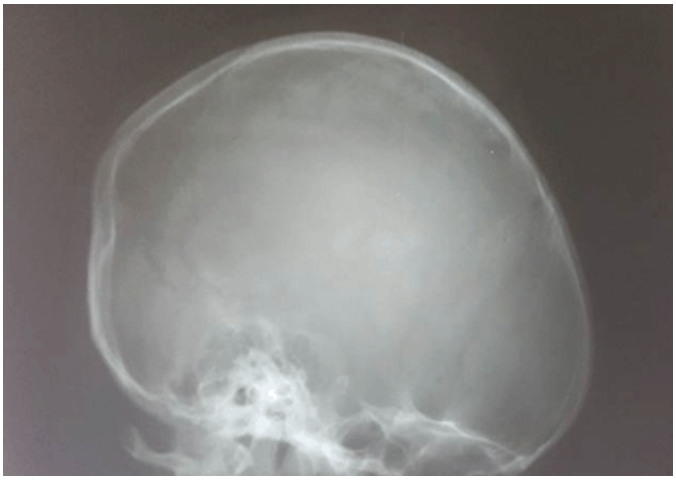


Figure 4: Normal skull X-ray which indicates no obvious bony involvement from the scalp mass.

made available by an international donor agency is given free of charge, however, her parents declined to proceed with any further treatment despite adequate counseling. She was continued on hydroxyurea and regular physiotherapy for her cranial palsy. She remained stable during regular follow-up, however, there was still residual seventh nerve palsy at third month follow-up.

DISCUSSION

Neurological complications in patients with leukemia may result acutely either from the disease at the time of diagnosis or from relapse of the disease or as complication of treatment procedure [6, 7]. Advance CNS involvement presents with features such as irritability, headache, seizures and coma. Overt CNS involvement is defined as the presence of white blood cell of five microliters or more in the CSF with evidence of blast cells and or presence of cranial nerve palsy or cerebral mass. This is seen in about 3% of children presenting with leukemia at the time of diagnosis [6]. The poor CNS perfusion following leukostasis or thrombosis in cases presenting with hyperleukocytosis may manifests with altered mental status. The high leukocrit associated with the raised total white blood cell count results in increased blood viscosity, consequently, the large non deformable blast cells results in reduce blood flow in marginally sized vessels and occlusion of microvasculature in the lungs and CNS [6]. The mainstay of management of symptomatic patients presenting with hyperleukocytosis consist of supportive treatment with aggressive hydration, allopurinol to prevent tumor lysis syndrome, urine alkalinization and appropriate respiratory support, followed by cytoreduction treatment using leukapheresis or chemotherapeutic agents like hydroxyurea [1, 2].

In the index case, the patient presented with hyperleukocytosis, which was complicated by facial palsy. The facial nerve is the commonest involve in cranial

nerve neuropathy [8, 9]. There are few reports of children with AML presenting with facial nerve palsy at diagnosis [9, 10]. However, it is rare manifestation in children with CML at the initial presentation. The time from the appearance of facial nerve palsy to diagnosis of leukemia varies from I day to I month; and usually improves one to six month following chemotherapy [10]. Allogenic stem cell transplantation with whole brain irradiation has been shown to be effective in facial nerve palsy in children with AML [10]. However, there is no definite therapeutic management in patients with CML, timely and prompt intervention in patients presenting with hyperleukocytosis can reverse some of the complications [6]. Of note in the index case is the cutaneous hemorrhage which may be related to the markedly raised platelets count, and the partial left eye drooling with an otherwise normal third nerve function; in addition, the patient had evidence of alveolar infiltrates on chest X-ray and other symptoms suggestive of hyperviscosity, which improved dramatically within few days of supportive treatment and hydroxyurea.

CONCLUSION

In conclusion, presence of cranial nerve neuropathy at the initial presentation of patients with hyperleukocytosis indicates central nervous system involvement and requires prompt intervention to help reverse some of the features. Timely and aggressive intervention is particularly critical in poor resource setups like ours, where facility and expertise in leukapheresis is not readily available and therefore supportive management must be optimized.

****

Author Contributions

Adama Isah Ladu – Substantial contributions to concept and design of report, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Aisha Mohammed Abba – Substantial contributions to concept and design of report, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Jain R, Bansal D, Marwaha RK. Hyperleukocytosis: Emergency management. Indian J Pediatr 2013 Feb;80(2):144–8.
2. Ganzel C, Becker J, Mintz PD, Lazarus HM, Rowe JM. Hyperleukocytosis, leukostasis and leukapheresis: Practice management. Blood Rev 2012 May;26(3):117–22.
3. Gokce M, Unal S, Bayrakçi B, Tuncer M. Chronic myeloid leukemia presenting with visual and auditory impairment in an adolescent: An insight to management strategies. Indian J Hematol Blood Transfus 2010 Sep;26(3):96–8.
4. Laningham FH, Kun LE, Reddick WE, Ogg RJ, Morris EB, Pui CH. Childhood central nervous system leukemia: Historical perspectives, current therapy, and acute neurological sequelae. Neuroradiology 2007 Nov;49(11):873–88.
5. Pfeifer H, Wassmann B, Hofmann WK, et al. Risk and prognosis of central nervous system leukemia in patients with Philadelphia chromosome-positive acute leukemias treated with imatinib mesylate. Clin Cancer Res 2003 Oct 15;9(13):4674–81.
6. Piro E, Carillo G, Levato L, Kropp M, Molica S. Reversal of leucostasis-related pulmonary distress syndrome after leukopharesis and low-dose chemotherapy in acute myleloid leukaemia. Journal of clinical oncology 2011;29(26):e725–e6.
7. Kun LE. Leukaemias in children. Pediatric Radiology Oncology 2007:15–39.
8. Ino-Ekanem M, Ekwere TA. Generalized chloromas with multiple cranial nerves palsies in a patient with chronic myeloid leukemia in a tertiary institution in South-south Nigeria: A case report. Am J Hematol 2003 Nov;74(3):200–1.
9. Sood BR, Sharma B, Kumar S, Gupta D, Sharma A. Facial palsy as first presentation of acute myeloid leukemia. Am J Hematol 2003 Nov;74(3):200–1.
10. Baek HJ, Han DK, Kim YO, Choi IS, Hwang TJ. Facial palsy as the presenting symptom of acute myeloid leukaemia in children: Three cases with stem cell transplantation. Korean J Pediatr 2009;52(6):713–9.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Cardiac tamponade as a life-threatening complication after laparoscopic mesh hiatal hernia repair

Dimitar Penchev, Georgi Kotashev, Georgi Popivanov, Ventsislav Mutafchiyski

ABSTRACT

Introduction: Anti-reflux surgery becomes one of the most common laparoscopic procedures during the last decade, with perioperative morbidity rates up to 19% and mortality around 0.1%. The increasing usage of prosthetic mesh in large hiatal hernias led to a new type of complications such as mesh migration or infection, rejection and life-threatening cardiac tamponade. As of today, only few cases with cardiac tamponade after laparoscopic hiatal hernia repair are reported in the English literature. Here we present a case with cardiac tamponade after laparoscopic mesh-repair in order to increase the awareness of this rare complication. Case Report: We present a 72-year-old female with a large symptomatic type II hiatal hernia and grade II reflux disease by Savary-Miler classification. Nissen's fundoplication was performed along with relaxing incision of the left crus. Omiramesh® mesh was used and fixed with Secure strap® to the diaphragm. After six hours because of dyspnea, anxiety and retrosternal chest pain, desaturation, atrial fibrillation, significant pericardial effusion on echocardiography and hemodynamic instability she was consulted with thoracic surgeon and pericardial puncture with evacuation of 120 ml blood was performed. The patient was transferred immediately to the operative room for thoracotomy because of lack of effect, but she died before the thoracic procedure. Conclusion: The early diagnosis and proper management are crucial in case of cardiac tamponade. Understanding the mechanism of cardiac tamponade and the proper fixation of prosthetic material only to the diaphragmatic crus is the way to avoid this complication. Using glue may be a safe alternative to staples.

Keywords: Cardiac tamponade, Hiatal hernia, Laparoscopic fundoplication, Mesh repair

How to cite this article

Penchev D, Kotashev G, Popivanov G, Mutafchiyski V. Cardiac tamponade as a life-threatening complication after laparoscopic mesh hiatal hernia repair. Int J Case Rep Images 2017;8(8):528–532.

Article ID: Z01201708CR10815DP

****

doi:10.5348/ijcri-201776-CR-10815

Dimitar Penchev[1], Georgi Kotashev[1], Georgi Popivanov[1], Ventsislav Mutafchiyski[1]

Affiliation: [1]Clinic of Endoscopic, Endocrine surgery and Coloproctology, Military Medical Academy, Sofia, Bulgaria.

Corresponding Author: Georgi Popivanov, Clinic of Endoscopic, Endocrine surgery and Coloproctology, Military Medical Academy, Sofia, Bulgaria, Georgi Sofiiski Str. 3; Email: gerasimpopivanov@rocketmail.com

Received: 09 February 2017
Accepted: 27 May 2017
Published: 01 August 2017

INTRODUCTION

Surgical treatment of gastroesophageal reflux disease (GERD) is an alternative second line therapy after failed conservative treatment. Anti-reflux surgery becomes one of the most common laparoscopic procedures during the last decade, with perioperative morbidity rates up to 19% and mortality around 0.1% [1]. Most of the complications after anti-reflux surgery are minor, without any need of surgical intervention. Nevertheless, serious complications

may occur such as spontaneous pneumothorax, spleen injury, severe bleeding and esophageal perforation or as wittily wrote Prof. Swanström "the only way to die from GERD is to be operated on for it". The increasing usage of prosthetic mesh-reinforcement for prevention of recurrence leads to a new type of complications such as mesh migration, rejection reaction, mesh infection and cardiac tamponade [2]. The cardiac tamponade is a rare but life-threatening complication associated with significant mortality. As of today, there are only few cases reported in the English literature [3]. Here we present a case report of patient with cardiac tamponade after mesh-repair of large hiatal hernia in order to increase the awareness of this rare complication.

CASE REPORT

A 72-year-old female was admitted in our institution for surgical repair of GERD with a large symptomatic hiatal hernia. Conservative treatment, for more the one year, did not show any significant improvement of the regurgitation, dysphagia, chronic cough and oral intake limitations. She had a past history of arterial hypertension, thrombocytosis, chronic heart failure and osteoporosis. Endoscopic and contrast X-ray examinations showed multiple duodenal diverticula and a large, type II hiatal hernia containing 1/3 of the stomach into mediastinum, with diameter of 10 cm and reflux disease Grade II by Savary-Miler classification. Laparoscopy confirmed large hiatal hernia with diastasis and sclerosis of the diaphragmatic crus (Figure 1).

Nissen's fundoplication was performed along with relaxing incision of the left crus. Omiramesh® (Ethicon) 10x15 cm mesh was placed with Secure strap® (Ethicon, Norderstedt, Germany) to the diaphragm (Figure 2 and Figure 3).

After surgery patient was transferred to intensive care unit for postoperative monitoring. Several hours after procedure patient complained of mild dyspnea, anxiety and retrosternal chest pain. Desaturation, atrial fibrillation on ECG and pleural effusion on X-ray were noted and acute heart failure, pulmonary embolism and myocardial infarction were suspected. Due to normal troponin levels, significant pericardial effusion on echocardiography and hemodynamic instability the patient was consulted with thoracic surgeon and pericardial puncture with evacuation of 120 ml blood was performed. The patient was transferred immediately to the operative room for thoracotomy and pericardial fenestration because of lack of effect. Unfortunately, she died before the thoracic procedure.

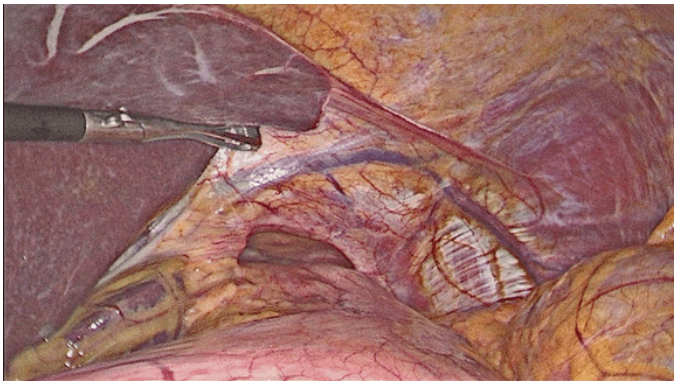


Figure 1: Intraoperative view.

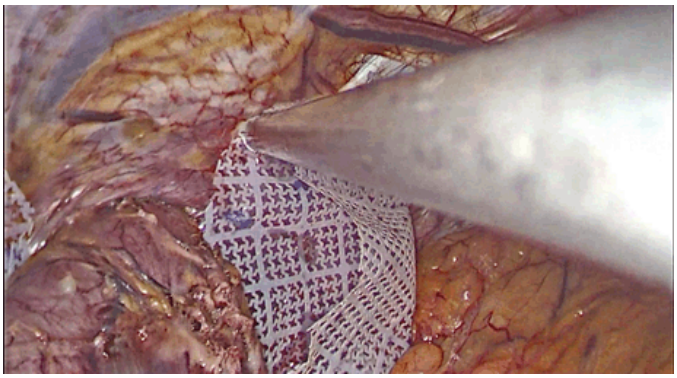


Figure 2: Instrument against tissue for fixation of the mesh – thin down the tissue and increase the potential for underlying damage.

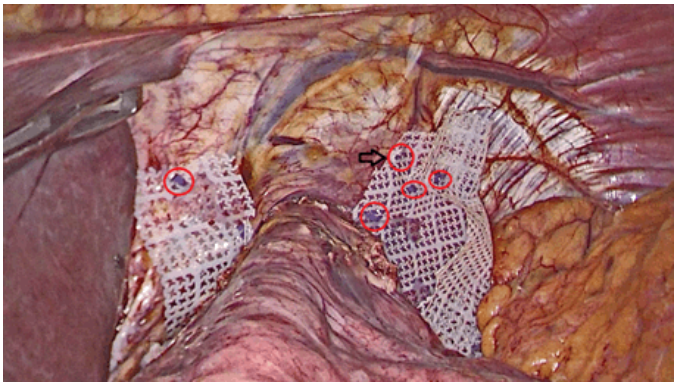


Figure 3: Mesh fixed to the diaphragm with red circles. Black arrow shows the most dangerous tack at the diaphragmatic tendon near to the heart.

DISCUSSION

Treatment of large hiatal hernia is required, because of the potential for significant complications such as volvulus, anemia, obstruction, strangulation or perforation. Cardiac tamponade is a rare, but life-threatening complication after anti-reflux surgery. The awareness about possible complications, which can occur in postoperative period, is crucial for early diagnosis and successful management. McClellan et al. reported a case with cardiac injury managed successfully [1].

Most of the studies analyzing the possible complications are retrospective reporting morbidity rate up to 19%. Most complications occur are minor; grade 1–2 according to Clavien–Dindo classification. Only

few retrospective trials took into account the severe life-threatening complications, such as injuries to the esophagus, stomach, spleen, pleura, aorta and heart [3]. Lundell published a review with mortality rate 0.15%, which was related to pneumothorax, hemorrhage, perforations, and splenectomy [4]. The most common cause of lethal outcome was pneumothorax. Mesh-related complications are a quite new area in hiatal hernia management. Almost 10% of surgeons use mesh in hiatal hernia and 45% of them are for large ones. Several reports reported the possible aortic and cardiac injury during procedures, related to laparoscopic tacks, sutures or dissection around the hiatus [1, 3, 5–8].

The understanding of mechanism of cardiac tamponade after hiatal hernia repair with mesh has a significant practical importance. The thickness of the diaphragm ranges from 1.5–5.4 mm. The central tendon averages from 2.9–3 mm [3]. Secure strap® tacks are 7.2 mm in length and even angled tacks have more than 4 mm length. The pressure on the instrument against the tissue thin down the tissues thus additionally contributing to this complication.

Perforated ventricular marginal branch vessel and tearing myocardium are possible sources of bleeding. In most of the reported cases spiral tacks were used, but there are reports after mesh fixation with angle tacks and sutures. The review of Frantzides et al. reported 15 cases —two with sutures, eleven with spiral tackers and one with angle tacker— with 66% mortality [3].

Other possible mechanism is by compressing hematoma around the heart [5]. The main cause of hematoma is extensive dissection and postoperative mediastinal bleeding. Usual symptoms of cardiac tamponade are hypotension, tachycardia, desaturation, anxiety, dyspnea, tachycardia, atrial fibrillation, distended neck veins and muffled heart sounds. They can be easily confused by other common postoperative complications, such as bleeding, pulmonary embolism, acute heart failure, pneumothorax, atelectasis and myocardial infarction. Some authors reported a case with cardiac tamponade which was primarily wrongly diagnosed with inferior myocardial infarction, because of ECG findings and elevated troponin levels [6]. Clinical manifestation may occur at any time in the postoperative course. Sugumar et al. reported two cases with cardiac tamponade on fifth week and on 37th day after surgery, respectively [7]. Usually, symptoms occur from few hours after operation up to 14 days. The most accurate method for diagnosis is transesophageal echocardiography, but in most cases, especially in non-cardiac surgery centers, standard postoperative monitoring includes ECG, echocardiography and serum troponin levels. The early and accurate diagnosis needs to be followed by a proper management by an experienced cardio-thoracic surgeon. Generally, there are three options to manage cardiac tamponade – the first one is conservatively to put a suction drain in pericardium to evacuate the blood. McClellan et al. reported a case with successful management of patient, using 6 French suction drain in pericardium with 370 ml blood evacuation for one day and resolving the symptoms immediately after drainage [1]. In our case, we tried the similar approach without success, because the symptoms still remained and the patient became unstable. The second treatment option is laparotomy with creating of subxiphoid window and drainage of pericardium into abdominal cavity with or without drain placement such in the case of Stich et al. [7]. The third and most aggressive approach is thoracotomy with pericardial fenestration and drainage. Surgical approaches are more preferable, because they allow making a definitive hemostasis if necessary. Recently, several surgeons use glue for mesh fixation to evade the potential risk of injuries [9]. The fear of complications or negative experience does not preclude the mesh reinforcement in large hiatal hernia repair [10].

CONCLUSION

Severe life-threatening complications are underestimated and need more discussion and recommendation for risk assessment, diagnosis and management. Early diagnosis and proper management are crucial in case of cardiac tamponade. The understanding of mechanism of cardiac tamponade and proper fixation of prosthetic material only to the diaphragmatic crus are of crucial importance to evade this type of complication. Using glue is a safe alternative to staples.

****

Author Contributions

Dimitar Penchev – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Georgi Kotashev – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Georgi Popivanov – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Ventsislav Mutafchiyski – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

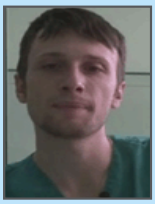
Authors declare no conflict of interest.

REFERENCES

1. McClellan JM, Nelson D, Martin M. Hemopericardium after laparoscopic perihiatal procedures: High index of suspicion facilitates early diagnosis and successful nonoperative management. Surg Obes Relat Dis 2016 Mar–Apr;12(3):e27–31.
2. Stadlhuber RJ, Sherif AE, Mittal SK, et al. Mesh complications after prosthetic reinforcement of hiatal closure: A 28-case series. Surg Endosc 2009 Jun;23(6):1219–26.
3. Frantzides CT, Welle SN. Cardiac tamponade as a life-threatening complication in hernia repair. Surgery 2012 Jul;152(1):133–5.
4. Lundell L. Complications after anti-reflux surgery. Best Pract Res Clin Gastroenterol 2004 Oct;18(5):935–45.
5. Fernandez Mdel C, Diaz M, López F, Martí-Obiol R, Ortega J. Cardiac complications after laparoscopic large hiatal hernia repair. Is it related with staple fixation of the mesh? –Report of three cases. Ann Med Surg (Lond) 2015 Oct 14;4(4):395–8.
6. Müller-Stich BP, Linke G, Leemann B, Lange J, Zerz A. Cardiac tamponade as a life-threatening complication in antireflux surgery. Am J Surg 2006 Jan;191(1):139–41.
7. Sugumar H, Kearney LG, Srivastava PM. Pericardial tamponade: A life threatening complication of laparoscopic gastro-oesophageal surgery. Heart Lung Circ 2012 Apr;21(4):237–9.
8. Puchakayala MR, Abbey K, Haft J, Orringer MB. Delayed pericardial tamponade following transthoracic hiatal hernia repair. J Cardiothorac Vasc Anesth 2006 Apr;20(2):245–6.
9. Losi P, Burchielli S, Spiller D, et al. Cyanoacrylate surgical glue as an alternative to suture threads for mesh fixation in hernia repair. J Surg Res 2010 Oct;163(2):e53–8.
10. Müller-Stich BP, Kenngott HG, Gondan M, et al. Use of mesh in laparoscopic paraesophageal hernia repair: A meta-analysis and risk-benefit analysis. PLoS One 2015 Oct 15;10(10):e0139547.

ABOUT THE AUTHORS

Article citation: Penchev D, Kotashev G, Popivanov G, Mutafchiyski V. Cardiac tamponade as a life-threatening complication after laparoscopic mesh hiatal hernia repair. Int J Case Rep Images 2017;8(8):528–532.



Dimitar Penchev is a Resident at Clinic of Endoscopic, Endocrine Surgery and Coloproctology, Military Medical Academy, Sofia, Bulgaria. His research interests include laparoscopic surgery of upper gastrointestinal tract and colorectal cancer.
Email: d.k.penchev@gmail.com

Georgi Kotashev is a Consultant and Senior Surgeon at Clinic of Endoscopic, Endocrine Surgery and Coloproctology, Military Medical Academy, Sofia, Bulgaria. His research interests include laparoscopic surgery of ventral and hiatal hernia, stomach and colon/rectum.
Email: kotashev@gmail.bg

Georgi Popivanov is an Assistant Professor at Clinic of Endoscopic, Endocrine Surgery and Coloproctology, Military Medical Academy, Sofia, Bulgaria. His research interests include military surgery, trauma and hepatobiliary surgery, coloproctology and oncology.
Email: gerasimpopivanov@rocketmail.com

Ventsislav Mutafchiyski, Dsci, FACS is a Chief of Military Medical Academy, Sofia, Bulgaria. His research interests include military surgery, trauma, hepatobiliary and pancreatic surgery, coloproctology, liver transplantation, open abdomen. He has published more than 300 research papers in national and international academic journals and authored 30 chapter of books and monographies.
Email: ventzimm@gmail.com

CASE REPORT PEER REVIEWED | OPEN ACCESS

Primary thyroid diffuse large B cell lymphoma: A case report

Tej Prakash Soni, Upendra Nandwana, Lalit Mohan Sharma, Anil Kumar Gupta, Naresh Jakhotia, Shantanu Sharma

ABSTRACT

Introduction: Primary thyroid lymphoma is a unique and rare entity. We report a rare case of primary thyroid lymphoma (diffuse large B cell lymphoma) in a male patient. Case Report: A 40-year-old male presented with complaints of rapidly progressive mass in left thyroid region neck. True cut biopsy from the thyroid lesion and immunohistochemistry study was reported as primary thyroid lymphoma. He was treated with chemotherapy followed by radiotherapy. After one year of the treatment his disease is well controlled with no evidence of recurrence or residual tumor. Differential diagnosis includes Hashimoto's thyroiditis, anaplastic thyroid carcinoma, and secondary thyroid lymphoma. Hashimoto's thyroiditis is commonly associated with primary thyroid lymphoma. Conclusion: Accurate and correct diagnosis of primary thyroid lymphoma is very important as its management is different from other thyroid malignancies. It is treated by combination of chemotherapy and radiotherapy. The role of surgery is questionable. Palliative thyroidectomy can be done in patients with obstructive symptoms.

Keywords: Differential diagnosis, Primary Thyroid lymphoma, Rare

How to cite this article

Soni TP, Nandwana U, Sharma LM, Gupta AK, Jakhotia N, Sharma S. Primary thyroid diffuse large B cell lymphoma: A case report. Int J Case Rep Images 2017;8(8):533–537.

Article ID: Z01201708CR10816TS

****

doi:10.5348/ijcri-201777-CR-10816

Tej Prakash Soni[1], Upendra Nandwana[2], Lalit Mohan Sharma[3], Anil Kumar Gupta[4], Naresh Jakhotia[1], Shantanu Sharma[5]

Affiliations: [1]Consultant, Department of Radiation Oncology, Bhagwan Mahaveer Cancer Hospital and Research Centre, Jaipur, Rajasthan, India; [2]Senior Resident, Department of Radiation Oncology, Bhagwan Mahaveer Cancer Hospital and Research Centre, Jaipur, Rajasthan, India; [3]Senior Consultant, Department of Medical Oncology, Bhagwan Mahaveer Cancer Hospital and Research Centre, Jaipur, Rajasthan, India; [4]Senior Consultant, Department of Surgical Oncology, Bhagwan Mahaveer Cancer Hospital and Research Centre, Jaipur, Rajasthan, India; [5]Associate Professor, Department of Radiotherapy, S.M.S. Medical College and Hospital, Jaipur, Rajasthan, India.

Corresponding Author: Tej Prakash Soni, Consultant, Department of Radiation Oncology, Bhagwan Mahaveer Cancer Hospital and Research Centre, JLN Road, Jaipur, Rajasthan, India, Pincode 302017; Email: drtejprakashsoni@yahoo.co.in

Received: 20 April 2017
Accepted: 27 May 2017
Published: 01 August 2017

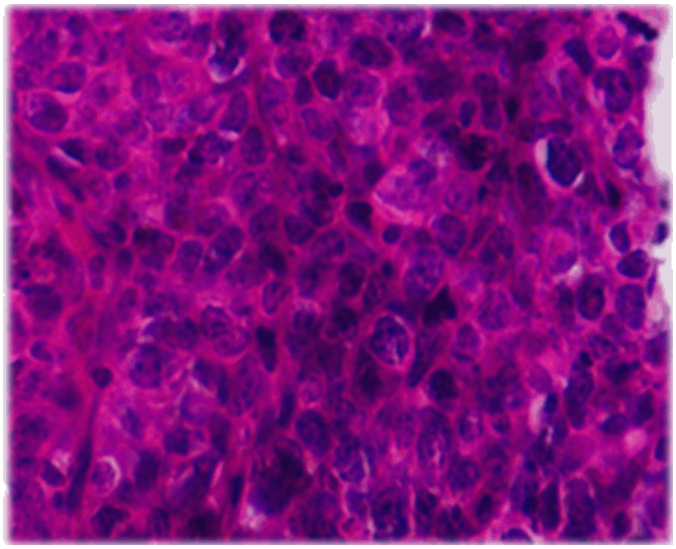
INTRODUCTION

Primary thyroid lymphoma is a rare entity. To diagnose it accurately and to differentiate with other thyroid malignancies is very important as primary thyroid lymphoma is highly curable without thyroidectomy. Most primary thyroid lymphomas are B cell non-Hodgkin's lymphoma and usually treated by combination of chemotherapy and radiotherapy.

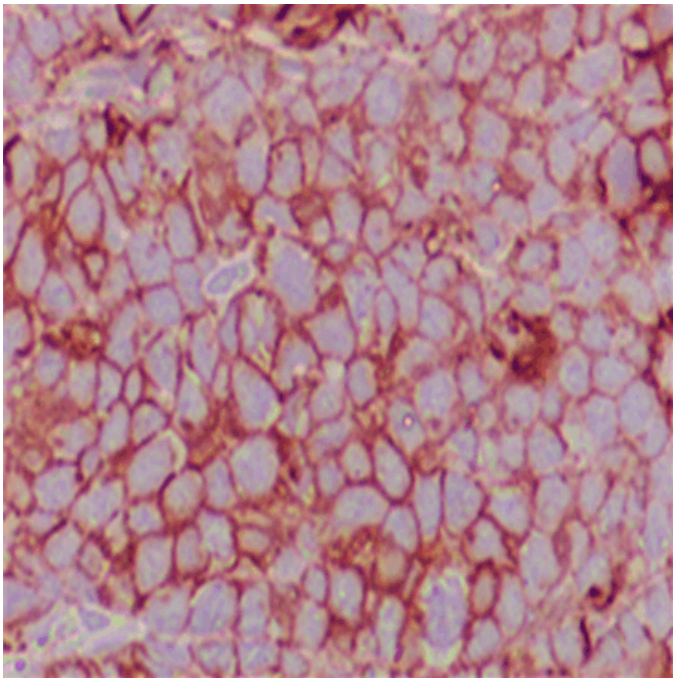
CASE REPORT

A 40-year-old male with history of hypothyroidism, taking tab levothyroxine 100 μg once daily since one year presented to our hospital with the presenting complaint of a swelling in left side of neck, with progressive increase in size since last two months. On examination, a diffuse non tender lump measuring 6×3 cm in root of left side neck, moving with deglutition was noted with no palpable lymphadenopathy elsewhere. Serum T3, T4 and TSH levels were within normal limits. All other routine blood investigations were normal. Ultrasonography (USG) of neck showed mixed echogenic mass lesion in left lobe thyroid measuring 56x29 mm. Ultrasonography guided fine-needle aspiration cytology from the left lobe of thyroid lesion showed lymphoid large mononuclear cells. Trucut biopsy from the left thyroid lobe lesion was suggestive of non-Hodgkin's lymphoma as lymphoid tissue with diffuse medium to large sized atypical cells with prominent round nucleoli and condensed chromatin with scanty cytoplasm and mitotic figures were present (Figure 1). Immunohistochemical analysis was reported as CD20 strongly positive, MIB-1 labeling index 60–65%, CD3 negative, CD5 negative, CD10 negative, CD23 negative, cycline D1 negative and Pan-Cytokeratin negative, suggestive of diffuse large B cell lymphoma (DLBCL) thyroid (Figure 2). The PET-CT scan showed FDG avid hypodense mass lesion involving the left lobe of thyroid gland measuring 63x38x31 mm with SUV Max 31.11 with no evidence of other abnormal lesion in the body (Figure 3). Bone marrow aspirate and biopsy were normal. Based on these investigations, final diagnosis was non-Hodgkin's lymphoma-Thyroid (DLBCL, stage IEA).

This case was discussed in multidisciplinary tumor board meeting. Chemotherapy followed by involved site radiotherapy (ISRT) as per our institute's protocol was planned for him. He received four cycles of chemotherapy (retuximab, cyclophosphamide, doxorubicine, prednisolone: R-CHOP) under medical oncology unit. He tolerated the chemotherapy well. After chemotherapy there was complete remission of thyroid lesion. Involved site radiotherapy (ISRT) was given to the thyroid region



Figures 1: Lymphoid tissue with diffuse medium to large sized atypical cells with prominent round nucleoli and condensed chromatin with scanty cytoplasm and mitotic figures (H&E stain, x400).



Figures 2: Immunophenotype features (x400) suggesting of large neoplastic lymphoid cells with strong and diffuse membranous staining of CD 20.

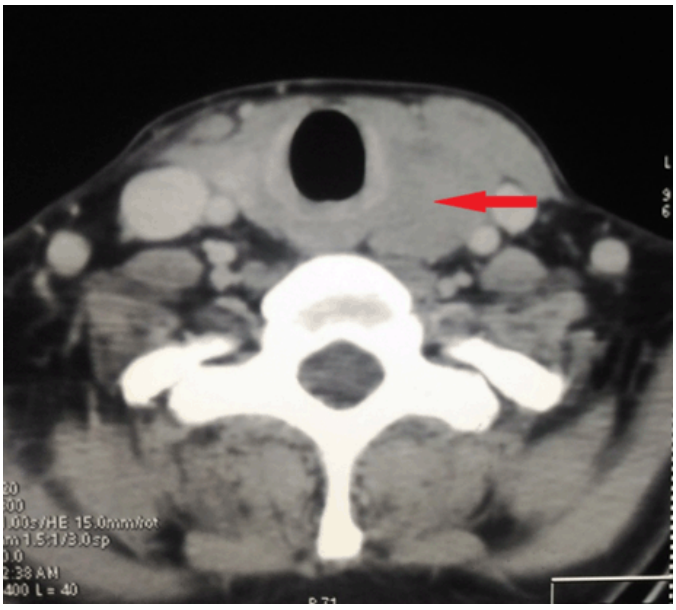


Figure 3: Axial view of contrast enhanced computed tomography scan of neck showing a large hypodense lesion (size max. 63 mm) in left lobe of thyroid.

to a dose 30 Gy in 15 fractions in three weeks. He is on regular follow-up. After one year of follow-up his disease is well controlled.

DISCUSSION

Primary thyroid lymphoma is a rare tumor and makes less than 5% of thyroid malignancies [1]. It constitutes no more than 2.5% of all lymphomas [1]. Most thyroid lymphomas are of B cell origin non-Hodgkin's lymphoma [2]. Diffuse large B cell lymphoma (DLBCL) constitutes 50–80% of the primary thyroid lymphoma and remaining 20-30% are extra nodal mucosa associated lymphoid tissue (MALT) lymphomas. Other rare subtypes include follicular lymphoma (12%), Hodgkin's disease (7%) [2]. Primary thyroid lymphoma is more common in 60–70 years old age group females (Female:male ratio 3:1) [1]. The most common presenting sign and symptom includes recently growing painless mass in neck thyroid region. It may be associated with fever, sweating and weight loss (Classic B-type symptoms) up to 20% of patients [3]. Dysphagia, stridor, hoarseness of voice may be seen in 30% cases due to compression by the mass over adjacent structures [3]. About 10–30% cases may also have hypothyroidism at the time of presentation [3].

Ultrasongraphy (USG) guided fine-needle aspiration cytology (FNAC) of the thyroid lesion is a simple, cost effective, accurate and rapid to perform investigation test for thyroid lesions. Fine niddle aspiration biopsy added with flow cytometry and immunohistochemical studies may be more sensitive and specific. Trucut needle biopsy is recommended to confirm the FNAC results or if FNAC results are insufficient or inclusive. Ultrasonography of the thyroid is very useful to characterize the thyroid lesion such as size and shape of the lesion, micro or coarse calcification, solid or cystic appearance, texture of the gland, blood flow within the lesion, proximity to blood vessels etc. Computed tomography scan neck is done to confirm the Ultrasonography findings, assessment of local invasion, lymph node metastasis, and evaluation of oro-naso-laryngopharynx and esophagus. The PET scan is useful for staging, detection of distant metastasis or second primary, radiotherapy treatment planning and post treatment response evaluation. Bone marrow biopsy is also required for staging. Differential diagnosis of primary thyroid lymphoma includes Hashimoto's thyroiditis (HT), secondary thyroid lymphoma, anaplastic thyroid carcinoma. Hashimoto's thyroiditis (autoimmune chronic lymphocytic thyroiditis) is the most prevalent autoimmune thyroid disorder [4]. Hashimoto's thyroiditis characteristic feature is lymphocytic infiltration of the thyroid gland followed by fibrous replacement of the thyroid parenchymal tissue. Hashimoto's thyroiditis is a well known risk factor and associated with 40-fold increased risk of developing primary thyroid lymphoma when compared to the general population [4]. Hashimoto's thyroiditis is associated with more than 90% of the primary thyroid lymphoma and due to this close association often both can be misdiagnosed for one another on FNAC [4].

Advanced and disseminated lymphoma can involve thyroid as secondary thyroid lymphoma. As with widespread disease burden, secondary lymphoma of the thyroid has poor treatment outcome compared to primary thyroid lymphoma [5]. It is very important to differentiate between anaplastic thyroid carcinoma and primary thyroid lymphoma. Anaplastic thyroid carcinoma is one of the most aggressive malignancies, with a poor prognosis and mean survival six months while primary thyroid lymphoma has excellent treatment outcomes. Primary thyroid lymphoma is treated mainly by chemotherapy with radiotherapy while surgical resection is needed for resectable anaplastic carcinoma thyroid. The presence of non-cohesive atypical large cells, irregular nuclear membrane, vesicular nuclei with prominent nucleoli and presence of lymphoglandular bodies in the background as cytology features, supported by CK and EMA negativity in IHC goes against the diagnosis of anaplastic carcinoma thyroid [6].

Primary thyroid MALT lymphoma has indolent course with superior clinical outcome (five-year survival more than 90%) compared to DLBCL of thyroid [7]. When MALT lymphoma (stage IE) is incidental diagnosis after thyroidectomy done for another reason there is no need of adjuvant chemotherapy or radiotherapy [7]. Thyroid lymphoma staging is done according to Ann-Arbor classification. Thyroidectomy is not recommended for the treatment of primary thyroid lymphoma [8]. Chemotherapy (CHOP: Cyclophosphamide, doxorubicin, vincristine, prednisone) followed by radiotherapy is the mainstay of the treatment for primary thyroid lymphoma [3, 7]. Addition of rituximab for CD20 positive DLBCL in combination with CHOP chemotherapy increases disease-free survival and life expectancy [9]. Combination of chemotherapy and radiotherapy has shown superior results in compare to chemotherapy alone [10]. Doria et al. [10] reported the superior results with combination of chemotherapy and radiotherapy in a retrospective study of 211 patients of stage IE and IIE thyroid lymphoma with the relapse rates of 7.7%, 37.1% and 43% for chemoradiotherapy, radiotherapy and chemotherapy alone, respectively. Local recurrence rates were 2.6%, 12.6% and 23%, respectively in favor of combination therapy. The five-year life expectancy after the treatment of primary thyroid DLBCL is 75% with excellent treatment outcomes [7, 10]. Stage III or IV, tumor size over 10 cm, lymph node involvement, mediastinal involvement, and presence of B symptoms are poor prognostic factors [7].

CONCLUSION

Primary thyroid lymphoma is a rare and unique disease, more commonly seen in females of 60–70 years age group. They present with rapidly growing masses

in the thyroid gland region. Hashimoto's thyroiditis is commonly associated with primary thyroid lymphoma. Ultrasonography guided fine-needle aspiration cytology and true cut needle biopsy from the thyroid lesion is required for the diagnosis and to differentiate with anaplastic carcinoma thyroid. Surgery/thyroidectomy is not recommended for primary thyroid lymphoma as chemotherapy and radiotherapy has excellent results and treatment outcomes.

****

Author Contributions

Tej Prakash Soni – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Upendra Nandwana – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Lalit Mohan Sharma – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Anil Kumar Gupta – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Naresh Jakhotia – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Shantanu Sharma – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Pedersen RK, Pedersen NT. Primary non-Hodgkin's lymphoma of the thyroid gland: A population based study. Histopathology 1996 Jan;28(1):25–32.
2. Chen C, Yang Y, Jin L, Dong L, Zhang X, Xiang Y. Primary thyroid T-lymphoblastic lymphoma: A case report and review of the literature. Int J Clin Exp Pathol 2013 Dec 15;7(1):443–50.
3. Katna R, Shet T, Sengar M, et al. Clinicopathologic study and outcome analysis of thyroid lymphomas: Experience from a tertiary cancer center. Head Neck 2013 Feb;35(2):165–71.
4. Sakorafas GH, Kokkoris P, Farley DR. Primary thyroid lymphoma (correction of lympoma): Diagnostic and therapeutic dilemmas. Surg Oncol 2010 Dec;19(4):e124–9.
5. Takashima S, Takayama F, Momose M, Shingu K, Sone S. Secondary malignant lymphoma which simulated primary thyroid cancer. Clin Imaging 2000 May-Jun;24(3):162–5.
6. Daneshbod Y, Omidvari S, Daneshbod K, Negahban S, Dehghani M. Diffuse large B cell lymphoma of thyroid as a masquerader of anaplastic carcinoma of thyroid, diagnosed by FNA: A case report. Cytojournal 2006 Oct 19;3:23.
7. Graff-Baker A, Roman SA, Thomas DC, Udelsman R, Sosa JA. Prognosis of primary thyroid lymphoma: Demographic, clinical, and pathologic predictors of survival in 1,408 cases. Surgery 2009 Dec;146(6):1105–15.
8. Pyke CM, Grant CS, Habermann TM, et al. Non-Hodgkin's lymphoma of the thyroid: Is more than biopsy necessary? World J Surg 1992 Jul–Aug;16(4):604–9; discussion 609–10.
9. Hiddemann W, Kneba M, Dreyling M, et al. Frontline therapy with rituximab added to the combination of cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP) significantly improves the outcome for patients with advanced-stage follicular lymphoma compared with therapy with CHOP alone: Results of a prospective randomized study of the German low-grade lymphoma study group. Blood 2005 Dec 1;106(12):3725–32.
10. Doria R, Jekel JF, Cooper DL. Thyroid lymphoma: The case for combined modality therapy. Cancer 1994 Jan 1;73(1):200–6.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Postherpetic spinal segmental paralysis

Lee Hui Jean, Pranav Kumar

ABSTRACT

Herpes zoster is a common viral infection, which is frequently seen in elderly or immunocompromised patients. It usually presented with painful neuropathy. However, it can rarely manifest as motor weakness. An 82-year-old male presented with L2–L4 myotomal weakness two-week post shingles. Diagnosis was made based on only history taking and physical examination. Other possible differential diagnoses were rule out through imaging and blood tests at that time. The patient made good recovery through analgesia and physiotherapy. Postherpetic spinal segmental paralysis is a rare complication arising from shingles. There was no consensus on diagnostic criteria and the underlying pathophysiology is still poorly understood. The main treatment modality of this condition is intensive physiotherapy and pain management.

Keywords: Herpes zoster, Myotome, Postherpetic spinal segmental paralysis

How to cite this article

Jean LH, Kumar P. Postherpetic spinal segmental paralysis. Int J Case Rep Images 2017;8(8):538–540.

Lee Hui Jean[1], Pranav Kumar[2]

Affiliations: [1]Medical Registrar, Redcliffe Hospital; [2]Staff Specialist, Mackay Base Hospital.

Corresponding Author: Dr. Pranav Kumar, Senior staff Specialist, Mackay Base Hospital, 4740, Australia; Email: drpranavkumar@gmail.com

Received: 07 February 2017
Accepted: 24 May 2017
Published: 01 August 2017

Article ID: Z01201708CR10817LJ

****

doi:10.5348/ijcri-201778-CR-10817

INTRODUCTION

Herpes zoster is a common viral infection and occurs in the sensory ganglia due to varicella zoster virus. It usually manifests with radicular pain and vesicular cutaneous eruptions along a single dermatome. It is a disease of sensory ganglia but motor involvement is rare and transient paresis can develop**.** Herpes zoster is frequently seen in elderly patients or those with compromised immune systems. Despite the advent of the zoster vaccine, an estimated 50% of those living to the age of 85 years will experience an episode of the disease [1]. This occurs in 3–5% of cases [1–3].

CASE REPORT

An 82-year-old male presented to a regional hospital with right leg numbness and weakness resulting recurrent mechanical falls. Two weeks ago, he was diagnosed with shingles by his general practitioner, involving L2 to L4 dermatome of his right lower limb. His significant past medical history includes non-metastatic prostate adenocarcinoma under remission, of which radiotherapy treatment was completed one year ago. He was not on any chemotherapy or immunosuppressive medications.

On systemic review, there were no sign and symptom suggestive of spinal cord compression, abscess or stroke. The patient was active with no physical limitation prior to hospitalization. Physical examination revealed crusting vesicles over the left lower back, left posterior and anteromedial thigh in L2 to 4 dermatomal distribution. He had normal vitals and afebrile. Lower limb neurological examination findings are given in Table 1. Plain films

Table 1: Lower limb neurological examination findings

Lower Limbs	Left	Right
Tone	Normal	Decreased
Hip flexion	5/5	3/5
Extension	5/5	2/5
Adduction	5/5	4/5
Abduction	5/5	4/5
Knee flexion	5/5	4/5
Extension	5/5	2/5
Ankle plantar flexion	5/5	5/5
Dorsiflexion	5/5	5/5
Reflexes patellar	++	-
Calcaneal	++	++
Babinski sign	down going	down going
Coordination foot tap	NAD	NAD
Heel shin test	NAD	Not assessed (due to weakness)
Sensation light touch	Normal	Diminished entire thigh and leg until proximal to ankle joint

and MRI scans did not report any evidence of transverse myelitis, spinal or nerve root compression. Bloods test was unremarkable, white cell count was normal and no further inflammatory marker was ordered at that time.

The patient was started on pregabalin and amitriptyline for neuropathic pain management, and was referred to rehabilitation program. Two weeks later, the patient mobility had significantly improved and he was discharged home with community follow-up.

DISCUSSION

Herpes zoster is a neurocutaneous condition caused by the reactivation of the varicella zoster virus. This commonly presents in people over 60 years of age as neuropathic pain follow by blister formation, which follows a dermatomal pattern.

Postherpetic neuralgia is the most common chronic condition arising from shingles, which may last from weeks to months after the blisters have healed. Herpes zoster ophthalmicus, bacterial super infection of the lesions, and cranial nerve or peripheral nerve palsies are less common sequela that could present post infection.

Postherpetic motor neuropathy is a rare complication of shingles which predominantly affects the facial nerves, followed by upper limbs, then lower limbs in incidence. As of the patient presentation, he elicited myotomal weakness of his right lower limb. This pattern of presentation therefore termed postherpetic spinal segmental paralysis, which is the subgroup of postherpetic motor neuropathy. In a case series of herpes zoster infection, Cohen reported only 0.8% of the patients had lower motor neuron paralysis [4]. Latency period of limb paralysis post initial onset of vesicular formation can range from one day to four months. The underlying pathophysiology of this condition is still poorly understood. Some literature postulates that the underlying pathophysiology is due to inflammatory demyelinating process and post infectious immune mediated motor root damage [5]. Some described this could due to hypervascularity in the perineural structure or disruption of blood nerve barrier secondary to inflammation [6, 7].

There was no consensus on the diagnostic criteria of this condition although multiple case reports in the literature had performed further investigations, such as inflammatory marker, CSF serology, nerve conduction study and electromyogram as part of their diagnostic workup. A Spanish retrospective study had collectively recruited patients who suffered from segmental motor paralysis analyzing the clinical findings, complementary investigations and their functional prognosis. Fifty percent of the patients have positive plasma or CSF varicella zoster serology and neurophysiological study showed denervation of the myotomes involved [8]. As of our case, neurophysiology facilities were unavailable in our hospital. We diagnosed the patient with postherpetic spinal segmental paralysis purely base on history and physical examination.

In general, 90% of the cases were just like the patient, who had motor weakness of the affected myotome corresponds to the dermatomal distribution of skin eruption [8]. However, we acknowledge there were existing literature suggest there could be no correlation between involved dermatome and myotome [9]. This might impose diagnostic challenge to the treating clinicians.

In our case, the affected myotome was L2 to L4 and associated with patchy sensory loss of which did not follow any dermatomal distribution. However, we acknowledge sensory loss in neurological examination can be highly subjective and easily skewed by his underlying painful neuropathy.

To date, there is not enough evidence in the literature suggesting commencement of steroid or antivirals therapy to promote functional recovery for established radiculopathy. Although, there is case series suggested the use of anti-viral could possibly prevent occurrence of paresis [10]. As of our case, the patient showed significant improvement in his mobility after having only intensive rehabilitation activities and pain management.

The differential diagnosis of lower limb polyradiculopathy is extensive. A thorough history taking and physical examination is warranted to establish the diagnosis.

CONCLUSION

Postherpetic spinal segmental paralysis is a rare complication of herpes zoster infection. Thorough

history taking and physical examination are essential in establishing diagnosis for this rare condition. Clinicians need to be aware not all patients have motor paralysis corresponds to dermatomal distribution of blisters. There is no evidence suggests antiviral therapy or steroid promotes functional recovery for patients who suffered from postherpetic spinal segmental paralysis.

****

Author Contributions

Hui Jean Lee – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Pranav Kumar – Substantial contributions to conception and design, Acquisition of data, Drafting the article, revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Johnson RW, Whitton TL. Management of herpes zoster (shingles) and postherpetic neuralgia. Expert Opin Pharmacother 2004 Mar;5(3):551–9.
2. Rastegar S, Mahdavi SB, Mahmoudi F, Basiri K. Herpes zoster segmental paresis in an immunocompromised breast cancer woman. Adv Biomed Res 2015 Aug 10;4:170.
3. Sifuentes Giraldo WA, de la Puente Bujidos C, de Blas Beorlegui G, López San Román A, Peña Arrebola A. Herpes zoster motor neuropathy in a patient with previous motor paresis secondary to Vogt-Koyanagi-Harada disease. Am J Phys Med Rehabil 2013 Apr;92(4):351–6.
4. Cohen JI. Clinical practice: Herpes zoster. N Engl J Med 2013 Jul 18;369(3):255–63.
5. Wilson JF. In the clinic: Herpes zoster. Ann Intern Med 2011 Mar 1;154(5):ITC31–15; quiz ITC316.
6. Hata A, Kuniyoshi M, Ohkusa Y. Risk of Herpes zoster in patients with underlying diseases: A retrospective hospital-based cohort study. Infection 2011 Dec;39(6):537–44.
7. Hanakawa T, Hashimoto S, Kawamura J, Nakamura M, Suenaga T, Matsuo M. Magnetic resonance imaging in a patient with segmental zoster paresis. Neurology 1997 Aug;49(2):631–2.
8. Lu PJ, O'Halloran A, Williams WW, Harpaz R. National and state-specific shingles vaccination among adults aged =60 Years. Am J Prev Med 2017 Mar;52(3):362–72.
9. Cockerell OC, Ormerod IE. Focal weakness following herpes zoster. J Neurol Neurosurg Psychiatry 1993 Sep;56(9):1001–3.
10. Sampathkumar P, Drage LA, Martin DP. Herpes zoster (shingles) and postherpetic neuralgia. Mayo Clin Proc 2009 Mar;84(3):274–80.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Immunosuppression and infectious complications: A hemorrhagic dilemma

Isabelle Malhamé, Amanda Farag, Zhao Gao, Alan Barkun

ABSTRACT

Introduction: Patients treated with immunosuppressive agents for autoimmune diseases are a unique patient population. Simultaneous management of an underlying autoimmune disease and of an opportunistic infection can present a therapeutic dilemma. Case Report: A 68-year-old female presented with signs and symptoms of vasculitis involving the gastrointestinal tract. After initiation of immunosuppressive treatment, she developed an infectious complication that led to intractable gastrointestinal bleeding. Conclusion: Current recommendations addressing the management of cytomegalovirus infection in the context of a severe vasculitis are lacking, and our approach to this case may benefit physicians dealing with this clinical dilemma in the future.

Keywords: Cytomegalovirus, Immunosuppression, Vasculitis

Isabelle Malhamé[1], Amanda Farag[2], Zhao Gao[3], Alan Barkun[2,4]

Affiliations: [1]MD, Department of Internal Medicine, McGill University Health Center; [2]MD, Division of Gastroenterology, McGill University Health Center; [3]MD, Department of Pathology, McGill University Health Center; [4]MD, Clinical Epidemiology, McGill University Health Center.

Corresponding Author: Alan Barkun, 1001 Decarie Blvd, Montreal, QC, Canada H4A 3J1; Email: Alan.barkun@muhc.mcgill.ca

Received: 07 April 2017
Accepted: 02 May 2017
Published: 01 August 2017

How to cite this article

Malhamé I, Farag A, Gao Z, Barkun A. Immunosuppression and infectious complications: A hemorrhagic dilemma. Int J Case Rep Images 2017;8(8):541–544.

Article ID: Z01201708CR10818IM

****

doi:10.5348/ijcri-201779-CR-10818

INTRODUCTION

Patients treated with immunosuppressive agents for autoimmune diseases affecting the gastrointestinal tract present a unique and complex challenge. When the symptoms of an opportunistic infection are similar to the underlying autoimmune disease, clinical management becomes even more difficult. We believe that our approach to this case may benefit physicians caring for such patients.

CASE REPORT

A 68-year-old female presented to the emergency department of a peripheral hospital for fatigue and palpable purpurae. Her past medical history included mild hypertension, hypothyroidism, and diabetes, for which she was taking calcium channel blockers, thyroxine, and an oral anti-hyperglycemic. One week after her initial visit, she developed polyarthritis, melena and bright red blood per rectum. Despite initiation of 60 mg of oral prednisone, her symptoms did not improve and she went on to develop hypoxic respiratory failure. In view of her clinical deterioration, she was transferred to a tertiary care hospital. Upon admission, she had both

upper and lower gastrointestinal bleeding symptoms requiring volume resuscitation as well as transfusions.

Given the constellation of polyarthritis, palpable purpurae, melena, and possible alveolar hemorrhage, we strongly suspected a vasculitic process with gastrointestinal involvement such as an eosinophilic granulomatosis with polyangiitis (EGPA), granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), polyarteritis nodosa (PAN), and rheumatoid arthritis associated vasculitis (RAAV) [1]. Pertinent laboratory examinations revealed normal platelet count, INR, eosinophil count, immunoglobulin profile, C3-4 levels, as well as negative C-anca, P-anca, anti-GBM, cryoglobulin, lupus anticoagulants, and rheumatoid factor. She also had negative HIV, hepatitis C and B screens. The patient's bleeding originated from the distal ileum on capsule endoscopy, CT-scan with contrast, and colonoscopy. She also had diffuse mucosal ulcerations throughout the digestive tract (Figure 1). Given the strong suspicion of underlying vasculitis, the treating team initiated a three days course of 1 g of intravenous methylprednisolone daily. Despite the steroid treatment, and two attempts at arterial embolization, the patient continued to pass large amounts of bloody stools. On day-7 of her admission, given the growing risk of bowel ischemia and perforation associated with subsequent arterial embolization, the patient underwent urgent surgical exploration with resection of the involved bowel. Intra-operative endoscopy visualized the entire small bowel lumen with active arterial bleeding and ulcerations in the proximal and terminal ileum. She had a right hemi-colectomy and loop ileostomy with extended resection of the terminal ileum. The biopsy, which resulted on the second week of hospitalization, suggested a polyarteritis nodosa (PAN) involving the ileum and colon with superimposed cytomegalovirus infection (Figure 2).

The patient was treated with ganciclovir for her infection and rituximab for PAN as a less aggressive treatment alternative to cyclophosphamide. The bleeding persisted from the ileostomy despite a decreasing cytomegalovirus viremia. Given the active PAN, which was thought to be responsible for the ongoing hemorrhage, a course of cyclophosphamide was initiated. The patient remained in the ICU for one month, during which she developed hospital-acquired infections including repeated bacteremiae with *Staphylococcus aureus*, and *Candida dubliniensis*.

After a brief resolution of the intestinal hemorrhage, the patient had a recurrence of profuse bleeding through the ileostomy. While a repeat gastroscopy and ileoscopy demonstrated deep ulcerations compatible with ischemia of both duodenum and ileum, an explorative laparotomy did not demonstrate transmural necrosis; resection was therefore not carried out. In view of her overall deteriorating condition despite ongoing therapy, it was decided with the patient's family to pursue a palliative approach and the patient died on day-70 day of admission.

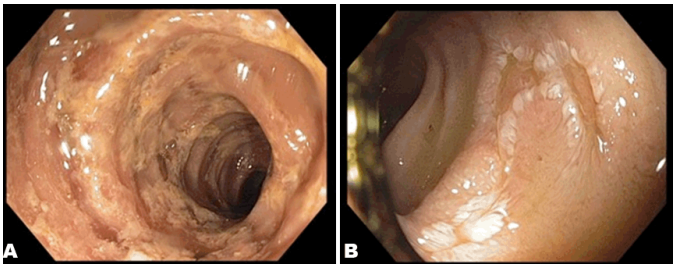


Figure 1: Endoscopic appearance of diffuse (A) Duodenal and (B) Colonic mucosal ulcerations.

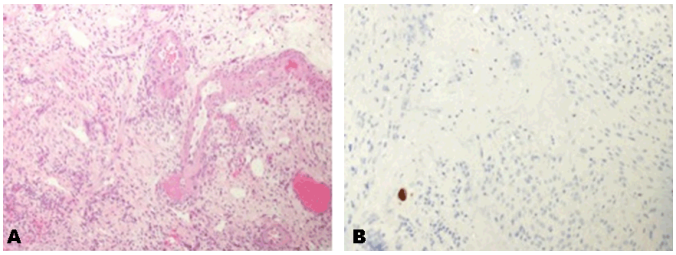


Figure 2: Presence of co-existing vasculitis and cytomegalovirus infection as evidenced by (A) Adjacent arteries showing fibrinoid necrosis of a segment of vessel wall in the small bowel, and (B) Scattered cytomegalovirus positive cells in the colonic submucosa.

DISCUSSION

Polyarteritis nodosa is a necrotizing, focal segmental vasculitis of medium-sized arteries [1]. The outcome of patients treated for PAN has greatly improved in the last two decades [2]. In this cohort, 37.9% of patients diagnosed with PAN had gastrointestinal involvement, and gastrointestinal manifestations requiring surgery at diagnosis were independent predictors of mortality [2].

Traditionally, hepatitis B has been identified as a possible viral trigger, but infections with cytomegalovirus have also been reported [3–5]. In a multi-centric retrospective survey on cytomegalovirus infection among patients hospitalized with autoimmune diseases in Japan, oral prednisone, pulsed methylprednisolone, and cyclophosphamide were most associated with reactivation [6]. Reactivation of cytomegalovirus, whether previously documented or not, should therefore be considered to be a serious potential complication of the mainstay treatment of vasculitis. In the case of our patient, it is reasonable to believe that initial treatment of her vasculitis with prednisone and methylprednisolone precipitated the cytomegalovirus reactivation.

Expert opinion on vasculitides in the context of viral infections suggests prompt intravenous antiviral treatment and discourages the use of corticosteroid and/or immune-suppressants for patients who are already immunosuppressed [3]. However, the combination of corticosteroids with cyclophosphamide remains the cornerstone of treatment, and herein lays the dilemma [7]. If we were to solely treat the cytomegalovirus we might be undertreating the vasculitis, which could worsen the

patient's prognosis. Similarly, only treating PAN could provoke more profound immunosuppression and lead to potentially fatal progression of the cytomegalovirus infection. In this context, we treated the PAN and the cytomegalovirus simultaneously and we monitored the clinical activity of the infection via serial measurements of the cytomegalovirus viral load. As the cytomegalovirus viremia was consistent with a resolution of the infection, we believe that uncontrolled PAN was the cause of the persistent hemorrhage.

The clinical course of the patient we present is thus characteristic of the rare difficult clinical scenario in which patients with a vasculitis have active cytomegalovirus infection, requiring both antiviral and aggressive immunosuppressive therapies. To the best of our knowledge, there have only been a handful of cases in the literature of active cytomegalovirus infection in the presence of an acute PAN presentation. The four patients described in these reports were treated with corticosteroids and/or antiviral drugs in the absence of any additional immunosuppressant therapy. Mayer et al. describes the effectiveness of less aggressive treatment without immuno-suppressants for ANCA-associated vasculitis with a concomitant viral infection. Unfortunately, recommendations remain lacking in this rare group of patients [4, 8].

CONCLUSION

In conclusion, the patient discussed highlights a unique diagnostic and therapeutic dilemma. The co-existence of active cytomegalovirus infection and severe polyarteritis nodosa resulted in having to choose between possible conflicting therapeutic aims. Clinicians should consider altering standard aggressive combination therapy for the vasculitis while administering antiviral treatment in such patients.

****

Author Contributions

Isabelle Malhamé – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Amanda Farag – Substantial contribution to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Zhao Gao – Substantial contribution to acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Alan Barkun – Substantial contribution to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

REFERENCES

1. Pagnoux C, Mahr A, Cohen P, Guillevin L. Presentation and outcome of gastrointestinal involvement in systemic necrotizing vasculitides: Analysis of 62 patients with polyarteritis nodosa, microscopic polyangiitis, Wegener granulomatosis, Churg-Strauss syndrome, or rheumatoid arthritis-associated vasculitis. Medicine (Baltimore) 2005 Mar;84(2):115–28.
2. Pagnoux C, Seror R, Henegar C, et al. Clinical features and outcomes in 348 patients with polyarteritis nodosa: A systematic retrospective study of patients diagnosed between 1963 and 2005 and entered into the French vasculitis study group database. Arthritis Rheum 2010 Feb;62(2):616–26.
3. Pagnoux C, Cohen P, Guillevin L. Vasculitides secondary to infections. Clin Exp Rheumatol 2006 Mar–Apr;24(2 Suppl 41):S71–81.
4. Doherty M, Bradfield JW. Polyarteritis nodosa associated with acute cytomegalovirus infection. Ann Rheum Dis 1981 Aug;40(4):419–21.
5. Kouchi M, Sato S, Kamono M, et al. A case of polyarteritis nodosa associated with cytomegalovirus infection. Case Rep Rheumatol 2014;2014:604874.
6. Takizawa Y, Inokuma S, Tanaka Y, et al. Clinical characteristics of cytomegalovirus infection in rheumatic diseases: multicentre survey in a large patient population. Rheumatology (Oxford) 2008 Sep;47(9):1373–8.
7. Guillevin L, Cohen P, Mahr A, et al. Treatment of polyarteritis nodosa and microscopic polyangiitis with poor prognosis factors: A prospective trial comparing glucocorticoids and six or twelve cyclophosphamide pulses in sixty-five patients. Arthritis Rheum 2003 Feb 15;49(1):93–100.
8. Meyer MF, Hellmich B, Kotterba S, Schatz H. Cytomegalovirus infection in systemic necrotizing vasculitis: Causative agent or opportunistic infection? Rheumatol Int 2000 Dec;20(1):35–8.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Morel–Lavallée lesions: A rare cause of post-traumatic lower back and hip pain

Yuzeng Shen

ABSTRACT

Introduction: Lower back and hip pain after falling is one of the common presenting complaints at the emergency department. Case Report: We present a rare case of an 80-year-old female with Morel–Lavallée lesions as a cause of lower back and hip pain after a fall. She was managed conservatively with improvement in her symptoms. Conclusion: Morel–Lavallée lesions are a rare cause for post-traumatic back and hip pain and may respond well to conservative non-operative management. It should be considered as a differential diagnosis for patients with persistent pain despite unremarkable plain X-rays done during initial evaluation at the emergency department.

Keywords: Closed degloving injury, Morel–Lavallée lesion, Soft tissue injury, Trauma

How to cite this article

Shen Y. Morel–Lavallée lesions: A rare cause of post-traumatic lower back and hip pain. Int J Case Rep Images 2017;8(8):545–548.

Article ID: Z01201708CR10819YS

Yuzeng Shen

Affiliation: Associate Consultant, Department of Emergency Medicine, Singapore General Hospital, Singapore.

Corresponding Author: Yuzeng Shen, Contact Address: Department of Emergency Medicine, Singapore General Hospital, Outram Road, Singapore - 169608; Email: shen.yuzeng@singhealth.com.sg

Received: 22 May 2017
Accepted: 05 June 2017
Published: 01 August 2017

****

doi:10.5348/ijcri-201780-CR-10819

INTRODUCTION

Lower back and hip pain after falling is one of the common presenting complaints at the emergency department.

The initial investigation of choice, depending on clinical history and physical examination findings, is the plain X-ray to rule out fractures and dislocations. Patients with unremarkable X-rays often have contusions or muscular-ligamentous sprains and strains, which respond well to analgesia and rest. In patients with persistently significant pain, advanced imaging is often performed to identify pathology not well seen on plain X-rays.

Morel–Lavallée lesions are associated with trauma and shearing injury, leading to a closed degloving injury where the inter-fascial planes between subcutaneous tissue and underlying fascia is separated. The underlying pre-fascial space may subsequently be filled with blood, lymph or serous fluid [1, 2]. We present a case of traumatic lower back and hip pain due to Morel–Lavallée lesions.

CASE REPORT

An 80-year-old female presented to the emergency department after sustaining a fall in the bathroom. The patient is currently taking an anti-platelet agent (clopidogrel) for presumptive ischemic heart disease. She slipped and fell off the toilet seat on the day prior to her emergency department visit, and landed on her hips. Since then, she complained of persistent lower back and right hip pain.

Vital signs at the patient are as follows: temperature 36.2°C, pulse rate of 83 beats per minute, respiratory rate at 18 breaths per minute, blood pressure 120/57 mmHg, pulse oximetry 96% on room air, and pain score of 8/10. On physical examination, significant findings were that

of midline tenderness over the lumbosacral spine, and pain on internal and external rotation of right hip. X-rays of the lumbar spine, pelvis and right hip did not show any obvious displaced fracture.

Patient was admitted to the orthopedics department for further observation and workup in view of persistent lower back and right hip pain despite trial of observation and analgesia at the emergency department. Subsequently, inpatient magnetic resonance imaging of the pelvis and bilateral hips showed pre-fascial fluid at the lateral aspect of bilateral hips and posteriorly at the level of the lumbosacral junction, suggestive of Morel–Lavallée lesions.

All identified Morel–Lavallée lesions were small (Figures 1 and 2). The right hip Morel–Lavallée lesion had a maximum width of 2.5 mm, and the left hip Morel–Lavallée lesion had a maximum width of 5 mm. Both collections above extend along the entire lateral aspect of the bilateral gluteus maximus. The third Morel–Lavallée lesion was superficial to the erector spinae muscles and extended from the level of the lumbosacral joint proximally with a maximum depth of 3.7 mm.

The patient was managed conservatively with oral paracetamol and oral tramadol tablets for analgesia. She did not undergo surgery or aspiration for the Morel–Lavallée lesions and was discharged after symptoms improved during her stay in hospital. No intervention was performed due to minimal depth of the identified Morel–Lavallée lesions. The patient was a tourist who wished to return to her home country for further review. Due to her continuing management within her home country, further follow-up regarding her progress was not possible.

DISCUSSION

Morel–Lavallée lesions, also known as closed degloving injury of the soft tissue, are rare and are usually due to shearing injury and disruption between subcutaneous tissue and the underlying fascia. Less commonly, it may present as a post-surgical complication, especially associated with liposuction. The space created when subcutaneous tissue separates from the underlying

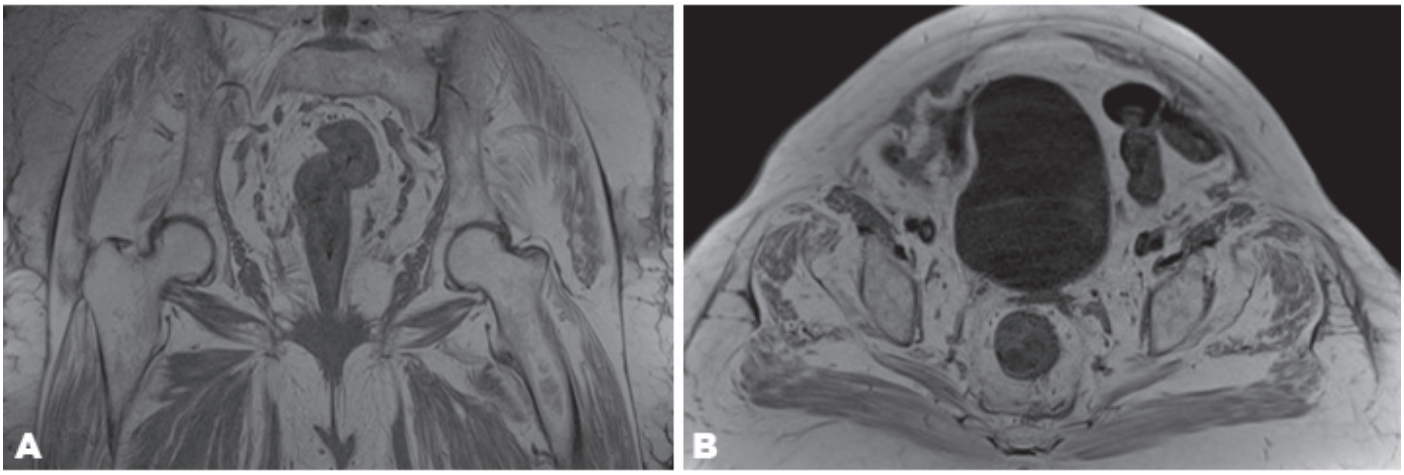


Figure 1: (A) Magnetic resonance imaging T1 turbo spin-echo (TSE) coronal cut, (B) Magnetic resonance imaging T1 turbo spin-echo (TSE) transverse cut.

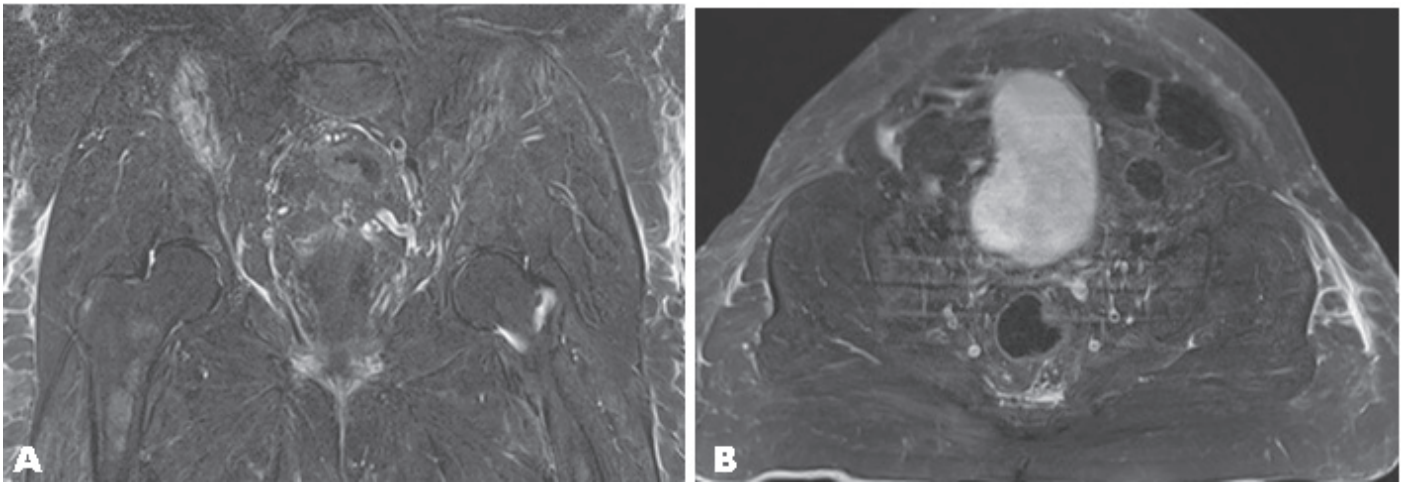


Figure 2: (A) Magnetic resonance imaging T2 turbo inversion recovery magnitude (TIRM) coronal cut, (B) Magnetic resonance imaging T2 turbo inversion recovery magnitude (TIRM) transverse cut.

fascia may subsequently be filled with blood, lymph or serous fluid [1]. The patient being on an anti-platelet agent may have contributed to increased bleeding risk and hematoma formation within the resultant space.

Common sites of injury include the greater trochanter and proximal thigh [3]. Possible mimics include hemorrhagic bursitis and soft tissue neoplasms. Advanced imaging with MRI is the investigation of choice for diagnosis [1]. Point of care ultrasound has been shown to be a useful and quick alternative in diagnosing Morel–Lavallée lesions [4].

Like this case, Morel–Lavallée lesions are commonly missed during initial evaluation [5], and may present with persistent pain and/or swelling despite rest and analgesia. They can be managed conservatively with compression therapy, physiotherapy, bed rest, or surgical aspiration, drainage or debridement [1, 6, 2]. Left untreated, complications include infection, expanding lesion and tissue necrosis due to mass effect.

There are no large studies providing quality evidence with regards to the management of Morel–Lavallée lesions. In a systemic review of 21 articles detailing 153 patients with Morel–Lavallée lesions of the peripelvic region [7], it was found that surgical intervention was better than conservative management. However, most articles within the systemic review were deemed as Level IV studies.

Nickerson et al. shared the creation of a practice management guideline for Morel–Lavallée lesions at the Mayo clinic [8], based on 79 patients with Morel–Lavallée lesions. The study recommended that aspiration of more than 50 ml of fluid from a Morel–Lavallée lesion would prompt surgical intervention, due to a higher rate of recurrence (83% vs 33% when less than 50 ml of fluid was aspirated). Patients can otherwise be conservatively managed with compression and observation. However, it was also mentioned that the study was limited to Level IV evidence, and would require further evaluation in the future.

CONCLUSION

This case report highlights a rare cause of post traumatic lower back and hip pain. Ultrasound evaluation is a useful investigation at the emergency department to identify Morel–Lavallée lesions, which may respond well to conservative nonoperative management. It should be considered as a differential diagnosis for patients with persistent pain despite unremarkable plain X-rays done during initial evaluation at the emergency department.

****

Author Contribution

Yuzeng Shen – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Bonilla-Yoon I, Masih S, Patel DB, et al. The Morel-Lavallée lesion: Pathophysiology, clinical presentation, imaging features, and treatment options. Emerg Radiol 2014 Feb;21(1):35–43.
2. Tejwani SG, Cohen SB, Bradley JP. Management of Morel–Lavallée lesion of the knee: Twenty-seven cases in the national football league. Am J Sports Med 2007 Jul;35(7):1162–7.
3. Hak DJ, Olson SA, Matta JM. Diagnosis and management of closed internal degloving injuries associated with pelvic and acetabular fractures: The Morel-Lavallée lesion. J Trauma 1997 Jun;42(6):1046–51.
4. LaTulip S, Rao RR, Sielaff A, Theyyunni N, Burkhardt J. Ultrasound utility in the diagnosis of a Morel-Lavallée lesion. Case Rep Emerg Med 2017;2017:3967587.
5. Kottmeier SA, Wilson SC, Born CT, Hanks GA, Iannacone WM, DeLong WG. Surgical management of soft tissue lesions associated with pelvic ring injury. Clin Orthop Relat Res 1996 Aug;(329):46–53.
6. Tejwani SG, Cohen SB, Bradley JP. Management of Morel–Lavallée lesion of the knee: twenty-seven cases in the national football league. Am J Sports Med 2007 Jul;35(7):1162–7.
7. Shen C, Peng JP, Chen XD. Efficacy of treatment in peri-pelvic Morel–Lavallée lesion: A systematic review of the literature. Arch Orthop Trauma Surg 2013 May;133(5):635–40.
8. Nickerson TP, Zielinski MD, Jenkins DH, Schiller HJ. The Mayo Clinic experience with Morel-Lavallée lesions: Establishment of a practice management guideline. J Trauma Acute Care Surg 2014 Feb;76(2):493–7.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Familial congenital hypothyroidism due to thyroid dysgenesis: A case report of largest family

Abhinav Kumar Gupta, Syed Mohd. Razi, Deepak Chand Gupta, Saqib Ahmad Khan, Pankaj Jain, Keshav Kumar Gupta

ABSTRACT

Introduction: Congenial hypothyroidism due to thyroid dysgenesis is usually regarded as sporadic. However, a small but significant proportion of familial cases have been identified (2%). Herein, we describe a case report of unusually large family of 10 siblings, out of which five were affected with congenital hypothyroidism, which is supposed to be the world's largest series of familial congenital hypothyroidism due to thyroid dysgenesis. Case Report: We describe a family of 10 siblings out of which five presented to endocrine department with complaints of lethargy, constipation, hoarseness of voice, edema, short stature and mental retardation. The eldest affected sibling was 25-year-old while the youngest affected child was 10-year-old. All the affected siblings had feeding difficulties, delayed fine motor, gross motor and social developmental milestones along with subnormal intelligence. On evaluation they were found to be grossly hypothyroid. Complete absence of thyroid tissue was found on ultrasonography and 99mTc pertechnetate scan. Conclusion: Familial congenital hypothyroidism due to thyroid dysgenesis is a rare finding; timely evaluation and proper genetic counseling can prevent disastrous consequences.

Keywords: Athyreosis, Familial congenital hypothyroidism, Sibling, Thyroid dysgenesis

Abhinav Kumar Gupta[1], Syed Mohd. Razi[2], Deepak Chand Gupta[1], Saqib Ahmad Khan[1], Pankaj Jain[3], Keshav Kumar Gupta[4]

Affiliations: [1]Senior resident, Department of Endocrinology, L.L.R.M. Medical College, Meerut, Uttar Pradesh, India; [2]Consultant endocrinologist, Department of Endocrinology, Sri Sai hospital, Muradabad, Uttar Pradesh, India; [3]Assistant Professor, Department of pediatrics, S.M.S. Medical College, Jaipur, Rajsthan, India; [4]Professor, Department of Endocrinology, L.L.R.M. Medical College, Meerut, Uttar Pradesh, India.

Corresponding Author: Abhinav Kumar Gupta, D-38, L.L.R.M. Medical College, Meerut, Uttar Pradesh, India, 250004; Email: abhinavgupta2026@gmail.com

Received: 29 March 2017
Accepted: 01 June 2017
Published: 01 August 2017

How to cite this article

Gupta AK, Mohd. Razi S, Gupta DC, Khan SA, Jain P, Gupta KK. Familial congenital hypothyroidism due to thyroid dysgenesis: A case report of largest family. Int J Case Rep Images 2017;8(8):549–554.

Article ID: Z01201708CR10820AG

****

doi:10.5348/ijcri-201781-CR-10820

INTRODUCTION

Congenital hypothyroidism (CH) is one of the most common preventable causes of mental retardation. Incidence of CH was initially reported to be in the range of 1:3000 to 1:4000. With the help of national screening programs, it has become apparent that the incidence varies by geographic location. A recent report showed that the incidence in the United States increased from 1:4094 in 1987 to 1:2373 in 2002. Incidence varies among different racial and ethnic groups, gender, birth weight, single versus multiple births and according to mother's age [1].

Eighty-five percent of cases of CH are caused by thyroid developmental defects (dysgenesis) and

remaining 15% of cases occur due to the defects in thyroid hormone biosynthesis (dyshormonogenesis). The pathogenesis of thyroid dysgenesis (TD) is still unknown, and the disease is usually regarded as sporadic with a female predominance. Possible roles of autoimmune or unidentified environmental factors have been suggested but not confirmed. Some familial cases of CH caused by thyroid dysgenesis have been reported with either athyreosis or ectopic gland in affected members. French study reported a significant important proportion of familial cases of CH due to thyroid dysgenesis (2%), affirming the existence of strong familial component in CH due to thyroid dysgenesis [2].

CASE REPORT

In this case report, we describe an Asian–Indian family of 10 siblings out of which five presented to endocrinology outpatient clinic with complaints of lethargy, constipation, hoarseness of voice, edema, short stature and mental retardation. The eldest affected sibling was 25-year-old while the youngest affected child was 10-year-old.

All the children were born to a hypothyroid mother, out of a non-consanguineous marriage with full term normal vaginal delivery. The mother did not give history of thyroxin intake during any of the pregnancies. All the pregnancies and ante-partum periods were uneventful. There was no history suggestive of delayed cry, birth hypoxia, prolonged jaundice, in any of the affected siblings. All the affected siblings had feeding difficulties, delayed fine motor, gross motor and social developmental milestones along with subnormal intelligence. Detailed family history revealed demise of two siblings at the ages of two years and 32 years respectively other than these five affected siblings, with the similar complaints while the remaining three are normal (Figure 1).

On general examination all the affected siblings were having typical puffy hypothyroid facies with cold dry skin, sparse fragile hair, lateral madarosis, short round nose, macroglossia, edema, hoarseness of voice and bradycardia (Figure 2). Calf muscle pseudo-hypertrophy and umbilical hernia were present in two of the affected siblings (Figures 3 and 4). There was absence of goiter, hearing difficulty, cleft palate, spiky hair, low posterior hairline, low set ear and abnormal movement. On anthropometric examination all affected siblings had significant short stature. Sexual maturity was corresponding with the chronological age in all affected siblings. Systemic examination of all patients was unremarkable except for delayed relaxation of deep tendon reflexes and pseudomyotonia.

Laboratory investigations revealed normal hemogram, renal function test, hepatic function test, electrolyte, LH, FSH and prolactin. Tests also revealed very high level of TSH along with very low levels of T4 and T3. Anti-thyroid peroxidase antibody (Anti-TPO) and anti-thyroglobulin (Anti-TG) were negative in all siblings. Serum thyroglobulin was very low in all the affected siblings (Table 1). X-ray of left hand revealed significant delay in bone age and epiphyseal dysplasia (Figure 5). Ultrasonography of the thyroid gland revealed complete absence of thyroid tissue in all affected siblings. Maternal laboratory testing revealed normal anti-TPO and normally located thyroid gland on ultrasonography. Father's thyroid function tests were normal. On Tc99m pertechnetate scan, there was no radioactive tracer uptake at any location. All the hormonal investigations were done by chemiluminescence immunoassay using Abbott ARCHITECT i1000sr immunoassay analyzer (USA).

On the basis of typical clinical history, physical examination and investigations all five siblings were diagnosed as cases of CH. They were started on thyroxine replacement and dose titrated by periodic laboratory investigation.

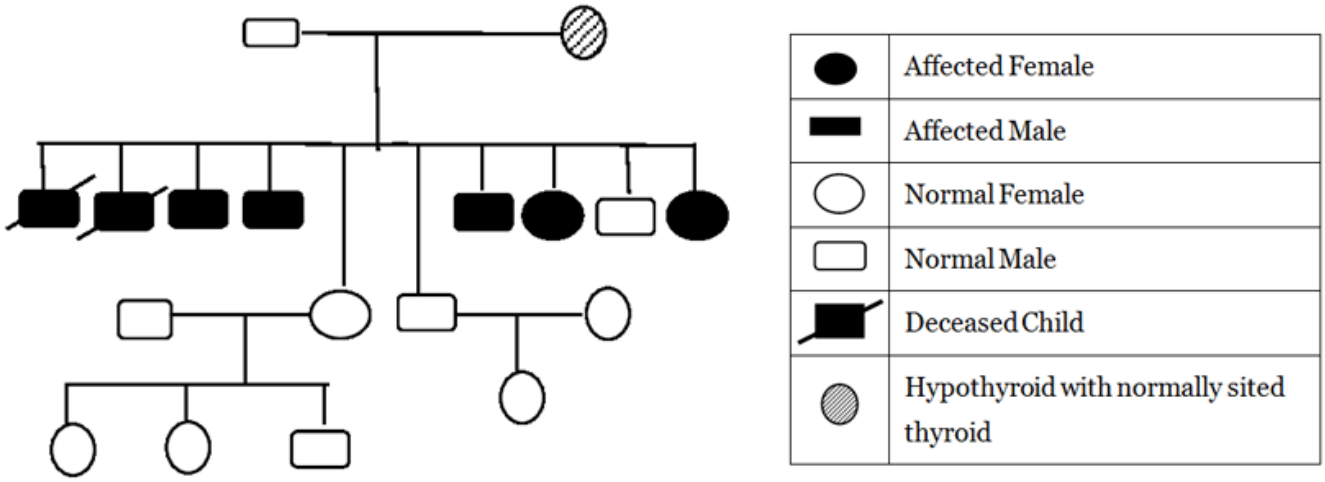


Figure 1: Family pedigree.

Figure 2: The characteristic clinical features: Coarse facial features, puffiness of face, short stature.

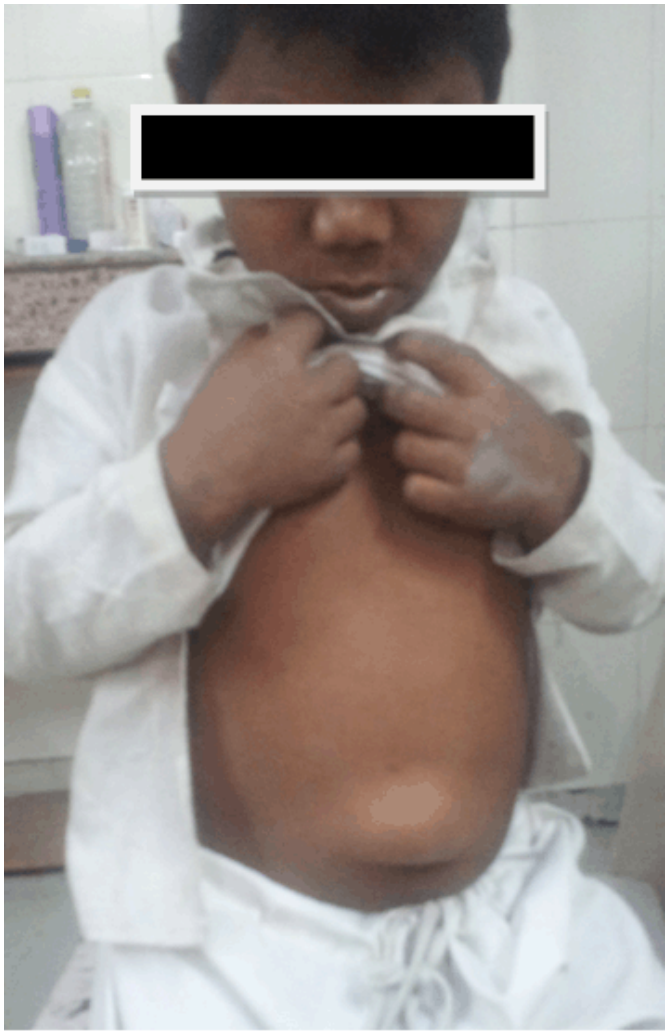


Figure 3: Umbilical hernia.

DISCUSSION

Congenital hypothyroidism is defined as reduced level of thyroid hormones, which may be symptomatic at birth or later. It may be either due to thyroid dysgenesis

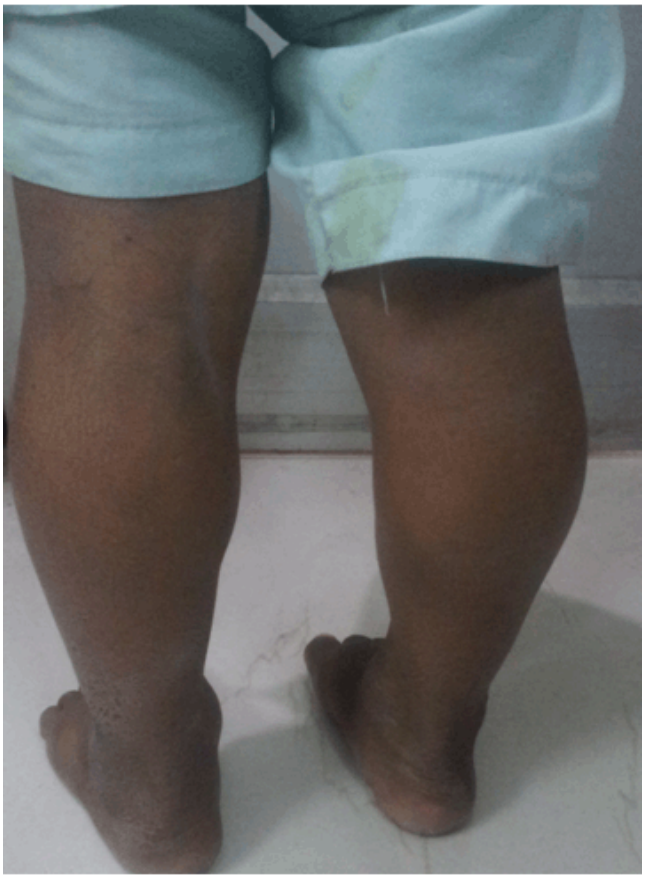


Figure 4: Pseudohypertrophy of calf muscles.

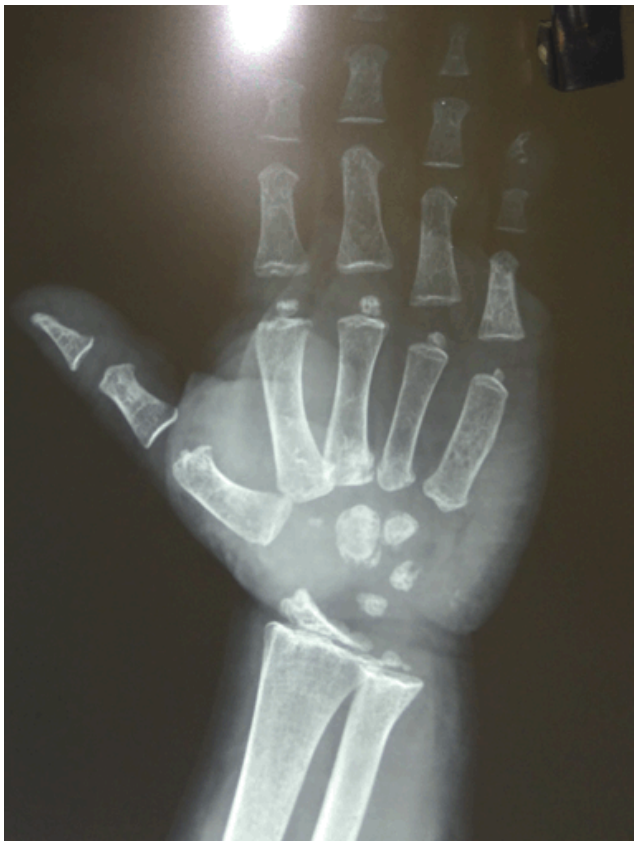


Figure 5: X-ray left hand showing epiphysis dysplasia.

Table 1: Anthropometry and hormonal profile

Parameter	1st is Male	2nd is Male	3rd is Female	4th is Male	5th is Female	Normal Range
Age (years)	25	22	19	17	10	
Weight (kg)	20	24	37	28	15	
Height (cm)	100	113	122	124	98	
TSH	424	478	470	318	398	0.34–4.25 µIU/ml
T4	1.67	1.23	1.43	1.98	1.97	5.4–11.7 µg/dl
T3	<0.25	<0.25	<0.25	<0.25	<0.25	77–135 ng/dl
Anti-TPO	0.66	0.59	0.62	0.75	0.52	<35 IU/l
Thyroglobulin	5.29	3.2	4.3	5.8	6.4	13–318 ng/ml
Antithyroglobulin	<15	<15	<15	<15	<15	<40 IU/ml
Luteinizing Hormone	1.47	2.59	0.10	1.8	0.05	Prepubertal:<3.36 mIU/ml Pubertal: 0.69–7.15 mIU/ml
FSH	6.14	5.34	5.45	1.7	5.86	Prepubertal: 0.3–1.67 mIU/ml Pubertal:0.4-6.5 mIU/ml
Prolactin	27.4	31.5	19.3	24.2	32	0.68–0.12.4 ng/ml
Testosterone	922	643	-	431	-	269–1066 ng/dl
Estradiol	-	-	29		<10	Prepubertal: <6 pg/ml

Abbreviations: TSH: Thyroid Stimulating Hormone, Anti-TPO: Anti-Thyroid Peroxidase Antibodies, FSH: Follicular Stimulating Hormone

(abnormal gland development) or dyshormonogenesis (disorder of thyroid hormone biosynthesis). Congenital hypothyroidism is classified as permanent and transient. Permanent CH refers to a persistent deficiency of thyroid hormone requiring life-long replacement. Transient CH is deficiency of thyroid hormone at birth which recovers in neonatal period or later. Causes of transient CH include iodine deficiency, transfer of maternal blocking antibodies, fetal exposure to anti-thyroid drugs, maternal iodine exposure, neonatal iodine exposure and mutation in DUOX2 and DUOXA2 [1].

Most of the cases of permanent CH are related to TD due to abnormal thyroid gland development involving various defects, such as thyroid ectopy, athyreosis and thyroid hypoplasia. About 5% of the cases have been shown to result from the mutations in genes involved in thyroid development, but most cases of TD are sporadic and their pathogenesis remains unknown. A high frequency of congenital cardiac defect in TD support the role of genetic component and reports of some familial cases of CH due to TD resulting from athyreosis and ectopic gland is consistent with the notion of an inherited disease [3].

Anger and Kelley described unusual occurrence of athyreotic cretinism in three siblings of Maxican-American descent who were born out of consanguineous marriage [4]. Cross et al. observed two sisters with cretinism and Kocher–Debre–Semelaigne syndrome (myotonia and muscular pseudohypertrophy). Although no thyroid tissue was palpable but on investigation small thyroid tissue was present [5]. Kaplan et al. described two non-consanguineous Ashkenazi Jewish families, in each of which, a brother and sister had hypothyroidism, associated with ectopia and hypoplasia of the thyroid [6]. Yano et al. described athyreotic CH in two sisters born out of non-consanguineous parents in Japan [7].

Positive family history was found in 2% of CH patients with TD [8]. Castanet et al. compared familial and sporadic cases of CH. He identified 67 patients were having family history of CH with TD, out of them only two families had three or four affected members, all other families were having ≤2 affected members. He observed significantly higher number of familial cases than it would occur by chance only, hence drawn the conclusion that genetic factors could be involved in TD [2]. Almost 20 families of CH due to TD have been described in last 40 years, which again implies that TD may be familial.

A number of genes which are expressed during thyroid embryogenesis have been implicated in TD, which includes paired box gene eight (PAX8), TTF-2, NKX2.1 and NKX 2.5.

Seven members of a non-consanguineous hypothyroid family with autosomal dominant mode of inheritance studied by Grasberger et al. had striking variable presentation. The proband and her brother who had elevated TSH and low free T4 on neonatal screening had normally placed gland on scintigraphy. At age of 37 years, their mother was found to have mild hypothyroidism on routine investigation. A female cousin had athyreosis along with elevated TSH and very low freeT4 on neonatal screening. Her five-year brother found to have elevated TSH and normal free T4 while their father was a hypothyroid since five years of age. A 67-year-old grandmother of the cousins had moderate thyroid

failure on biochemical study. Sequence analysis revealed heterozygosity for PAX8 gene mutations among affected family members [9].

In this case report, we describe a family with five siblings who are diagnosed to have CH due to TD. This is supposed to be the family with maximum number of affected individuals reported so far in the medical literature. All siblings had athyreosis as a cause of CH. Mother is also hypothyroid with normally situated thyroid gland. Although common unidentified environmental factors cannot be ruled out, the involvement of genetic factor strongly suggested. Although genetic analysis could not be done in the present case but there is affair chance of PAX gene mutation responsible for CH as PAX mutation is autosomal dominant with incomplete penetrance.

CONCLUSION

Familial congenital hypothyroidism (CH) due to thyroid dysgenesis is a rare though an important cause of permanent CH. Proper genetic counseling and universal screening for CH can prevent devastating later consequences.

****

Author Contributions

Abhinav Kumar Gupta – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Syed Mohd. Razi – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Deepak Chand Gupta – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Saqib Ahmad Khan – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Pankaj Jain – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Keshav Kumar Gupta – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Rastogi MV, LaFranchi SH. Congenital hypothyroidism. Orphanet J Rare Dis 2010 Jun 10;5:17.
2. Castanet M, Polak M, Bonaïti-Pellié C, Nineteen years of national screening for congenital hypothyroidism: Familial cases with thyroid dysgenesis suggest the involvement of genetic factors. J Clin Endocrinol Metab 2001 May;86(5):2009–14.
3. Castanet M, Marinovic D, Polak M, Léger J. Epidemiology of thyroid dysgenesis: The familial component. Horm Res Paediatr 2010;73(4):231–7.
4. Ainger LE, Kelley VC. Familial athyreotic cretinism: Report of 3 cases. J Clin Endocrinol Metab 1955 Apr;15(4):469–75.
5. Cross HE, Hollander CS, Rimoin DL, McKusick VA. Familial agoitrous cretinism accompanied by musclar hypertrophy. Pediatrics 1968 Feb;41(2):413–20.
6. Kaplan M, Kauli R, Raviv U, Lubin E, Laron Z. Hypothyroidism due to ectopy in siblings. Am J Dis Child 1977 Nov;131(11):1264–5.
7. Yano K, Itoh Y, Inyaku F, Taguchi T, Takimoto M, Okuno A. Athyreotic congenital hypothyroidism in two sisters. [Article in Japanese]. Nihon Naibunpi Gakkai Zasshi 1992 Nov 20;68(11):1197–204.
8. Castanet M, Lyonnet S, Bonaïti-Pellié C, Polak M, Czernichow P, Léger J. Familial forms of thyroid dysgenesis among infants with congenital hypothyroidism. N Engl J Med 2000 Aug 10;343(6):441–2.
9. Grasberger H, Ringkananont U, Lefrancois P, Abramowicz M, Vassart G, Refetoff S. Thyroid transcription factor 1 rescues PAX8/p300 synergism impaired by a natural PAX8 paired domain mutation with dominant negative activity. Mol Endocrinol 2005 Jul;19(7):1779–91.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Churg–Strauss syndrome (eosinophilic granulomatosis with polyangiitis): A case report

M. West, P. Kumar

ABSTRACT

Churg–Strauss syndrome (CSS) is a rare systemic vasculitis of the small and medium sized blood vessels. The triad of asthma, sinusitis and hypereosinophilia is characteristic of CSS. However, it can affect any organ system with predominance for the skin, respiratory, neurological, gastrointestinal and cardiovascular systems. The natural history of the condition has been described in three phases: prodromal, eosinophilic and vasculitic. Depending on the organ system affected and stage of the disease, the presentation of CSS can be varied and the diagnosis can be challenging. The most frequently used criteria for diagnosing CSS are those developed by the American College of Rheumatology in 1990. Such classification criteria can assist in making the diagnosis of CSS and differentiating the condition from other diseases that cause pulmonary infiltrates with eosinophilia including allergic bronchopulmonary aspergillosis, acute and chronic eosinophilic pneumonia, idiopathic hypereosinophilic syndrome, certain parasitic infections and drug reactions. We present a case that is characterized by hypereosinophilia, vasculitis involving mainly the pulmonary and nervous systems, with a history of allergic rhinitis and sinusitis. Early diagnosis is crucial so that systemic glucocorticoids (the mainstay of treatment) can be commenced and can help to prevent organ damage and mortality.

Keywords: Churg–Strauss syndrome, Eosinophilic granulomatosis polyangiitis, Pulmonary infiltrates

M. West[1], P. Kumar[1]

Affiliation: [1]Department of Medicine, Mackay Base Hospital, Australia.

Corresponding Author: Pranav Kumar, Department of Medicine, Mackay Base Hospital, Australia; Email: drpranavkumar@gmail.com

Received: 12 May 2017
Accepted: 05 June 2017
Published: 01 August 2017

How to cite this article

West M, Kumar P. Churg–Strauss syndrome (eosinophilic granulomatosis with polyangiitis): A case report. Int J Case Rep Images 2017;8(8):555–560.

Article ID: Z01201708CR10821MW

****

doi:10.5348/ijcri-201782-CR-10821

INTRODUCTION

Eosinophilic granulomatosis with polyangiitis (EGPA), or more commonly known as Churg-Strauss syndrome (CSS) is a necrotizing small and medium vessel vasculitis associated with blood and tissue hypereosinophilia and usually occurring in people with a history of asthma [1].

Churg–Strauss syndrome is a rare disease with the annual incidence ranging from 2.4–6.8/1 million in the general population. It has equal gender distribution, average affected age ranges from 40–60 years old and there is no ethnic or familial predisposition [2].

Pathologists Dr Jacob Churg and Dr Lotte Strauss first described this condition as a distinctive clinical entity in 1951 [3, 4]. Based on their pathological examinations and postmortem studies, they noted a syndrome of asthma, hypereosinophilia, necrotizing vasculitis and extravascular granulomas [3]. They identified it as related to, but distinct from Polyarteritis Nodosa (PAN), which they termed allergic granulomatosis and angiitis [3, 5, 6].

The pathogenesis of CSS is still unknown [2, 7]. It is believed to be allergic or immune mediated due to the presence of allergic symptoms, elevated IgE levels and immune complex mediation [7, 8] with 40–60% being antineutrophil cytoplasmic antibody (ANCA) positive (predominantly perinuclear ANCA with anti-myeloperoxidase specificity) [1].

Herein, we report on a case that fits the clinical criteria for diagnosis of Churg–Strauss syndrome, supported by classic laboratory findings (not always present) and imaging results.

CASE REPORT

A 52-year-old female was referred by her general practitioner to the respiratory outpatients clinic (Mackay Base Hospital, Australia) for the evaluation of progressive dyspnea and wheeze. She reported a three-month history of increasing shortness of breath, with persistent wheeze, chest tightness and a dry cough. She also reported paroxysmal nocturnal dyspnea, but denied orthopnea or peripheral pitting edema. The patient had been treated with bronchodilators and antibiotics with minimal relief.

On review of systems, the patient denied skin rashes, joint swelling, myalgias, arthralgias, sore throat, dysphagia, odynophagia, abdominal pain, and changes in bowel habits or urinary symptoms. She did report tingling, numbness and pain in her left arm. She also endorsed some weight loss of five kilograms over the last three months. However, there was no history of fever or night sweats.

The patient's past medical history was significant for allergic rhinitis and sinusitis for the last two years. There was no history of asthma. She was also recently diagnosed with Grade 2 cervical intramedullary astrocytoma. The tumor was not resectable, and so she was treated with radiotherapy and chemotherapy (temozolomide) with neurosurgical follow-up. She has also had a splenectomy in the past for idiopathic thrombocytopenic purpura. She suffered depression as well.

There was no family history of asthma or atopy. Her mother had cervical cancer and her brother had hypertension. The patient had previously worked as a nurse. There was no exposure to organic or mineral dust, fumes or pets. There was no history of travel. She was a lifelong non-smoker, and rarely consumed alcohol.

On examination, the patient had a pulse rate of 72 beats/min, blood pressure of 132/84 mmHg, saturations of 98% on room air and she was afebrile. On auscultation of the chest, there were vesicular breath sounds with a few bibasal crepitations and bilateral rhonchi throughout the lungs. Her heart sounds were dual with no murmurs. Her abdomen was soft and non-tender. Her calves were soft, non-tender and there was no pitting edema. No skin rashes were noted.

Initial blood tests (reference ranges in defined opacities in the left lower lobe parentheses) of the patient showed hemoglobin level of 12.0 (11.5–16.0 g/dl), a white blood cell count of 10.3 cm^3 (4–11 cm^3) with a raised eosinophil count of 3.3 (<0.60).

Electrolytes, urea, creatinine, liver function tests and coagulation profile of the patient were within normal ranges. The C-reactive protein (CRP) was 11 (<5), and erythrocyte sedimentation rate (ESR) was 21 (<19). Urine examination was normal.

The patient's routine chest X-ray was normal. Computed tomography (CT) scan of the chest revealed a focal ill-defined alveolar opacity in the peribronchial region of the left lower lobe apical segment and another smaller similar opacity in the left upper lobe centrally (Figure 1). Computed tomography scan of the sinuses (Figure 2) showed near complete opacification of the frontal and the visualized ethmoid sinuses with mild mucosal thickening throughout the maxillary, sphenoid, and ethmoid sinuses and hypertrophy of the turbinates.

Spirometry showed a moderate obstructive pattern with forced expiratory volume (FEV1) of 58% predicted, forced vital capacity (FVC) of 74% predicted, and FEV1/FVC of 79%. There was severe small airway obstruction with maximum midexpiratory flow (MMEF 75/25) of 30% predicted. There was gas trapping with residual volume (RV) of 142%. Gas diffusion was normal with diffusing capacity (DLCO) being 89% and transfer coefficient of carbon monoxide (KCO) being 85%.

A transthoracic echocardiogram showed normal left ventricular systolic function, ejection fraction of 65% and normal diastolic function. No valvular abnormalities.

Subsequent bloods showed serum IgE was markedly elevated at 1140. Serum precipitins against *Aspergillus fumigatus* were negative. Bloods were positive for perinuclear antineutrophil cytoplasmic antibodies (p-ANCA) at a level of 640 by indirect immunofluorescence. The ANCA subsets directed towards the proteins myeloperoxidase (MPO-ANCA) were also positive at 417. Bloods were negative for cytoplasmic antineutrophil cytoplasmic antibodies (c-ANCA) and also negative for the subset directed towards proteinase-3 (PR3-ANCA).

A bronchoscopy and bronchoalveolar lavage was done which showed normal respiratory flora and a cell count less than 10. No open lung biopsy was performed, as there were sufficient criteria for a clinical diagnosis of eosinophilic granulomatosis with polyangiitis or Churg–Strauss syndrome to be made.

The patient was commenced on prednisolone (50 mg/day) and this was associated with a marked improvement in her symptoms. She reported no dyspnea or wheeze, and the neuropathic pain and paresthesia in her left arm decreased substantially. The prednisolone dose was slowly tapered and is still ongoing. Her last computed tomography scan of the chest (performed after three months of treatment with corticosteroids) shows complete resolution of the previously visualized areas of ground glass opacities in the left lower lobe.

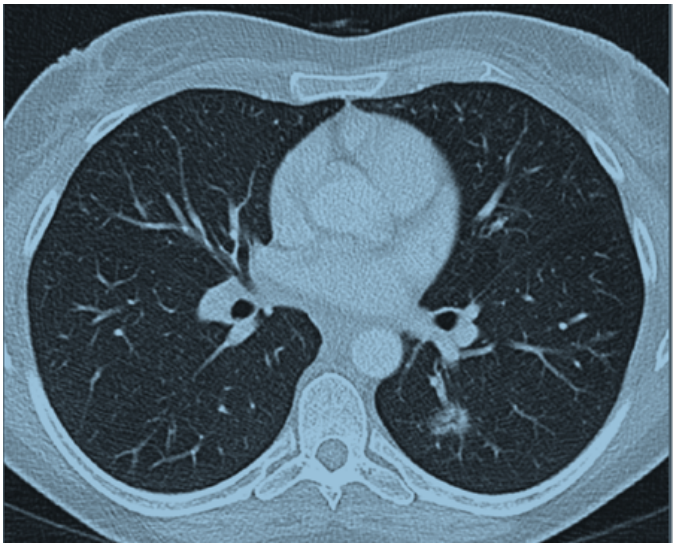


Figure 1: Computed tomography of the chest showing focal ill-defined opacities in the left lower lobe.

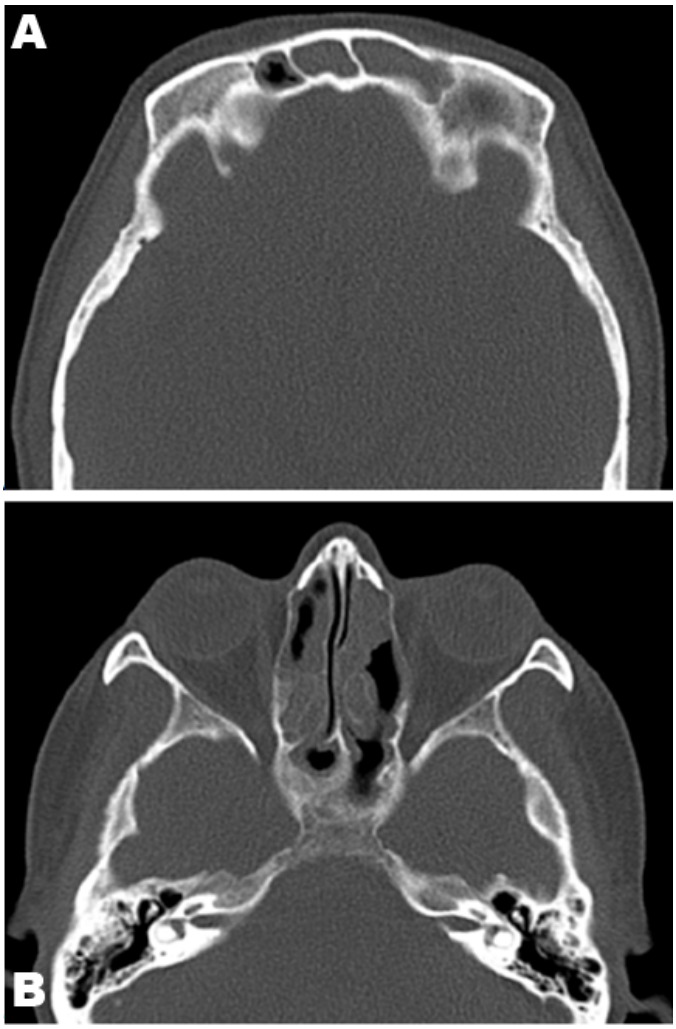


Figure 2: Computed tomography scan of the sinuses showing near complete opacification of the frontal and the visualized ethmoid sinuses.

DISCUSSION

The triad of asthma, sinusitis and hypereosinophilia is suggestive of Churg–Strauss syndrome [4]. Churg–Strauss syndrome however can be highly variable in its presentation and course and depending on the organ systems involved [6].

The natural history of the disease has been described in three phases [1]:

- **Prodromal phase**: It is characterized by atopic disease and may last for many years [1, 5]. Asthma is the cardinal feature of CSS, and is generally of late onset, chronic and severe, requiring multiple courses of systemic corticosteroids [1, 7]. However, upper airway conditions such as allergic rhinitis, nasal polyposis and sinusitis are also quite common and are known to affect 70% of patients [1, 9].
- **Eosinophilic phase**: It is characterized by hypereosinophilia in peripheral blood and eosinophilic infiltration of organs [1]. Eosinophils normally comprise less than 5% of leukocytes in the peripheral blood [10], however, may exceed 10% of the total leukocyte count in CSS [11]. Histological findings in CSS can include infiltrates rich in eosinophils in the walls of small and medium sized vessels and in surrounding tissues, as well as extravascular eosinophilic granulomas [2].
- **Vasculitic phase**: It is usually heralded with the onset of constitutional symptoms (fatigue, malaise, fever, weight loss, arthralgias) and the emergence of symptoms and possibly life-threatening sequelae of systemic vasculitis [1, 8].

As a systemic vasculitis, CSS can involve any organ system but predominantly involves skin, [1] respiratory, neurological, gastrointestinal and cardiovascular systems [4]. Poor prognosis is usually associated with gastrointestinal and cardiovascular involvement [1]. Common manifestations related to involvement of different organ systems are outlined below:

- **Skin:** Skin manifestations commonly include palpable purpura, as well as tender subcutaneous nodules, cutaneous infarctions, petechial rashes, urticarial lesions and livedo reticularis [2, 6, 8]. Skin involvement is present in half to two-thirds of patients [2].
- **Respiratory:** As mentioned, asthma is usually a cardinal feature of the disease [1] and often accompanied by rhinitis, sinusitis and nasal polyposis [2]. With a history of asthma, the presence of pulmonary infiltrates makes the diagnosis of CSS very likely, and this combination is present in two-thirds of patients. The infiltrates are classically patchy, lacking lobar and segmental distribution, and are transient, rapidly disappearing after corticosteroid treatment [2, 8].

- **Neurological:** Nervous system involvement may be present in up to three-quarters of patients and usually manifests as mononeuritis multiplex or peripheral polyneuropathy [2]. Less commonly there is central nervous system involvement (which can include cranial nerve palsies, hemorrhagic or ischemic strokes, seizures and psychiatric disturbance) [2].
- **Gastrointestinal:** Involvement of the gastrointestinal tract may affect up to half of all CSS patients. It can commonly cause abdominal pain, but may also cause bowel ischemia, perforation, obstruction, ulcerations, fistulas and hemorrhage [2]. It has also been associated with pancreatitis and acute acalculous cholecystitis [8].
- **Cardiovascular:** A wide range of cardiac complications related to CSS have been described including myocarditis, pericarditis, cardiomyopathy, arrhythmias, intraventricular thrombi, valvular abnormalities, coronary arteritis and sudden cardiac death. The frequency of cardiac involvement varies between 16–92%, with significant mortality if untreated [2].
- **Renal:** The Kidneys may be involved in 25–47% of patients with focal segmental glomerulonephritis that may lead to hematuria, proteinuria and hypertension, although rarely cause renal failure [2].

This case of CSS was characterized by hypereosinophilia, vasculitis involving mainly the pulmonary and nervous systems and with a history of allergic rhinitis and sinusitis. Interestingly, there was no history of asthma, which is considered the hallmark of CSS [12].

The diagnosis of CSS can be difficult especially given the variable presentation, and depending on the stage of CSS [1, 2]. The laboratory findings are non-specific and include eosinophilia, high IgE and raised acute phase reactants. The ANCA positivity (particularly perinuclear ANCA of anti-myeloperoxidase specificity) is found in approximately 40–60% of patients [1]. Again, this is not specific to CSS, and can be associated with microscopic polyangiitis (MPA), another one of the vasculitides [8].

In this case, the patient had peripheral eosinophilia (3.3), markedly elevated serum IgE (1140), and p-ANCA positive (640), specifically MPO-ANCA positive (417). Although these findings are not specific, it is useful in supporting the diagnosis in conjunction with the other clinical findings.

For clinical diagnosis of CSS, the diagnostic criteria published by the American College of Rheumatology (ACR) in 1990 are the most widely used [1, 11]. The criteria are: asthma, eosinophilia (greater than 10% on white blood cell differential count), paranasal sinusitis, pulmonary infiltrates (non-fixed), extravascular eosinophils on biopsy, and neuropathy (mononeuropathy including mononeuritis multiplex or polyneuropathy) [11]. At least four out of the six criteria are required for a confident diagnosis of CSS [1].

Our patient satisfied four of the six criteria with peripheral eosinophilia, paranasal sinusitis, mononeuropathy (with neuropathic pain, numbness and paresthesia in left arm), and pulmonary infiltrates. This makes the diagnosis of CSS with 85% sensitivity and 99.7% specificity [11].

The other classification systems include Lanham's criteria, which proposes a clinical diagnosis defined by the coexistence of asthma, blood eosinophilia (>1500 eosinophils/μl) and evidence of vasculitis in two or more extrapulmonary organs [2, 13]. However, as this involves at least two sites of vasculitic involvement, it can miss patients early in the course of the disease and delay treatment [2].

The Chapel Hill Consensus conference also established a new classification system, which defines CSS as: 'eosinophil-rich and granulomatous inflammation often involving the respiratory tract, and necrotizing vasculitis affecting small to medium-sized vessels associated with asthma and eosinophilia [14]. However, this definition has also been criticized as requires biopsy to establish diagnosis and lacks specificity in comparison to other classification systems [2].

The biopsy features proposed originally by Churg and Strauss [3] required the presence of:

i) necrotizing vasculitis of small and medium sized vessels
ii) eosinophilic infiltration
iii) extravascular granuloma formation

However, this triad hardly ever coexists at the same time in any one CSS patient, which can delay diagnosis [2]. Although easily accessible tissues should be biopsied, the clinical features of CSS are so distinctive that tissue biopsy is invariably not required [15]. In this case, the diagnosis had been made according to the clinical criteria published by the ACR and so biopsy was not required.

Churg–Strauss syndrome must be differentiated from other diseases that cause pulmonary infiltrates with eosinophilia including allergic bronchopulmonary aspergillosis, acute and chronic eosinophilic pneumonia, idiopathic hypereosinophilic syndrome, certain parasitic infections and drug reactions.

- Allergic bronchopulmonary aspergillosis (ABPA): The ABPA is a complex entity, resulting from a hypersensitivity response to persistent *Aspergillus fumigatus* in the airways [16]. The characteristic features are asthma, migratory pulmonary infiltrates, central bronchiectasis, elevated blood and alveolar eosinophils, high total IgE levels, positive immediate and late skin tests to the causative agent, and positive specific IgE and precipitins [17]. The absence of multisystem disease differentiates this condition from CSS [9].

- Idiopathic acute eosinophilic pneumonia (IAEP): The IAEP is associated with the rapid development of hypoxemic respiratory failure in previously well patients with no history of atopy, with diffuse bilateral air space or interstitial opacities on imaging. Peripheral blood eosinophils are usually normal, but a very high percentage of eosinophils (>25%) in bronchoalveolar lavage fluid is characteristic [18].
- Idiopathic chronic eosinophilic pneumonia (ICEP): The ICEP is characterized by a subacute or chronic presentation of cough, fever, weight loss and dyspnea [9, 17]. It occurs more commonly in women, and in those with a history of asthma [17]. There is alveolar and/or blood eosinophilia present in 90% of patients. It is differentiated from CSS by peripheral pulmonary infiltrates on imaging and an absence of multisystem involvement [9].
- Idiopathic hypereosinophilic syndrome (IHS): The IHS causes multisystem involvement (as does CSS) with signs and symptoms of organ damage related to eosinophilic infiltration [17]. However it can be distinguished from CSS by definition of blood eosinophilia greater than >1500/mm^3 for more than 6 months [9].
- Parasitic infections: Causative agents such as *Ascaris lumbricoides*, *Toxocara canis* and *Fasciola hepatica* are associated with blood and tissue eosinophilia and pulmonary infiltrates [17, 18].
- Drugs: Several drugs including non-steroidal anti-inflammatory agents, salicylates, and antibiotics can cause eosinophilic lung disease [17].

Early diagnosis and treatment is important for prevention of organ damage and mortality [1]. Systemic glucocorticoids are the mainstay of treatment [2]. However in a subset of patients, combined therapy with a cytotoxic agent (i.e. cyclophosphamide, azathioprine) may be required to control the disease [8]. Biologic agents such as rituximab, interferon-alpha, infliximab and etanercept are the focus of clinical trials for their potential role in CSS treatment in the future [2]. The prognosis of CSS is quite good in comparison to other systemic vasculitides, with a survival rate of 70% at five years [1].

CONCLUSION

In this case, the patient was commenced on high dose prednisolone with a subsequent improvement in symptoms, blood eosinophil count, inflammatory markers and resolution of pulmonary infiltrates on imaging. We will continue to slowly wean her prednisolone dose, with the possibility of commencing a steroid-sparing agent in the future if required.

****

Author Contributions

M. West – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

P. Kumar – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

REFERENCES

1. Manglunia A, Mathur V, Garg P, Jat RK, Saxena GN. Churg Strauss Syndrome: A Case Report. IOSR-JDMS 2015; 14(3):27–8.
2. Szczeklik W, Jakiela B, Adamek D, Musial J. Cutting edge issues in the Churg–Strauss syndrome. Clin Rev Allergy Immunol 2013 Feb;44(1):39–50.
3. Churg J, Strauss L. Allergic granulomatosis, allergic angiitis, and periarteritis nodosa. Am J Pathol 1951 Mar–Apr;27(2):277–301.
4. Gorska L, Chelminska M, Kuziemski K, et al. Analysis of safety, risk factors and pretreatment methods during rush hymenoptera venom immunotherapy. Int Arch Allergy Immunol 2008;147(3):241–5.
5. Lhote F, Cohen P, Généreau T, Gayraud M, Guillevin L. Microscopic polyangiitis: clinical aspects and treatment. Ann Med Interne (Paris) 1996;147(3):165–77.
6. Della Rossa A, Baldini C, Tavoni A, et al. Churg-Strauss syndrome: Clinical and serological features of 19 patients from a single Italian centre. Rheumatology (Oxford) 2002 Nov;41(11):1286–94.
7. Masjedi MR, Tafti SF, Cheraghvandi A, Fayazi N, Talischi F, Mokri B. Churg–Strauss syndrome following cessation of allergic desensitization vaccination: A case report. J Med Case Rep 2010 Jun 22;4:188.
8. Cottin V, Cordier JF. Churg–Strauss syndrome. Allergy 1999;54(6):535–51.

9. Mokri B, Tafti SF, Talischi F. Churg–Strauss syndrome with broad spectrum clinical presentations: A report of 3 cases. East Mediterr Health J 2011 Oct;17(10):798–800.
10. Fulkerson PC, Rothenberg ME. Targeting eosinophils in allergy, inflammation and beyond. Nat Rev Drug Discov 2013 Feb;12(2):117–29.
11. Masi AT, Hunder GG, Lie JT, et al. The American College of Rheumatology 1990 criteria for the classification of Churg–Strauss syndrome (allergic granulomatosis and angiitis). Arthritis Rheum 1990 Aug;33(8):1094–100.
12. Sharma BK, Daga MK, Sharma M. A limited form of Churg–Strauss syndrome presenting without asthma and eosinophilia. Med J Aust 2004 Nov 1;181(9):498-9.
13. Lanham JG, Elkon KB, Pusey CD, Hughes GR. Systemic vasculitis with asthma and eosinophilia: A clinical approach to the Churg–Strauss syndrome. Medicine (Baltimore) 1984 Mar;63(2):65–81.
14. Jennette JC, Falk RJ, Andrassy K, et al. Nomenclature of systemic vasculitides: Proposal of an international consensus conference. Arthritis Rheum 1994 Feb;37(2):187–92.
15. Leatherman JW. The lung in systemic vasculitis. Semin Respir Infect 1988 Sep;3(3):274–88.
16. Patterson K, Strek ME. Allergic bronchopulmonary aspergillosis. Proc Am Thorac Soc 2010 May;7(3):237–44.
17. Marchand E, Cordier JF. Idiopathic chronic eosinophilic pneumonia. Orphanet J Rare Dis. 2006 Apr 6;1:11.
18. Jeong YJ, Kim KI, Seo IJ, et al. Eosinophilic lung diseases: A clinical, radiologic, and pathologic overview. Radiographics 2007 May–Jun;27(3):617–37; discussion 637–9.

CLINICAL IMAGE PEER REVIEWED | OPEN ACCESS

Acquired cystic kidney disease: A benign yet potentially fatal condition

Vikrampal Bhatti, Abhilash Koratala, Ashutosh M. Shukla

CASE REPORT

A 48-year-old African-American male with end stage renal disease (ESRD) on hemodialysis for 14 years developed acute abdominal pain during his outpatient dialysis session and was sent to the emergency room. The patient was found to be hypotensive with a blood pressure of 93/72 mmHg requiring fluid resuscitation, which later deteriorated further requiring vasopressors. He was found to have acute anemia with a hemoglobin level of 7.5 g/dL. Abdominal computed tomography (CT) scan revealed large hemoperitoneum with a right perinephric sentinel clot, bilateral multiple renal cysts and features suspicious of bleeding originating from the upper anterior pole of right kidney (Figure 1). The largest of the cysts measured ~3.2 cm in diameter. Renal angiogram showed active ongoing extravasation from multiple branches of the right renal artery (Figure 2). Selective right renal artery embolization was unsuccessful. An emergent laparotomy revealed a torn right renal capsule with subcapsular hemorrhage and a 1.4 cm ragged defect in the inferior portion of the kidney. He underwent right nephrectomy and pathology was negative for malignancy. It essentially showed end stage kidney with arteriolonephrosclerosis, tubular atrophy, interstitial fibrosis, thyroidization, cysts and chronic interstitial inflammation with associated intra-parenchymal and perirenal hemorrhage. Patient refused blood transfusion because of religious reasons and unfortunately died of hemorrhagic shock after the surgery.

Vikrampal Bhatti[1], Abhilash Koratala[1], Ashutosh M. Shukla[1]

Affiliation: [1]University of Florida, Gainesville, USA.

Corresponding Author: Abhilash Koratala, MD, Division of Nephrology, Hypertension and Renal Transplantation, P.O. Box 100224, Gainesville, FL 32610, USA; Email: akoratsla@ufl.edu

Received: 01 March 2017
Accepted: 13 July 2017
Published: 01 August 2017

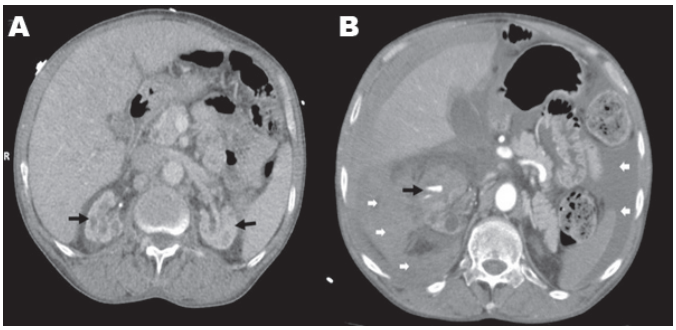


Figure 1: (A) Abdominal computed tomography scan one year prior to presentation showing bilateral renal cysts (arrows) (B) Abdominal computed tomography scan with contrast demonstrating large hemoperitoneum (white arrows) and features suggestive of bleeding originating from the upper anterior pole of right kidney, with extravasation of contrast (black arrow).

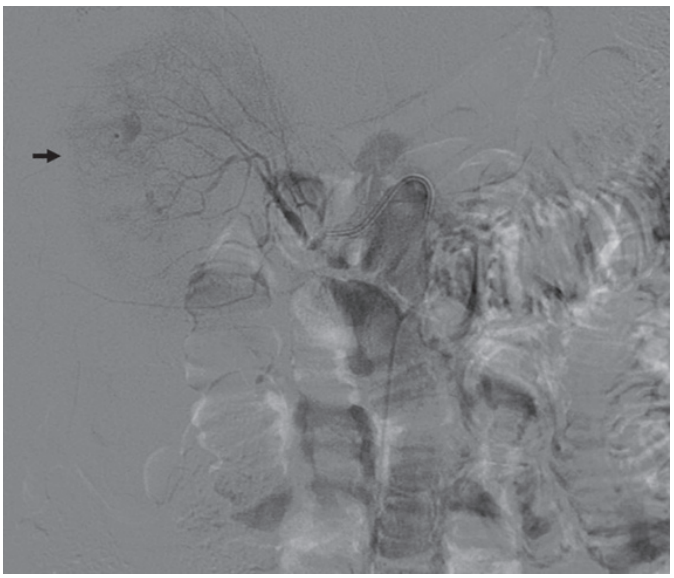


Figure 2: Renal angiogram demonstrating the bleeding vessels inside the right kidney.

DISCUSSION

Acquired cystic kidney disease (ACKD) in patients with advanced chronic kidney disease (CKD) and end

stage renal disease, unlike other forms of cystic renal diseases, is largely considered a benign pathology with no clear recommendations for follow-up care. Our case represents a rare but fatal complication of ACKD. Definition of ACKD usually requires three or more cysts in each kidney in a patient with CKD or end stage renal disease who does not have a hereditary cause of cystic disease such as autosomal-dominant polycystic kidney disease or tuberous sclerosis [1]. It is estimated that hemorrhagic cysts are seen in approximately 50% of the patients with ACKD and perinephric hematomas in about 13% of patients [2], which can generally be managed conservatively. Severe bleeding can be fatal and may require interventions such as transcatheter embolization or surgery. More studies are needed to determine the benefit of periodic screening in patients with ACKD to prevent potentially fatal complications.

Without clear guidelines for follow-up and majority of the studies on cystic renal diseases excluding patients with ACKD, the true incidence of complications in these patients is not known. We believe an observational cohort study on the lines of Bosniak classification, in patients with advanced CKD and end stage renal disease should be considered such that natural history of this condition, including potential complications e.g. malignancy [3] and bleeding can be better predicted.

CONCLUSION

Acquired cystic kidney disease (ACKD) is characterized by development of numerous cysts in both the kidneys in individuals without history of hereditary cystic disease. Hemorrhagic cysts can sometimes lead to severe bleeding that can be life-threatening. High index of suspicion is required for this condition when end stage renal disease patients present with abdominal pain and drop in hemoglobin. Until clear guidelines are established, it would be prudent to monitor ACKD patients with periodic renal imaging.

****

Keywords: Acquired cystic kidney disease, Hemoperitoneum, Renal disease

How to cite this article

Bhatti V, Koratala A, Shukla AM. Acquired cystic kidney disease: A benign yet potentially fatal condition. Int J Case Rep Images 2017;8(8):561–563.

Article ID: Z01201708CL10129VB

****

doi:10.5348/ijcri-201719-CL-10129

****

Author Contributions

Vikrampal Bhatti – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Abhilash Koratala – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Ashutosh M. Shukla – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Levine E. Acquired cystic kidney disease. Radiol Clin North Am 1996 Sep;34(5):947–64.
2. Choyke PL. Acquired cystic kidney disease. Eur Radiol 2000;10(11):1716–21.
3. Bhatti V, Vangapalli A, Bhattacharya D, Koratala A. Renal cell carcinoma of the native kidney in a renal transplant recipient. Int J Case Rep Images 2017;8(2):165–7.

CLINICAL IMAGE PEER REVIEWED | OPEN ACCESS

Successful treatment of pneumomediastinum in a patient with interstitial lung disease due to anti-synthetase syndrome: A case report

Roy Cho, Erhan H. Dincer, Rade Tomic, Hyun Kim

CASE REPORT

A 35-year-old female with interstitial lung disease due to anti-synthetase syndrome presented with cough, hoarseness and facial swelling. Chest computed tomography scan demonstrated significant pneumomediastinum (Figure 1). Bronchoscopy and esophagram were unremarkable. We began high-concentration oxygen (10 L/min with non-rebreather face mask for 12-hours/day), withheld mycophenolate mofetil, reduced prednisone and began dextromethorphan. After one-month, there was complete resolution of her symptoms and pneumomediastinum.

DISCUSSION

Pneumomediastinum (PNM) is a rare complication of anti-synthetase syndrome with only three cases reported since 1986 [1–3]. The associated one-month mortality is 25%, which highlights the need for effective management [4]. The management of PNM in interstitial lung disease is based on case reports and experience forming expert opinion. However, the best management requires understanding the pathogenesis [5]. Several mechanisms of PNM in interstitial lung disease have been proposed including the Macklin effect, worsening vasculitis leading to airway rupture, and weakened alveolar walls from immunosuppression [6]. In this case, reducing immunosuppression suggested that weakened alveolar walls was a factor and using high-concentration oxygen and cough suppressant decidedly contributed to the full recovery. Notably, this is the second case in literature that has reported success using high-concentration oxygen for the treatment of PNM [7].

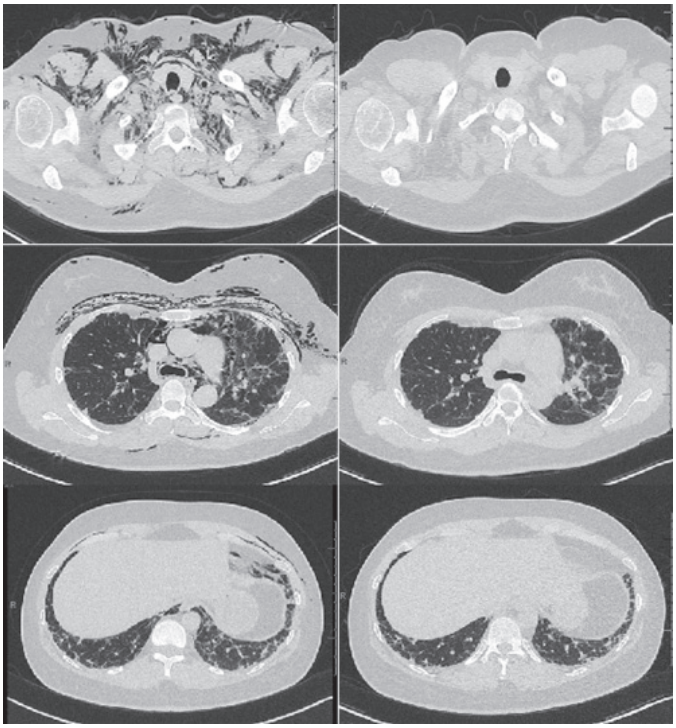


Figure 1: Chest computed tomography (CT) scan at the level of the neck, heart and lung base demonstrating subcutaneous air and pneumomediastinum (left). After one-month of high-concentration oxygen, anti-tussives and reduction in immunosuppression; there was complete resolution (right).

CONCLUSION

We report a case of pneumomediastinum in a patient with anti-synthetase syndrome who was successfully

Roy Cho[1], Erhan H. Dincer[2], Rade Tomic[2], Hyun Kim[2]

Affiliations: [1]Assistant Professor, University of Minnesota, Department of Pulmonary, Allergy, Critical Care and Sleep Medicine; [2]Associate Professor, University of Minnesota, Department of Pulmonary, Allergy, Critical Care and Sleep Medicine.

Corresponding Author: Roy Cho, MD, Assistant Professor, University of Minnesota, Department of Pulmonary, Allergy, Critical Care and Sleep Medicine; Email: choxx548@umn.edu

Received: 14 April 2017
Accepted: 13 July 2017
Published: 01 August 2017

treated with reduction in immunosuppression, high-concentration oxygen, and cough suppression.

****

Keywords: Anti-synthetase syndrome, Dermatomyositis, Interstitial lung disease, Pneumomediastinum

How to cite this article

Cho R, Dincer EH, Tomic R, Kim H. Successful treatment of pneumomediastinum in a patient with interstitial lung disease due to anti-synthetase syndrome: A case report. Int J Case Rep Images 2017;8(8):564–566.

Article ID: Z01201708CL10130RC

****

doi:10.5348/ijcri-201720-CL-10130

****

Author Contributions

Roy Cho – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Erhan H. Dincer – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Rade Tomic – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Hyun Kim – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Bradley JD. Spontaneous pneumomediastinum in adult dermatomyositis. Ann Rheum Dis 1986 Sep;45(9):780–2.
2. Carmody E, McNicholl J, Chadwick G, Bresnihan B, Fitzgerald MX. Prolonged spontaneous pneumomediastinum in adult dermatomyositis. Ann Rheum Dis 1987 Jul;46(7):566.
3. Cicuttini FM, Fraser KJ. Recurrent pneumomediastinum in adult dermatomyositis. J Rheumatol 1989 Mar;16(3):384–6.
4. Ma X, Chen Z, Hu W, et al. Clinical and serological features of patients with dermatomyositis complicated by spontaneous pneumomediastinum. Clin Rheumatol 2016 Feb;35(2):489–93.
5. Tang R, Millett CR, Green JJ. Amyopathic dermatomyositis complicated by pneumomediastinum. J Clin Aesthet Dermatol 2013 Mar;6(3):40–3.
6. Macklin MT, Macklin CC. Malignant interstitial emphysema of the lungs and mediastinum as an important occult complication in many respiratory diseases and other conditions: An interpretation of the clinical literature in the light of laboratory experiment. Medicine 1944;23(4):281–358.
7. Patel A, Kesler B, Wise RA. Persistent pneumomediastinum in interstitial fibrosis associated with rheumatoid arthritis: Treatment with high-concentration oxygen. Chest 2000 Jun;117(6):1809–13.