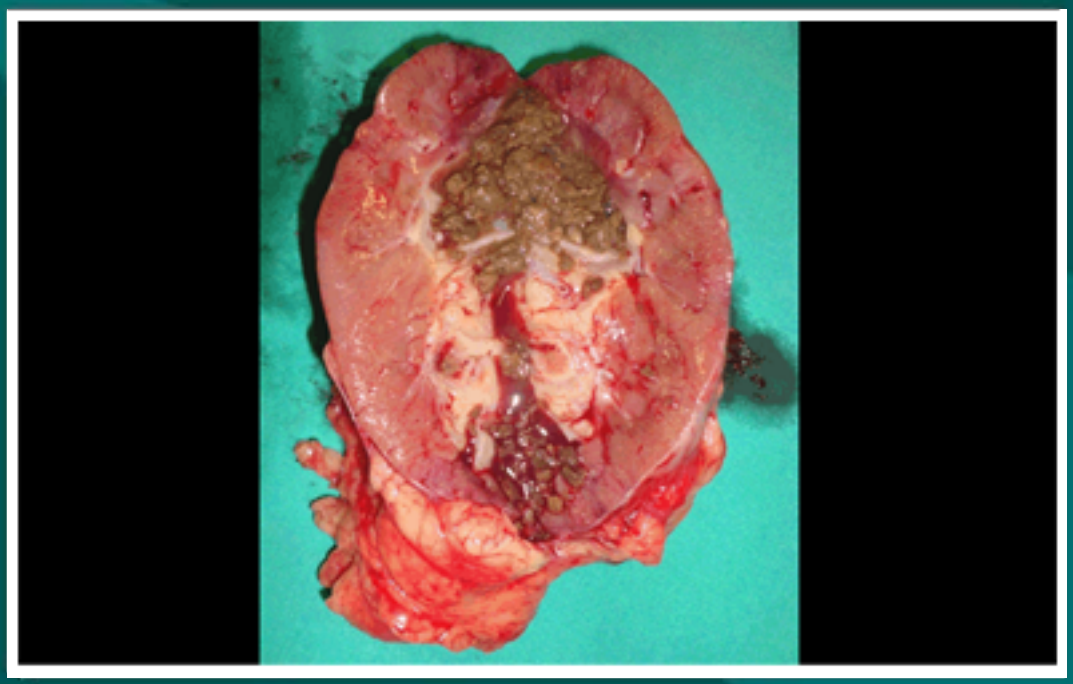
March 2015; Volume 6; Number 3

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

EDITORIAL BOARD

International Journal of Case Reports and Images (IJCRI)

Contact Details:

Editorial Office

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

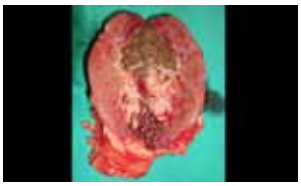
Guidelines for Authors

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 Figure:



All Articles:

Open Access

Contents

Vol. 6, No. 3 (March 2015)

Cover Image

Case Report

Clinical Images

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

CASE REPORT PEER REVIEWED | OPEN ACCESS

Osteochondroma of the mandibular condyle: A case report

Jayakumar K., Soumithran C.S., Manoj Joseph Michael, Pallav Kumar Kinra, Ambadas Kulkarni, Tushar Lamsoge

ABSTRACT

Introduction: Osteochondromas or osteocartilaginous exostoses are the most common benign tumors of the bones. It is characterized as a type of overgrowth that can occur in any bone where cartilage forms bone. It is uncommon in this part of body because of intramembranous origin of craniofacial bones. Osteochondromas do not result from any injury and the exact cause remains unknown. Recent research has indicated that multiple osteochondromas is an autosomal dominant inherited disease. The treatment choice for osteochondroma is surgical removal of solitary lesion or partial excision of the outgrowth when symptoms cause motion limitations or nerve and blood vessel impingements. Osteochondroma of the mandibular condyle is extremely rare. Case Report: A 45-year-old female presented to our department with diffuse swelling in her left side of face and pain in her left ear while opening the mouth since last six months. Clinically, mouth opening was limited with deviation of mandible towards right side while opening mouth. There was unilateral posterior crossbite on the right side. Protrusive movement and lateral excursions of mandible were restricted. The lesion appeared to be benign bony lesion and complete surgical excision of the whole tumor mass along with condylectomy was performed under general anesthesia. Conclusion: As osteochondroma is a benign neoplasm, various treatment modalities include resection of tumor along with condylectomy, condylectomy with reconstruction of the resected condyle if indicated or selected tumor removal without condylectomy. The prognosis of osteochondroma is usually excellent after adequate excision. This case showed no recurrence after the treatment. Malignant transformation of the lesion is exceedingly rare.

Keywords: Condyle, Mandible, Osteochondroma, Temporomandibular joint

Jayakumar K.[1], Soumithran C.S.[2], Manoj Joseph Michael1, Pallav Kumar Kinra[3], Ambadas Kulkarni[4], Tushar Lamsoge[5]

Affiliations: [1]Associate Professor, Department of Oral and Maxillofacial Surgery, Govt. Dental College, Kozhikode, Kerala, India; [2]Professor and HOD, Department of Oral and Maxillofacial Surgery, Govt. Dental College, Kozhikode, Kerala, India; [3]Junior Resident, Department of Oral and Maxillofacial Surgery, Govt. Dental College, Kozhikode, Kerala, India; [4]Senior Resident, Department of Oral and Maxillofacial Surgery, Govt. Dental College, Kozhikode, Kerala, India; [5]Senior Resident, Department of Oral and Maxillofacial Surgery, Govt. Dental College, Alleppey, Kerala, India.

Corresponding Author: Dr. Pallav Kumar Kinra, Junior Resident, Department of Oral and Maxillofacial Surgery, Govt. Dental College, Kozhikode, Kerala, India 673008; Ph: 918943560527; Email: pallav_kinra163@yahoo.com

Received: 08 October 2014
Accepted: 19 November 2014
Published: 01 March 2015

How to cite this article

Jayakumar K, Soumithran CS, Michael MJ, Kinra PK, Kulkarni A, Lamsoge T. Osteochondroma of the mandibular condyle: A case report. Int J Case Rep Images 2015;6(3):127–131.

doi:10.5348/ijcri-201523-CR-10484

INTRODUCTION

Osteochondroma, also known as osteocartilaginous exostosis is a benign, cartilage capped osseous lesion that projects from the surface of the bone, usually near its growth center [1–4]. The affected bone may be abnormally wide and somewhat deformed at the level of the lesion.

The appearance is similar to that of an epiphyseal plate before closure. Although osteochondroma is predominantly an osseous lesion, still it is considered as one of the cartilaginous tumors because bony mass is produced by progressive endochondral ossification of its growing cartilaginous cap. Osteochondroma is one of the most common benign tumors of the axial skeleton [4–8] representing 35–50% of all benign bone tumors and 8–15% of all primary bone tumors [4, 7]. Only 1% of these occur in head and neck region. In 2014, Erdem et al. revealed literature and mentioned that only 72 cases of osteochondroma of mandibular condyle has been reported till now.

It is rarely found in the facial bones, because most of the craniomaxillofacial bones develop by intramembranous ossification [5–8]. When present, the tumor is most often reported to affect the mandibular coronoid process as it is of the embryonic cartilaginous origin [6]. The other sites where osteochondroma have been reported are at the skull base, in the posterior maxilla, maxillary sinus, ramus, body and symphyseal region of the mandible [7–10]. Osteochondroma of the mandibular condyle is extremely rare [9]. Osteochondroma of the mandibular condyle is a slow growing lesion. Therefore, symptoms may develop over a long period. These symptoms include occlusal disturbances, facial asymmetry restricted mandibular movements, pain with varying intensity, clicking, popping, and crepitation of the affected joint and changes in the condylar morphology. The treatment of a condylar osteochondroma involves primarily resection and in large lesions where functional or cosmetic deformity results, immediate reconstruction is indicated.

CASE REPORT

A 45-year-old female presented to our department with diffuse swelling in her left side of face and pain in her left ear while opening the mouth since last six months (Figure 1). She gave a history of slowly progressing facial asymmetry with difficulty in chewing. There was no significant family history, no one in her family had the same problem earlier. On clinical examination, deviation of mandible was present towards right side and mouth opening was limited to 20 mm (Figure 2). There was a unilateral posterior crossbite on the right side. Protrusive movement and lateral excursions of mandible were restricted. On palpation there as a firm swelling over the left side of the face in front of the tragus extending along the ramus of mandible towards temporal area crossing the zygomatic arch. There was tenderness over masseter muscle and at the lateral and dorsal aspects of the left condyle. On palpation of cervical lymph nodes no enlargement was present and were normal in consistency. Panoramic radiographic examination showed pedunculated lesion originating from anterior part of left condyle. On the coronal and axial CT images, it was clearly distinguished that there was a cartilaginous or bony lesion attached to the anteromedial surface of the left condyle and extending medially to involve left parapharyngeal space and masticatory space displacing the lateral pterygoid muscle anteriorly. Superiorly the lesion is causing thinning and erosion of greater wing of sphenoid (Figure 3).

The surgery was performed under general anesthesia. Preauricular incision was given and extended from the superior portion of helix to the inferior portion of the

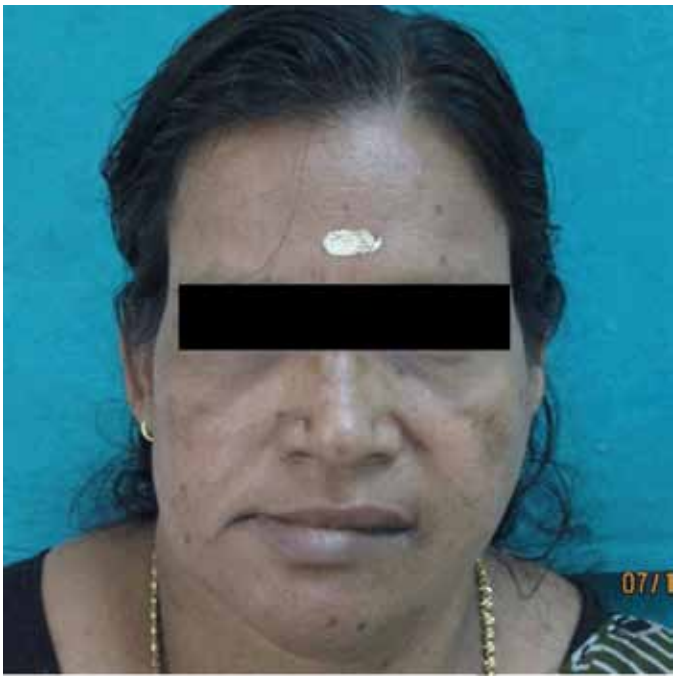


Figure 1: Preoperative picture of patient with deviated mandible towards right side.

Figure 2: Clinical picture with limited mouth opening.

ear lobe. After the skin incision was done, the underlying subcutaneous tissue, temporal fascia and muscle were carefully dissected. In the temporal region the incision was up to the superficial layer of the temporalis fascia. At the root of the zygomatic arch, the superficial layer of temporalis fascia was incised anterosuperiorly. The periosteum was then elevated to expose the zygomatic arch and then subperiosteal dissection was carried further downwards to expose temporomandibular joint region. After getting a wide exposure of the area zygomatic arch was sectioned; leaving it attached to the masseter muscle, and was put aside. Excision of the whole tumor mass was performed on the table with condylectomy (Figure 4). Zygomatic arch was placed back and rigid fixation was done with interosseous wires.

On histological examination, an outer lining composed of a broad layer of partially loose periosteal collagen tissue was found, attached by small amounts of cartilaginous differentiated tissue. Adjacent cancellous bone with trabeculae of variable size and surrounded by cartilaginous tissue was visible. Chondrocytes of the cartilaginous cap were arranged in clusters parallel to lacunar spaces. On the basis of histological findings definitive diagnosis of osteochondroma was established (Figure 5A–B). Postoperative course was uneventful. Follow-up at second month, the patient's maximum mouth opening had increased to 35 mm. On two years follow- up patient is asymptomatic and there is no deviation of mandible towards opposite side (Figure 6). Till now there is no recurrence reported.

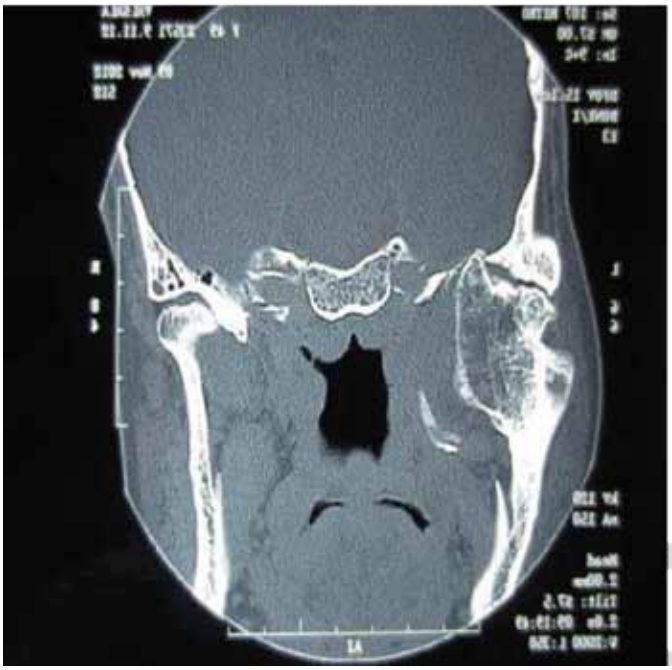


Figure 3: Computed tomography scan image of the lesion involving left condyle.

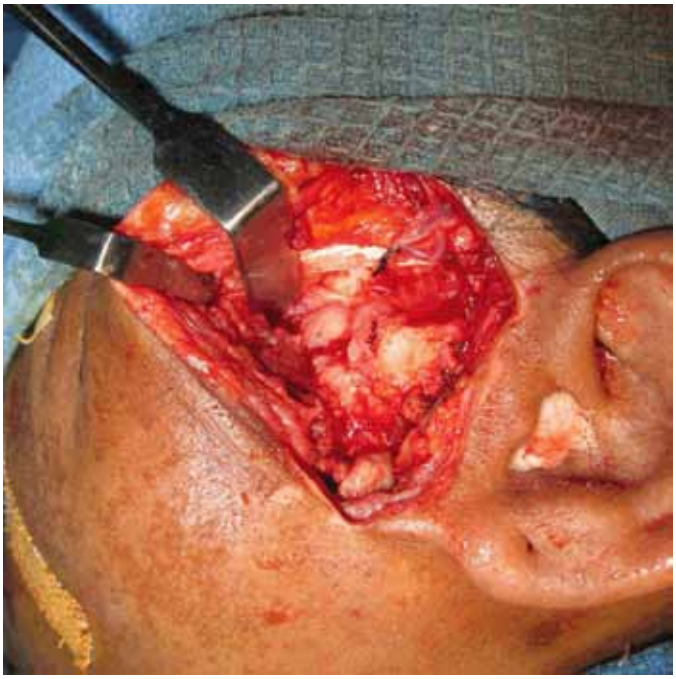


Figure 4: Intraoperative view of lesion being excised.

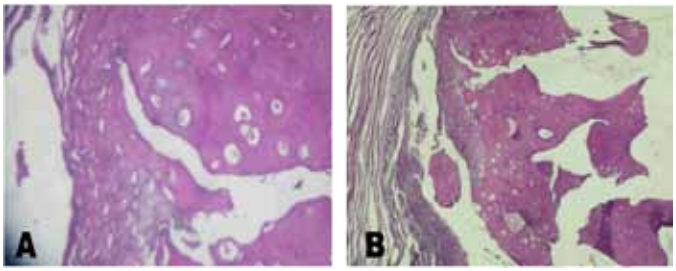


Figure 5: (A, B) Histological view confirming the diagnosis.

Figure 6: Postoperative, after two years, view of patient with improved mouth opening.

DISCUSSION

Cartilage-capped tumor like, exophytic growths of bone, termed osteochondroma, has been subjected to many debates regarding their origin and have been considered as developmental malformations, hyperplasia, or neoplastic disorders. Langenskiold postulated that osteochondroma occurs when limited portions of the undifferentiated cell layer of the growth cartilage are displaced peripherally towards the metaphysis [11]. Lichtenstein's theory favored a neoplastic origin, but did not attribute it to the growth cartilage [12]. He suggested that periosteum had a potential to form chondroblasts and osteoblasts, and a perverted activity of the periosteum to form metaplastic cartilage may give rise to osteochondromas. A relatively high frequency of osteochondromas around the temporomandibular joint can be easily explained embryologically when it is considered that the region from the mandibular lingula to the anterior process of the malleus is derived from the part of Meckel's cartilage not replaced by mandibular bone and that remnants of this embryonic tissue may still persist and give rise to tumor growth [13]. This theory is also applicable to the occurrence of osteochondromas or other chondrogenic tumors in the tongue, where remnants of brachial arch cartilage may potentially persist [14].

Trauma and inflammation have been specifically implicated either as initiating or as predisposing factors for mandibular condyle osteochondromas". Porter and Simpson suggested that a genetic component might also be involved in the neoplastic pathogenesis due to somatic mutations found in chromosomes 8 and 11. Osteochondromas are frequently seen in 2nd and 3rd decades of life. They are more common in men, with a male to female predilection of 1.6 to 1. The clinical findings associated with osteochondromas of the mandibular condyle usually develop over the course of several months to years. Patients most commonly present with the facial asymmetry, disturbed occlusion, posterior apertognathia (open bite) on the affected side, crossbite on the unaffected side, palpable painless temporomandibular area mass, together with limitation of mouth opening and mandibular movement. The differential diagnosis includes osteoma, chondroma, condylar hyperplasia, giant cell tumor, myxoma, fibro-osteoma, fibrous dysplasia, fibrosarcoma, chondrosarcoma and metastatic disease. Several methods have been suggested for the treatment of condylar osteochondromas. These include resection of tumor along with condylectomy, condylectomy with reconstruction if indicated or selected tumor removal without condylectomy. By providing extra space and exposure, condylectomy enables easier and safer removal of the lesion when the medially located vascular structures (for example, internal and external maxillary arteries) are concerned. Approximately, 75% of the patients with osteochondroma develop solitary lesions and 25% have multiple lesions. The solitary lesions develop sarcomatous changes in approximately 1% of the cases. However, all reported condylar osteochondromas have been histologically benign and malignant transformation has not been observed. The general recurrence rate of osteochondromas is approximately 2% and there is only one recurrence of condylar osteochondroma reported in literature, which occurred one year after its excision in multiple pieces [15].

CONCLUSION

As osteochondroma of mandibular condyle is extremely rare and benign neoplasm, patients with this tumor present mandibular movement deviation and alterations in dental occlusion, with a slow and asymptomatic growth of the lesion. Various treatment modalities include resection along with condylectomy, condylectomy with reconstruction of the resected part if indicated or selected tumor removal without condylectomy. The prognosis of osteochondroma is usually excellent after adequate excision. Imaging techniques are the valuable aid for accurately diagnosing neoplasm like condylar osteochondroma. Diagnosis is only confirmed by histopathological examination. Even though the recurrence of the tumor is rare, it is always better to follow-up the patient for the recurrence risk.

****

Acknowledgements

We are thankful to Dr. Akhilesh, Dr. Shalini, Dr. Bindudas, Dr. Jibin, Dr. Ikram, Dr. Seeja P. Chandran, Dr. Pravish, Dr. Aruldev DP, and Dr. Manu Mathew for their help and support in preparing this case report.

Author Contributions

Jayakumar K. – 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

Soumithran C.S. – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Manoj Joseph Michael – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Pallav Kumar Kinra – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Ambadas Kulkarni – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Tushar Lamsoge – Substantial contributions to conception

and design, 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. Miyawaki T, Kobayashi M, Takeishi M, Uchida M, Kurihara K. Osteochondroma of the Mandibular Body. Plast Reconstr Surg 2000 Apr;105(4):1426–8.
2. Muñoz M, Goizueta C, Gil-Díez JL, Díaz FJ. Osteocartilaginous exostosis of the mandibular condyle misdiagnosed as temporomandibular dysfunction. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1998 May;85(5):494–5.
3. Aydin MA, Kucukcelebi A, Sayilkan S, Celebioglu S. Osteochondroma of the Mandibular Condyle: Report of 2 cases treated with Conservative Surgery. J Oral Maxillofac Surg 2001 Sep;59(9):1082–9.
4. Vezeau PJ, Fridrich KL, Vincent SD. Osteochondroma of the Mandibular Condyle: Literature Review and Report of Two Atypical Cases. J Oral Maxillofac Surg 1995 Aug;53(8):954–63.
5. Gingrass DJ, Sadeghi EM. Osteochondroma of the Mandibular Condyle Mimicking TMJ syndrome: Clinical and Therapeutic Appraisal of a Case. J Orofac Pain 1993 Spring;7(2):214–9.
6. Goyal M, Sidhu SS. A massive osteochondroma of the mandibular condyle. Br J Oral Maxillofac Surg 1992 Feb;30(1):66–8.
7. Karras SC, Wolford LM, Cottrell DA. Concurrent Osteochondroma of the Mandibular Condyle and Ipsilateral Cranial Base resulting In Temporomandibular Joint Ankylosis: Report of a Case and Review of the Literature. J Oral Maxillofac Surg 1996 May;54(5):640–6.
8. Koole R, Steenks MH, Witkamp TD, Slootweg PJ, Shaefer J. Osteochondroma of the mandibular condyle. A case report. Int J Oral Maxillofac Surg 1996 Jun;25(3):203–5.
9. Gaines RE Jr, Lee MB, Crocker DJ. Osteochondroma of the Mandibular Condyle: Case Report and Review of the Literature. J Oral Maxillofac Surg 1992 Aug;50(8):899–3.
10. Kerscher A, Piette E, Tideman H, Wu PC. Osteochondroma of the coronoid process of the mandible - Report of a case and review of literature. Oral Surg Oral Med Oral Pathol 1993 May;75(5):559–64.
11. Langenskiold A. The development of multiple cartilaginous exostoses. Acta Orthop Scand 1967;38:259.
12. Lichtenstein L. Bone Tumors (Ed 5). St Louis, MO, Mosby 1977.
13. Kermer C, Rasse M, Undt G, Lang S. Cartilaginous exostoses of the mandible. Int J Oral Maxillofac Surg 1996 Oct;25(5):373–5.
14. Watson C, Crother JA, Stephen MR. Osteochondroma of the tongue. J Laryngol Otol 1990 Feb;104(2):138–40.
15. Iizuka T, Schroth G, Laeng RH, Lädrach K. Osteochondroma of the mandibular Condyle: Report of a Case. J Oral Maxillofac Surg 1996 Apr;54(4):495–1.

CASE REPORT PEER REVIEWED | OPEN ACCESS

A rare case of maxillary sinus osteomyelitis with intraorbital, extraconal abscess in a term low birth weight twin

Janampally Ravikiran, Swathi Chacham, Uppin Narayan Reddy, Jillalla Narsing Rao, Ekta Aggarwal, Anumula Soumya, Nori Madhavi

ABSTRACT

Introduction: Acute osteomyelitis in neonates is a rare infectious disorder of bone leading to diagnostic and therapeutic challenge, more so in neonates. While the long bones are frequently affected sites, maxilla is rarely involved. Physiological immaturity of the immune system coupled with invasive diagnostic and therapeutic procedures render them susceptible for this condition, more so in premature, low birth weight infants. The most common organism causing osteomyelitis in neonates is *Staphylococcus aureus*. Lesion of the orbit as well as contiguous faciomaxillary and sinonasal inflammatory pathology often lead to proptosis in neonates. Case Report: We report a 15-day-old term female neonate (twin 1) presenting with non-traumatic periorbital swelling and purulent nasal discharge. The neonate also had failure to thrive. On clinical examination, the neonate was sick with sclerema and shock. There was right eye proptosis with edema and erythema. Initial diagnosis of periorbital abscess or orbital tumor was considered and the infant was investigated. Sepsis screen was positive with marked polymorphic leukocytosis. Also, the blood culture and culture from sinus scraping grew *Staphylococcus aureus*. Magnetic resonance imaging scan of the brain including orbit revealed osteomyelitis of maxilla and intraorbital extraconal mass with restricted diffusion, suggesting intraorbital abscess. The neonate responded to intravenous antimicrobials and decompression of the abscess. Although, the source of this major infection is unidentified, low birth weight, failure to thrive and poor socioeconomic status might have contributed. Conclusion: We report a term, low birth weight, female neonate with culture positive sepsis, maxillary sinus osteomyelitis and orbital abscess. Osteomyelitis of the maxilla is a rare entity and an important etiological factor for orbital abscess.

Keywords: Maxillary sinus, Neonate, Orbital abscess, Osteomyelitis, Proptosis

Janampally Ravikiran[1], Swathi Chacham[2], Uppin Narayan Reddy[3], Jillalla Narsing Rao[4], Ekta Aggarwal[5], Anumula Soumya[1], Nori Madhavi[6]

Affiliations: [1]MBBS, (MD, Pediatrics), Junior Resident, Department of Pediatrics, Princess Esra Hospital, Deccan College of Medical Sciences, Hyderabad, Telangana, India; [2]MD Pediatrics, DM Neonatology (PGIMER, Chandigarh), Associate Professor, Department of Pediatrics, Princess Esra Hospital, Deccan College of Medical Sciences, Hyderabad, Telangana, India; [3]MD Pediatrics, Professor and Head, Department of Pediatrics, Princess Esra Hospital, Deccan College of Medical Sciences, Hyderabad, Telangana, India; [4]MD Pediatrics, Professor, Department of Pediatrics, Princess Esra Hospital, Deccan College of Medical Sciences, Hyderabad, Telangana, India; [5]MS Ophthalmology, Department of Ophthalmology, Vason Eye Hospital, Hyderabad, Telangana, India; [6]MD Radiology, Associate Professor, Department of Radiology, Princess Esra Hospital, Deccan College of Medical Sciences, Hyderabad, Telangana, India.

Corresponding Author: Janampally RaviKiran, Office Address: Princess Esra Hospital, Deccan College of Medical Sciences, Hyderabad, Telangana, India, Residence (Mailing Address): 9-7-72/1/1, Maruthi Nagar, Santosh Nagar, Behind Yadagiri Theater Hyderabad, Telangana, India. Postal Code: 500059; Ph: +919985164888; Email: janampalli.ravikiran@gmail.com

Received: 01 December 2014
Accepted: 30 December 2014
Published: 01 March 2015

How to cite this article

Ravikiran J, Chacham S, Reddy UN, Rao JN, Aggarwal E, Soumya A, Madhavi N. A rare case of maxillary sinus osteomyelitis with intraorbital, extraconal abscess in a term low birth weight twin. Int J Case Rep Images 2015;6(3):131–137.

doi:10.5348/ijcri-201524-CR-10485

INTRODUCTION

Acute osteomyelitis (OM), a rare infectious disorder of bone infrequently manifests in neonates. The reported incidence varies from 5–12 per 100,000 infants [1], with a very high mortality rate (7.3%) [2]. The most frequently affected joints are hip, knee and shoulder, while maxillary sinus is rarely involved [3, 4]. Physiological immaturity of the immune system in neonates renders them susceptible for this condition. Premature and low birth weight (LBW) neonates are more immunocompromised when compared to term and appropriate for gestational age (AGA) infants. Invasive diagnostic and therapeutic procedures add to the risk of OM in premature and LBW neonates [5]. Osteomyelitis of maxillary bone and sinus is a rare entity in pre-antibiotic era, more so in the current era of broad spectrum antibiotics [3, 4]. This resulted from extension of infection from neighboring paranasal sinuses and rarely, it had odontogenic origin [6]. In pre-antibiotic era, it was described as a syndrome of acute OM of superior maxilla in early infancy, manifesting with ophthalmological or sinonasal symptoms, by Lieberman and Brem [6]. While the most common organism causing OM is *Staphylococcus aureus*, *Klebsiella* and other gram negative organisms are rarely reported [7, 8]. Maxillary sinus OM can lead to ocular complications, upper airway obstruction and even death [9]. Ophthalmological complications of sinusitis were reported by Hubert and Chandler in pre-antibiotic era [10, 11]. These include orbital cellulitis, orbital abscess and cavernous sinus thrombosis.

CASE REPORT

We report a 15-day-old term, 1550 grams, female neonate (twin 1) presenting with right periorbital swelling of one week duration. There was no history of injury, administration of injections or application of topical agents to the eye. Also, there was no history of pustular lesions over the eye prior to the onset of periorbital swelling. The infant had fever, purulent nasal, conjunctival discharge, poor feeding, lethargy and poor weight gain. The neonate was fed with dilute formula as the breast milk was insufficient (due to twin gestation). This was born to a second-gravid with twin gestation by spontaneous vaginal delivery and had normal extra-uterine transition. There was no history of maternal pyrexia and rash during the first trimester. Also there was no pregnancy induced hypertension, gestational diabetes and premature rupture of membranes. It was a spontaneous conception. No history of maternal blood transfusion (to suspect retro-viral pathology) and maternal hepatitis B surface antigen (HBsAg), venereal disease research laboratory (VDRL) and human immunodeficiency virus (HIV) serology were negative. Family belonged to poor socio-economic class. However, the other twin was apparently normal.

On clinical examination, the neonate was sick with sclerema and mottling. The patient had septic shock manifesting with tachycardia (heart rate 180/min), hypotension (mean arterial pressure 36 mmHg) and poor tissue perfusion (capillary filling time >3 seconds). The neonate required dobutamine infusion and shock dose hydrocortisone. Local examination revealed right eye proptosis with inflammatory edema and bulbar conjunctival erythema along with purulent conjunctival and nasal discharge (Figure 1A–B). Initial differential diagnosis of periorbital abscess, orbital tumor and cavernous sinus thrombosis was considered and the child was investigated. Sepsis screen was positive (C-reactive protein 96 mg/L) with marked polymorphic leukocytosis (white blood cell count $2.9x10^4$ cells/mm^3, neutrophils 80%) and the blood culture has grown *Staphylococcus aureus*, sensitive to augmentin and ciprofloxacin, confirming the infectious nature of the lesion.

The neonate was further evaluated to detect the nature of orbital swelling. Magnetic resonance imaging (MRI) scan of the orbit and brain revealed thick walled fluid collection with rim enhancement (Figure 2A–B). This was present over the right maxilla extending along the nasal, ethmoid sinuses to the medial wall of orbit, suggesting intraorbital extraconal mass. Further, imaging with diffusion weighted MRI scan depicted restricted diffusion (Figure 3A–B) suggesting orbital abscess. However, the brain parenchyma and other structures were normal. The neonate responded to intravenous antimicrobials as per the sensitivity pattern. Decompression of the abscess revealed, copious amounts of purulent material, which grew *Staphylococcus aureus* with similar sensitivity pattern as the blood culture. The antimicrobials were continued for a period of six weeks as recommended. Maxillary sinus OM has been implicated for the orbital abscess. Magnetic resonances imaging scan revealed OM of maxillary sinus and orbital abscess. Also, the pus from

Figure 1: (A, B) Clinical photographs showing right eye proptosis with conjunctival edema (big arrow) and purulent nasal discharge (small arrow).

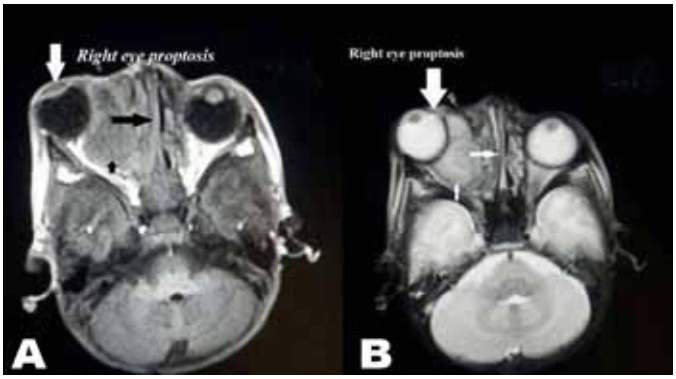


Figure 2: (A) Magnetic resonance imaging T1-weighted (axial image) scan of orbit and brain showing right eye proptosis with opacification of the right maxillary sinus (big black arrow) and a thick walled collection with rim enhancement extending from the right maxilla along the nasal, ethmoid sinuses to the medial wall of orbit, intraorbital extraconal mass (small arrow). Brain parenchyma and other structures is normal, (B) Magnetic resonance imaging T2-weighted (axial image) of orbit including brain showing similar findings (arrows).

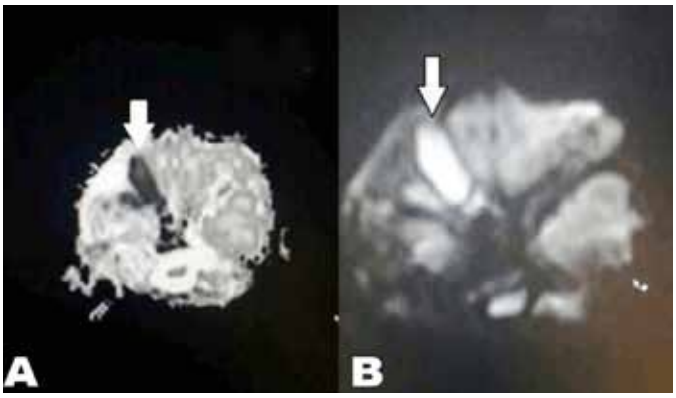


Figure 3: (A, B) Diffusion weighted magnetic resonance imaging orbit and brain revealing restricted diffusion in the right intraorbital extraconal mass suggesting orbital abscess (arrows).

orbital abscess grew *Staphylococcus aureus*. Hence, the coexistence of maxillary sinus OM and orbital abscess in a given patient can implicate the maxillary sinus OM to be the etiological factors for orbital abscess. Although, the exact cause of this major infection was unidentified, low birth weight, growth failure and poor socioeconomic status might have contributed.

DISCUSSION

In neonates, acute osteomyelitis (OM) is not only an infrequent complication, but also a diagnostic dilemma. The principal risk factors for OM in preterm and LBW infants include iatrogenic procedures like central veins lines, umbilical and intraarterial catheters. Similarly, the treatment modalities like total parental nutrition, artificial ventilation and their consequences like hospital acquired infection also predispose to OM in this high risk group [12, 13]. Being a LBW neonate, the index case was at risk for OM. However, there was no invasive monitoring and no history of umbilical catheterization in this case. The prime causes for periorbital swelling in infants and children include adjoining sinonasal and faciomaxillary infective lesions and their complications such as cavernous sinus thrombosis. These often manifest with orbital congestion and proptosis [6, 14]. The index neonate presented with fever, nasal discharge followed by periorbital swelling. Hence, the upper respiratory tract infection complicated by maxillary sinus OM might have contributed to the orbital abscess in this case. The anatomy and embryological development of the maxilla per se, predisposes it to infection from adjacent structures.

Maxillary antrum is present at birth with greater volume and high vascularity. Also, the lower border of the orbit forms the upper boundary of the maxilla, which extends down up till dental ridge with primordial follicles and embryonic teeth. Hence, this superior maxilla can acquire infection from maxillary antrum (an area highly prone for air borne infections in infants), embryonic teeth follicle and lachrymal duct or sac infection [14]. In the pre-antibiotic era, OM of maxilla as been reported in the infantile age group which had hematogenous origin.

Even in the current era, maxillary sinus OM is more prevalent in the infantile age group when compared to other age groups [14, 15]. The predominant route of neonatal OM is hematogenous. However, direct transmission of bacteria due to breach in the protecting skin and mucosae coupled with invasive procedures can occur in preterm and LBW infants [16]. But these were absent in this neonate. The commonly reported organisms for sinusitis complicated by infantile orbital cellulitis are *Staphylococcus aureus* and *Streptococcus anginosus* [17] which were present in the index neonate case. The C-reactive protein (CRP), a rapid indicator of systemic inflammation and tissue damage, is useful as an acute phase reactant, but not specific for skeletal infection. Elevated values of CRP and erythrocyte sedimentation rates could be used to monitor response to therapy or identify complications. Likewise, the index infant also had elevated CRP along with significant polymorphic leukocytosis. Radiological investigations confirm the suspicion of neonatal OM, define the infection site, differentiate between unifocal and multifocal disease patterns and identify secondary complications. Radiographs should be the first diagnostic assessment to be performed in patients with suspected OM. The first line of investigation in neonates with suspected OM is plain radiograph. This reveals involvement of the soft tissue with obliteration of fascial plains within a week of inception of OM [18]. However, deep seated OM as in the index case, might be missed in plain radiography, requiring other superior imaging modalities. Correspondingly, ultrasonography can detect features of acute OM several days earlier, than radiographs in children and infants with good tissue penetration [18]. Studies have reported

that, MRI scan is associated with good sensitivity (97%) and specificity (94%) in detecting OM, when compared to other diagnostic modalities. Also, it can demonstrate changes within 3–5 days after the commencement of infection. Likewise, it is the diagnostic modality of choice for OM in deep seated areas and rare sites like maxillary sinus [19]. In the index infant, MRI scan of orbit and brain was done to delineate the exact pathophysiology of the orbital mass and to note the route and the extension of the lesion. MRI scan showed thick walled fluid collection with rim enhancement. This was present over the right maxilla extending along the nasal, ethmoid sinuses to the medial wall of orbit, suggesting intraorbital extraconal mass. An abscess can be differentiated from necrotic neoplasm by its restricted diffusion in diffusion weighted MRI, which was noted in this case. As the brain parenchyma and other structures were normal in this case, the orbital abscess is likely to result from the adjoining maxillary sinus OM. Usually, the response to the antimicrobials is satisfactory in the pediatric age group [14]. Likewise, the orbital swelling resolved and the general condition improved with appropriate anti-microbial agents in the present case. Hence, the coexistence of maxillary sinus OM and orbital abscess in this infant implicate the maxillary sinus OM to be the etiological factors for orbital abscess.

CONCLUSION

We report a term low birth weight female (twin 1) neonate presenting with proptosis of the right eye and *Staphylococcus aureus* sepsis. Imaging revealed osteomyelitis of maxilla and intraorbital extraconal mass with restricted diffusion, suggesting orbital abscess secondary to maxillary sinus osteomyelitis. This maxillary osteomyelitis is a rare entity in infancy, and is the prime differential diagnosis for unilateral proptosis.

****

Acknowledgements
We thankful to Dr. Sana, Dr Shilpa Reddy and the NICU nursing staff or helping out with the case.

Author Contributions
Janampally Ravikiran – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Swathi Chacham – Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published
Uppin Narayan Reddy – Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published
Jillalla Narsing Rao – Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published
Ekta Aggarwal – Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published
Anumula Soumya – Acquisition of data, Analysis and interpretation of data, Drafting the article, Final approval of the version to be published
Nori Madhavi – 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.

REFERENCES

1. Rasool MN. Hematogenous osteomyelitis of the calcaneus in children. J Pediatr Orthop 2001 Nov-Dec;21(6):738–43.
2. Caksen H, Oztürk MK, Uzüm K, Yüksel S, Ustünbas HB, Per H. Septic arthritis in childhood. Pediatr Int 2000 Oct;42(5):534–40.
3. Loh FC, Ling SY. Acute osteomyelitis of the maxilla in the newborn. J Laryngol Otol 1993 Jul;107(7):627–8.
4. Skouteris CA, Velegrakis G, Christodoulou P, Helidonis E. Infantile osteomyelitis of the maxilla with concomitant subperiosteal orbital abscess: A case report. J Oral Maxillofac Surg 1995 Jan;53(1):67–70.
5. Mok PM, Reilly BJ, Ash JM. Osteomyelitis in the neonate. Clinical aspects and the role of radiography and scintigraphy in diagnosis and management. Radiology 1982 Dec;145(3):677–82.
6. Lieberman H, Brem J. Syndrome of acute osteomyelitis of the superior maxilla in Early Infancy. N Engl J Med 1959 Feb 12;260(7):318–22.
7. Qadir M, Ali SR, Lakhani M, Hashmi P, Amirali A. Klebsiella osteomyelitis of the right humerus involving the right shoulder joint in an infant. J Pak Med Assoc 2010 Sep;60(9):769–1.
8. Dunphy DL, Frazer JP. Acute osteomyelitis of the maxilla in a newborn; report of a case successfully treated with penicillin and sulfadiazine. Yale J Biol Med 1947 May;19(5):877–1.
9. Goodé-Jolly D, Besson D, Lopez A, Cabanis EA. A case of acute osteomyelitis of the maxilla in infants. Current prognosis of ocular and dental manifestations. Bull

Soc Ophtalmol Fr 1986 Oct;86(10):1141–4. [Article in French].
10. Hubert L. Orbital infections due to nasal sinusitis. NY State J Med 1937;37:1559–64.
11. Chandler JC, Lagenbrunner DL, Stevens ER. The pathogenesis of orbital complications in acute sinusitis. Laryngoscope 1970;80:1414–8.
12. Lim MO, Gresham EL, Franken EA Jr, Leake RD. Osteomyelitis as a complication of umbilical artery catheterization.(UAC). Am J Dis Child 1977 Feb;131(2):142–4.
13. Williamson JB, Galasko CS, Robinson MJ. Outcome after acute osteomyelitis in preterm infants. Arch Dis Child 1990 Oct;65(10 Spec No):1060–2.
14. Krishnan N, Ramamoorthy N, Panchanathan S, Balasundaram JS. Infantile maxillary sinus osteomyelitis mimicking orbital cellulitis. J Glob Infect Dis 2014 Jul;6(3):125–7.
15. Wilensky AO. Osteomyelitis of jaws in nurslings and infants. Ann Surg 1932 Jan;95(1):33–45.
16. Bergdahl S, Ekengren K, Eriksson M. Neonatal hematogenous osteomyelitis: Risk factors for long-term sequelae. J Pediatr Orthop 1985 Sep-Oct;5(5):564–8.
17. Kobayashi D, Givner LB, Yeatts RP, Anthony EY, Shetty AK. Infantile orbital cellulitis secondary to community-associated methicillin-resistant Staphylococcus aureus. J AAPOS 2011 Apr;15(2):208–10.
18. Pineda C, Espinosa R, Pena A. Radiographic imaging in osteomyelitis: The role of plain radiography, computed tomography, ultrasonography, magnetic resonance imaging, and scintigraphy. Semin Plast Surg 2009 May;23(2):80–9.
19. Mandell GA. Imaging in the diagnosis of musculoskeletal infections in children. Curr Probl Pediatr 1996 Aug;26(7):218–37.

ABOUT THE AUTHORS

Article citation: Ravikiran J, Chacham S, Reddy UN, Rao JN, Aggarwal E, Soumya A, Madhavi N. A rare case of maxillary sinus osteomyelitis with intraorbital, extraconal abscess in a term low birth weight twin. Int J Case Rep Images 2015;6(3):132–137.

Janampally Ravikiran is Junior Resident, Department of Pediatrics, Princess Esra Hospital, Deccan College of Medical Sciences, Hyderabad, Telangana, India. MBBS from Deccan College of Medical Sciences, Hyderabad, Telangana, India. Doing (MD) from Deccan College of Medical Sciences, Hyderabad, Telangana, India. He is a co-author in one research paper, and has published five case reports. He intends to pursue DM neonatology /cardiology in future.
Email: janampalli.ravikiran@gmail.com

Swathi Chacham is MD Pediatrics, DM Neonatology (PGIMER, Chandigarh), Associate Professor, Department of Pediatrics, Princess Esra Hospital, Deccan College of Medical Sciences, Hyderabad, Telangana, India.
Email: swathi.m.lahiri@gmail.com

Uppin Narayan Reddy is MD Pediatrics, Professor and Head, Department of Pediatrics, Princess Esra Hospital, Deccan College of Medical Sciences, Hyderabad, Telangana, India.
Email: drnarayanreddy@yahoo.co.in

Jillalla Narsing Rao is MD Pediatrics, Professor, Department of Pediatrics, Princess Esra Hospital, Deccan College of Medical Sciences, Hyderabad, Telangana, India.
Email: jillallanrao@yahoo.com

Ekta Aggarwal is MS Ophthalmology, Department of Ophthalmology, Vason Eye Hospital, Hyderabad, Telangana, India.
Email: drektaoph@gmail.com

Anumula Soumya is MD, Pediatrics, Junior Resident, Department of Pediatrics, Princess Esra Hospital, Deccan College of Medical Sciences, Hyderabad, Telangana, India

Nori Madhavi is MD Radiology, Associate Professor, Department of Radiology, Princess Esra Hospital, Deccan College of Medical Sciences, Hyderabad, Telangana, India.
Email: madhavi_dr@hotmail.com

CASE REPORT PEER REVIEWED | OPEN ACCESS

Emphysematous pyelonephritis in a non-diabetic patient associated with nephrolithiasis: A rare case report

Sônia M. H. A. Araújo, Mateus P. M. Feitosa, Priscila D. Evangelista, Sarah F. Sá, Jarinne C. L. Nasserala, Yuri Nóbrega, Pedro Miguel A. Pinheiro, Nicole Almeida de Alencar Araújo, Elizabeth F. Daher, Geraldo B. Silva Junior

ABSTRACT

Introduction: Emphysematous pyelonephritis (EP) is an acute renal infection, potentially fatal, with high mortality, which is more frequent in women at fifth decade of life. Herein, we describe a case of EP successfully treated with surgery. Case Report: A 50-year-old female was admitted with complaints of lumbar pain, irradiating to ipsilateral lower limb, associated with nausea and fever (38.9ºC), with chills. Abdominal computed tomography (CT) scan showed amorphous echogenic image, with gaseous component, inside the right kidney, and pyelogram showing lithiasis in the right renal pelvis and filling defect. Urine culture was positive for *Escherichia coli*. Antibiotic therapy was initiated with piperacillin-tazobactam and opioids. On 15th day of hospital stay, she underwent a right nephrectomy. The histopathological analysis evidenced numerous sclerosed glomeruli, tubules with atrophy, inflammatory infiltrate in the interstitium. She was discharged with complete resolution of the infection, asymptomatic and with normal renal function. Conclusion: Emphysematous pyelonephritis is a rare, atypical, and severe form of renal parenchyma infection. Early nephrectomy (<1 week) is associated with increased mortality in comparison to conservative treatment. In this case, the patient underwent an elective nephrectomy due to the chronicity of the disease, with successful recovery.

Keywords: Emphysematous pyelonephritis, Infection, Nephrolithiasis, Non-diabetic patient, Renal failure, Urinary infection

Sônia M. H. A. Araújo, Mateus P. M. Feitosa, Priscila D. Evangelista, Sarah F. Sá, Jarinne C. L. Nasserala, Yuri Nóbrega, Pedro Miguel A. Pinheiro, Nicole Almeida de Alencar Araújo, Elizabeth F. Daher, Geraldo B. Silva Junior

Affiliations: School of Medicine, University of Fortaleza. Fortaleza, Ceará, Brazil; School of Medicine, Federal University of Ceará. Fortaleza, Ceará, Brazil.

Corresponding Author: Sônia M.H.A. Araújo, Curso de Medicina. Universidade de Fortaleza. Av. Washington Soares, 1321. CEP: 60811-905. Fortaleza, Ceará, Brazil; Email: soniaholanda.unifor@gmail.com, geraldobezerrajr@yahoo.com.br

Received: 09 October 2014
Accepted: 19 November 2014
Published: 01 March 2015

How to cite this article

Araújo SMHA, Feitosa MPM, Evangelista PD, Sá SF, Nasserala JCL, Nóbrega Y, Pinheiro PMA, Araújo ADA, Daher EF, Silva Junior GB. Emphysematous pyelonephritis in a non-diabetic patient associated with nephrolithiasis: A rare case report. Int J Case Rep Images 2015;6(3):138–141.

doi:10.5348/ijcri-201525-CR-10486

INTRODUCTION

The first case of emphysematous pyelonephritis (EP) was described in 1898 by Kelly and MacCallum, and it was called 'pneumaturia'. This term was then replaced by EP in 1962 by Schultz and Klorfein [1].

Emphysematous pyelonephritis is an acute renal infection, potentially fatal, with mortality around 70–80% [2, 3], and there are approximately 200 cases reported in literature. The disease is more frequent in women at fifth decade of life, in the proportion 5.9:1 [1]. The main risk factors are diabetes mellitus (80–90%) and urinary tract obstruction (40%).

Its pathophysiology is not completely understood, but it is related to bacterial infection by gram negative and anaerobes, such as *Escherichia coli, Klebsiella*

pneumoniae, Proteus mirabilis and *Pseudomonas aeruginosa*. The production of carbon dioxide and hydrogen from the glucose fermentation by these bacteria causes inflammation and necrosis of renal parenchyma with progressive renal function loss.

The clinical manifestations are similar to an acute pyelonephritis, characterized by the triad: lumbar pain, fever and vomiting. However, a more severe evolution can be seen, mainly associated with thrombocytopenia, renal failure, sepsis and shock.

The gold-standard complimentary test for the diagnosis of EP is abdominal computed tomography (CT) scan without contrast with the finding of gas in the genitourinary tract. It is unilateral in 90% of cases. Approximately, half of the cases present is extra-renal involvement.

There is still no consensus about the best treatment for this infection, since the prevalence of this disorder is low and there are a few data in literature. Treatment can be conservative, with endovenous antibiotics and percutaneous drainage, or surgical, with nephrectomy [4].

CASE REPORT

A 50-years-old female was admitted with complaints of lumbar pain, irradiating to ipsilateral lower limb, associated with nausea and fever (38.9ºC), with chills, since 12 days ago. She referred an episode of pyelonephritis 18 years before, which has been successfully treated with antibiotics. She had also two other episodes of lumbar pain with hospitalization to treat pyelonephritis, and the last was three months before, when a hydronephrosis has been diagnosed, through ultrasound (Figure 1). Abdominal computed tomography (CT) scan revealed amorphous echogenic image, with gaseous component, inside the right kidney (Figure 2).

At physical examination she had blood pressure 110/80 mmHg, temperature 36ºC, heart rate 80 bpm, and painful lumbar percussion at right flank. Her laboratory tests showed: hemoglobin 11.5 g/dL, hematocrit 35.9%, leukocytes 4420/mm^3, platelets 2.52x10^5/mm^3, urea 39 mg/dL, creatinine 0.8 mg/dL, fast glucose 122 mg/dL, sodium 138 mEq/L, potassium 4.0 mEq/L, calcium 8.6 mg/dL, albumin 4.2 g/dL, VHS 34 mm, PCR 0.7. Urinalysis: Ph 5.0, protein traces, hemoglobin +++, leukocytes +++ (30/high power field), erythrocytes 25/high power field, moderate bacteriuria. Urine culture was positive for *Escherichia coli*, resistant to ciprofloxacin, amikacin and ceftriaxone. Antibiotic therapy was initiated with piperacillin-tazobactam and opioids. Despite antibiotic use, the patient persisted with dysuria, lumbar pain and purulent collection in the kidney. She had no signs of systemic inflammatory response syndrome, but the persistent pain and absence of regression of the collection with gas suggestive of emphysematous pyelonephritis (EP) refractory to clinical treatment. On 15th day of hospital stay, the patient underwent a right nephrectomy (Figure 3). The histopathological analysis evidenced numerous sclerosed glomeruli, tubules with atrophy, inflammatory infiltrate in the interstitium (Figure 4). Pyelocaliceal system was dilated and re-vested with squamous metaplasia, containing partially calcified material. These findings were compatible with chronic pyelonephritis, acute tubular necrosis and hydronephrosis. She was discharged with complete resolution of the infection, asymptomatic and with normal renal function.

DISCUSSION

Emphysematous pyelonephritis is a rare, atypical, and severe form of renal parenchyma infection [5]. This is an acute infection, potentially fatal, with mortality around 70–80%. Emphysematous pyelonephritis is a necrotizing infection of the renal parenchyma characterized by the production of gas in the intra- and perirenal tissues [6, 7]. It is believed that high levels of glucose, in association with

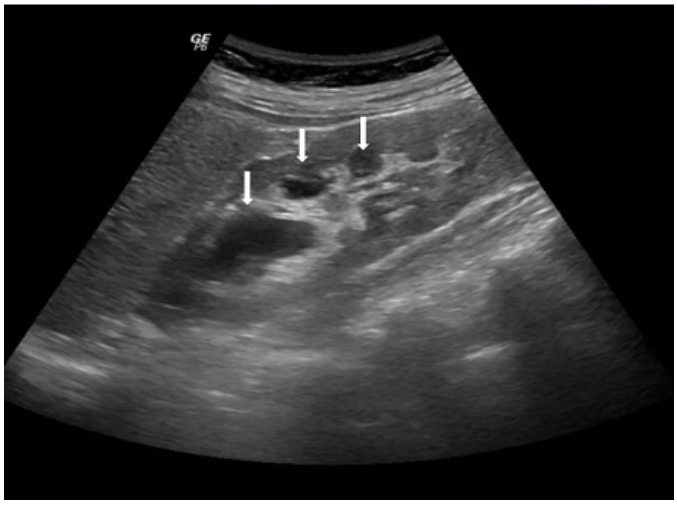


Figure 1: Ultrasound showing increased right kidney, with pyelocaliceal dilation (arrows).

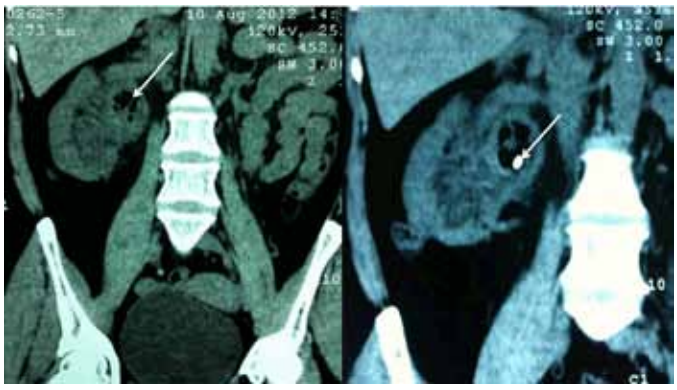


Figure 2: Computed tomography scan image showing increased right kidney, with heterogeneous lesion with gaseous component (left arrow) and calcified image (right arrow).

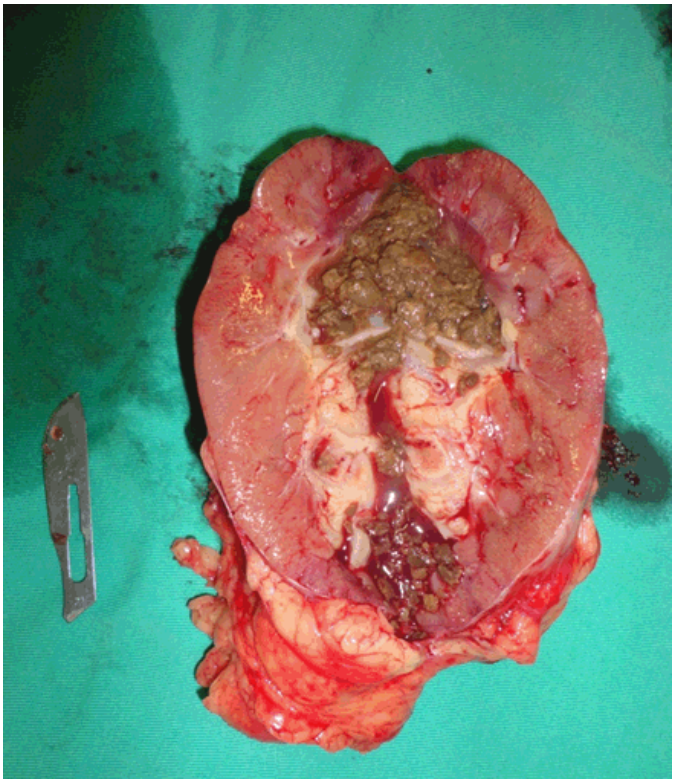


Figure 3: Macroscopic image of right kidney after nephrectomy showing purulent secretion and thickening of the renal parenchyma.

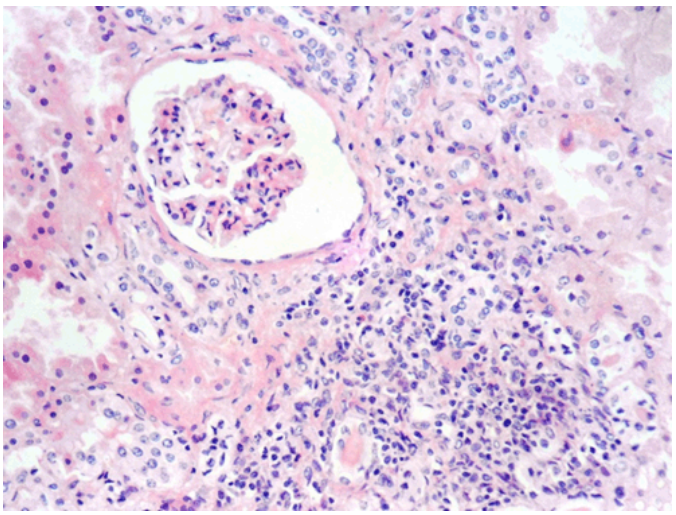


Figure 4: Histopathological analysis evidencing sclerosed glomerulus, tubules with atrophy and inflammatory infiltrate in the interstitium.

inadequate perfusion, lead to a favorable environment for the growth of anaerobic organisms. This disease affects individuals of all ages, but women are six times more likely to be affected [6, 7].

Emphysematous pyelonephritis is more common in patients with diabetes [1]. Our patient did not have diagnosis of diabetes, but had altered fast glucose. An obstructive factor was found in the present case, nephrolithiasis, which is a known risk factor for chronic kidney disease. She had history of recurrent urinary tract infections, with increased frequency in the last years. Emphysematous pyelonephritis was suspected after doing a CT scan, which showed a gaseous component in the right kidney.

Huang and Tseng [8] proposed a radiological classification for emphysematous pyelonephritis in four classes: (1) gas in the collecting system only, (2) gas in the renal parenchyma without extension to extrarenal space, (3A) extension of gas or abscess to perinephric space, (3B) extension of gas or abscess to pararenal space, and (4) bilateral EPN or solitary kidney with emphysematous pyelonephritis. According to this classification, the case presented here falls in class II, which is associated with a better prognosis. Some factors are associated with poor prognosis such as thrombocytopenia, renal failure, hyponatremia and sepsis. No one of these factors was present in our patient.

Based on the cases published by now, there is no consensus regarding the best treatment conservative management, with endovenous antibiotics and percutaneous drainage, or surgery, with nephrectomy of the involved kidney should be chosen according to each case. Early nephrectomy (<1 week) is associated with increased mortality in comparison to conservative treatment. In the present case, the patient underwent an elective nephrectomy due to the chronicity of the disease. Patients with chronic pyelonephritis and renal function <10% evidenced by scintigraphy benefits with nephrectomy.

CONCLUSION

In summary, we reported a rare case of emphysematous pyelonephritis in a non-diabetic patient. Renal lithiasis was evidenced in this case, and it represents a possible risk factor for pyelonephritis due to chronic urinary tract obstruction. Repeated history of pyelonephritis is another important factor for emphysematous pyelonephritis.

****

Author Contributions

Sônia M. H. A. Araújo – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Mateus P. M. Feitosa – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Priscila D. Evangelista – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Sarah F. Sá – Substantial contributions to conception and design, Drafting the article, Revising it critically

for important intellectual content, Final approval of the version to be published
Jarinne C. L. Nasserala – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Yuri Nóbrega – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Pedro Miguel A. Pinheiro – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Nicole Almeida de Alencar Araújo – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Elizabeth F. Daher – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
Geraldo B. Silva Junior – Substantial contributions to conception and design, 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. Carvalho M, Goulão J, Monteiro C, et al. Pielonefrite enfisematosa: revisão da literatura a propósito de um caso clínico. Acta Urol 2006;23:75–80.
2. Junior M, Ferreira G, Lopes H, Amaral K. Pielonefrite enfisematosa: revisão e atualização da abordagem terapêutica. HU Revista (Juiz de Fora) 2010;36:161–5.
3. Cavalli AC, Tambara Filho R, Cavalli RC, Bressan F. Emphysematous pyelonephritis: importance of early diagnosis. Rev Méd Paraná (Curitiba) 2006;64:13–15.
4. Eloubeidi MA, Fowler VG Jr. Images in clinical medicine. Emphysematous pyelonephritis. N Engl J Med 1999 Sep 2;341(10):737.
5. Lim CS, Kim WB, Kim YS, et al. Bilateral emphysematous pyelonephritis with perirenal abscess cured by conservative therapy. J Nephrol 2000 Mar-Apr;13(2):155–8.
6. McGorry DM, Kroser J, Taylor LT, Howard Lewis H, Gabale D. Emphysematous Pyelonephritis Presenting as an Acute Abdomen. Infect Urol 1999;12:162–5.
7. Jain SK, Agarwal N, Chaturvedi SK. Emphysematous pyelonephritis: A rare presentation. J Postgrad Med 2000 Jan-Mar;46(1):31–2.
8. Huang JJ, Tseng CC. Emphysematous pyelonephritis: clinicoradiological classification, management, prognosis, and pathogenesis. Arch Intern Med 2000 Mar 27;160(6):797–805.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Ureteroscopy assisted retrograde nephrostomy in patient with lower pole renal stone

Takashi Kawahara, Hiroki Ito, Hideyuki Terao, Hiroji Uemura, Yoshinobu Kubota, Junichi Matsuzaki

ABSTRACT

Introduction: We previously reported a new technique, ureteroscopy-assisted retrograde nephrostomy (UARN) for percutaneous nephrolithotomy (PCNL). This case report describes a new approach for treating renal stones in the lower calyx using the UARN technique. Case Report: A 68-year-old female was referred to our department for the treatment of a right renal stone. We performed PCNL using the technique for UARN. Conclusions: We herein report a new technique using UARN for lower pole renal stones. This method may represent a new treatment option for renal stones in the lower calyx.

Keywords: Nephro access sheath (NAS), Percutaneous nephrolithotomy (PCNL), Renal stones, Ureteral access sheath, Ureteroscopy, Ureteroscopy-assisted retrograde nephrostomy (UARN)

Takashi Kawahara[1], Hiroki Ito[2], Hideyuki Terao[1], Hiroji Uemura[3], Yoshinobu Kubota[3], Junichi Matsuzaki[1]

Affiliations: [1]Department of Urology, Ohguchi Higashi General Hospital, 2-19-1, Irie, Kanagawa-ku, Yokohama City, Kanagawa, Japan; [2]Department of Urology, Ohguchi Higashi General Hospital, Department of Urology, Yokohama City University Graduate School of Medicine, Japan; [3]Department of Urology, Yokohama City University Graduate School of Medicine, Japan.

Corresponding Author: Takashi Kawahara, MD, Department of Urology, Ohguchi Higashi General Hospital, 2-19-1, Irie, Kanagawa-ku, Yokohama City, Kanagawa, Japan, Facsimile: +81-45-431-6920, Phone: +81-45-401-2411; Email: takashi_tk2001@yahoo.co.jp

Received: 01 December 2014
Accepted: 19 December 2014
Published: 01 March 2015

How to cite this article

Kawahara T, Ito H, Terao H, Uemura H, Kubota Y, Matsuzaki J. Ureteroscopy assisted retrograde nephrostomy in patient with lower pole renal stone. Int J Case Rep Images 2015;6(3):142–144.

doi:10.5348/ijcri-201526-CR-10487

INTRODUCTION

For large renal stones in the lower calyx, percutaneous nephrolithotomy (PCNL) has become the preferred method of treatment in patients with a large or complex stone burden.

We previously reported a new technique, ureteroscopy-assisted retrograde nephrostomy (UARN) for PCNL, which allows for continuous visualization from puncture to the insertion of the nephro access sheath (NAS), resulting in a higher stone-free rate and fewer complications. Although the ureteroscope can usually be used to reach the target puncture spot, the URS with a puncture wire cannot reach the lower calyx because it is too stiff. This report describes a new approach for treating renal stones in the lower calyx using the UARN technique.

CASE REPORT

A 68-year-old female was referred to our department for the treatment of a right renal stone. In April 2012, the patient was admitted to our department for PCNL of a right lower pole renal stone. A preoperative kidney ureter bladder (KUB) film and computed tomography (CT) scan showed a maximum stone diameter of 23 mm, with no evidence of hydronephrosis (Figure 1A).

We did not consider that ureteroscopic lithotripsy would provide a stone-free outcome because the stone's diameter was more than 20 mm. Therefore, we performed UARN, following the method described in our previous manuscript [1]. Briefly, we confirmed that performing puncture from the lower pole was difficult. However, puncturing from the middle calyx was considered to be associated with a higher risk of not being able to reach the target stone with the nephroscope because the entrance to the lower calyx was occupied by the NAS (Figure 1B). Despite generating hydronephrosis using a high irrigation pressure supplied by the URS, performing percutaneous puncture under ultrasonography with fluoroscopic guidance was not successful, as the lower calyx was not sufficiently dilated for puncture. We first punctured the site from the middle calyx using the UARN technique, as described in our previous report [2]. Briefly, after the puncture wire was easily passed through the renal capsule and the skin at the posterior axillary line was tented, a 12-Fr safety catheter was inserted under visualization with the URS; the outer 12-Fr catheter was left in place to drain the irrigation flow. After conducting ureteroscopic lithotripsy using a holmium:yttrium aluminum garnet (Ho:YAG) laser to create stone fragments, the calculus fragments were washed out from the lower calyx into the renal collecting system using the irrigation flow (Figure 1C–E). Then, a 24-Fr NAS (X-Force® Nephrostomy Balloon Dilation Catheter, BARD, Murray Hill, NJ, USA) was inserted, and the stone fragments were retracted using a nephroscope. Finally, a ureteral stent was inserted at the conclusion of the PCNL procedure and subsequently removed two weeks after the operation. Postoperative nephrostomy was not performed. A stone-free status was confirmed on computed tomography (CT) scan; the stone composition included calcium oxalate monohydrate.

DISCUSSION

Treating renal stones in the lower calyx is challenging, even when shockwave lithotripsy can be used to successfully create stone fragments, due to the anatomical structure in this area [3]. URS is a candidate for a new therapeutic approach for treating renal stones in the lower calyx [4, 5]. The American Urological Association (AUA) and European Association Urology (EAU) guidelines both recommend PCNL as the first-line therapy for renal stones larger than 2 cm [4, 5].

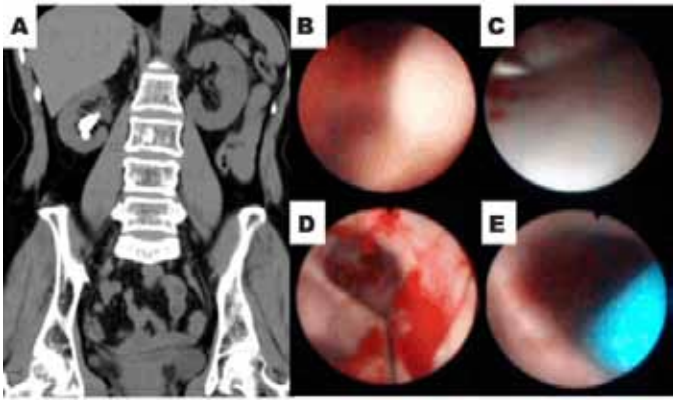


Figure 1: (A) Preoperative axial CT. Ureteroscopic findings, (B) The renal stone occupied the lower pole. (C, D) Puncturing from the upper calyx to the skin, and (E) Dilationof the nephrostomy under ureteroscopic guidance.

We previously described a new technique for UARN [1]. Using UARN, it is possible to continuously visualize the region from the initial puncture to the insertion of the NAS. The most important limitation of UARN is that it cannot be used to puncture from the lower calyx. Notably, the URS with the puncture wire is too stiff, making it impossible to reach the lower calyx. Therefore, in most cases without a horseshoe kidney, the target calyx is the middle calyx on the dorsal side [6]. However, when the target stone is located in the lower calyx, inserting the NAS may block the entrance to the lower calyx, which can also make it difficult or impossible to advance the nephroscope to the target stone [7].

In this case, before dilating the balloon, we inserted a 12-Fr catheter and created stone fragments using Ho:YAG laser lithotripsy. After washing out the stone fragments from the lower calyx via the irrigation flow, balloon dilation and NAS insertion was performed and the stone fragments were easily removed through the NAS. This procedure allowed for the use of a higher irrigation flow with less intrarenal pressure as a result of the drainage provided by the 12-Fr catheter. This method may help decrease the risk of postoperative urinary tract infections.

The most critical issue associated with PCNL is the proper selection of the puncture site to minimize the risk of hemorrhage, the most common major complication of this procedure [1, 8]. US-guided puncture to the renal collecting system with subsequent placement of a drainage tube under fluoroscopic guidance is the standard modality for performing PCNL. However, applying US-guided nephrostomy is difficult without dilating the renal collecting system, even if an occlusion balloon catheter is used to create hydronephrosis [9]. The limitation of this procedure is that it is only effective for relatively small stones after initial fragmentation using a Ho:YAG laser.

CONCLUSION

We herein reported a new technique using ureteroscopy-assisted retrograde nephrostomy (UARN) for lower pole renal stones. This method may represent a new treatment option for renal stones in the lower calyx.

****

Author Contributions

Takashi Kawahara – 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

Hiroki Ito – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Hideyuki Terao – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Hiroji Uemura – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Yoshinobu Kubota – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Junichi Matsuzaki – 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. Kawahara T, Ito H, Terao H, et al. Ureteroscopy assisted retrograde nephrostomy: A new technique for percutaneous nephrolithotomy (PCNL). BJU Int 2012 Aug;110(4):588–90.
2. Kawahara T, Matsuzaki J, Kubota Y. Ureteroscopy-assisted retrograde nephrostomy for an obese patient. Indian J Urol 2012 Oct;28(4):439–1.
3. de la Rosette J, Assimos D, Desai M, et al. The Clinical Research Office of the Endourological Society Percutaneous Nephrolithotomy Global Study: Indications, complications, and outcomes in 5803 patients. J Endourol 2011 Jan;25(1):11–7.
4. Fernstrom I, Johansson B. Percutaneous pyelolithotomy. A new extraction technique. Scand J Urol Nephrol 1976;10(3):257–9.
5. Skolarikos A, Alivizatos G, de la Rosette JJ. Percutaneous nephrolithotomy and its legacy. Eur Urol 2005 Jan;47(1):22–8.
6. Kawahara T, Ito H, Terao H, et al. Ureteral stent exchange under fluoroscopic guidance using the crochet hook technique in women. Urol Int 2012;88(3):322–5.
7. Fischbach F, Porsch M, Krenzien F, et al. MR imaging guided percutaneous nephrostomy using a 1.0 Tesla open MR scanner. Cardiovasc Intervent Radiol 2011 Aug;34(4):857–63.
8. Wynberg JB, Borin JF, Vicena JZ, Hannosh V, Salmon SA. Flexible Ureteroscopy-Directed Retrograde Nephrostomy for Percutaneous Nephrolithotomy: Description of a Technique. J Endourol 2012 Oct;26(10):1268–74.
9. Ibarluzea G, Scoffone CM, Cracco CM, et al. Supine Valdivia and modified lithotomy position for simultaneous anterograde and retrograde endourological access. BJU Int 2007 Jul;100(1):233–6.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Epigastric pain: Incarceration or rotation?

Ana Franky Carvalho, Ana João Rodrigues, Pedro Leão

ABSTRACT

Introduction: Acute intrathoracic gastric volvulus occurs when the stomach has a twist mesenteroaxial/organoaxial or chest cavity resulting in a dilatation or rupture of the diaphragmatic hiatus or diaphragmatic hernia. The purpose of this work is to show a interesting case of gastric volvulus in a patient with several comorbidities. Case Report: A 77-year-old female with past history of hiatal hernia and mental disease associated with diabetes and atrial fibrillation. Patient went to the emergency department due to vomiting associated with blood. Analytical parameters (WBC, HGB, PCR, metabolic panel and liver function), showed no significant alterations. Thoracic X-ray revealed an enlarged mediastinum due to herniation of the stomach. A computed tomography (CT) scan confirmed intrathoracic localization of the gastric antrum with twist. Patient's symptoms were relieved by nasogastric intubation and analgesia. After six months, the patient is still asymptomatic. Conclusion: In general, the treatment of an acute gastric volvulus requires an emergent surgical repair. In patients who are not surgical candidates (with comorbidities or an inability to tolerate anesthesia), endoscopic reduction should be attempted. Chronic gastric volvulus may be treated non-emergently, and surgical treatment is increasingly being performed using a laparoscopic approach. In this case, it is a chronic form that was solved with the placement of the nasogastric tube. A nasogastric decompression is an option in the chronic form of hiatal hernia associated to gastric volvulus in patients with serious comorbidities.

Keywords: Epigastric pain, Gastric volvulus, Hiatal hernia, Mesenteroaxial, Nasogastric tube

Ana Franky Carvalho[1], Ana João Rodrigues[2], Pedro Leão[3]

Affiliations: [1]MD, PhD, Serviço de Cirurgia Geral – Hospital de Braga, Life and Health Sciences Research Institute (ICVS), School of Health Sciences, University of Minho, Braga, ICVS/3B's - PT Government Associate Laboratory, Braga/Guimarães, Portugal; [2]PhD, Life and Health Sciences Research Institute (ICVS), School of Health Sciences, University of Minho, Braga, ICVS/3B's - PT Government Associate Laboratory, Braga/Guimarães, Portugal; [3]MD, Ph.D, Serviço de Cirurgia Geral – Hospital de Braga, Life and Health Sciences Research Institute (ICVS), School of Health Sciences, University of Minho, Braga, ICVS/3B's - PT Government Associate Laboratory, Braga/Guimarães, Portugal.

Corresponding Author: Pedro Leão, General Surgery, Hospital of Braga, 4701-965, Braga, Apartado 2242, Portugal; Ph: (+351) 915303818; Email: pedroleao@ecsaude.uminho.pt

Received: 30 June 2014
Accepted: 29 July 2014
Published: 01 March 2015

How to cite this article

Carvalho AF, Rodrigues AJ, Leão P. Epigastric pain: Incarceration or rotation? Int J Case Rep Images 2015;6(3):145–148.

doi:10.5348/ijcri-201527-CR-10488

INTRODUCTION

Gastric volvulus (GV) is an abnormal rotation of the stomach. The primary etiology is laxity or agenesis of the gastric ligaments. It may also develop adhesions, hiatal hernia and other causes [1], It was first described by Berti in 1866 [2]. According to the axis of rotation, it is classified into organoaxial:

(i) longitudinal axis parallel to the imaginary line; cardiopyloric being the most common (60%)

(ii) Mesentric axial transverse axis perpendicular to the line; cardiopyloric (30%) and

(iii) mixed style - a combination of the both [3]

The acute GV, represented by sudden abdominal pain, vomiting and triad Borchardt (epigastric distention, inability to pass the gastric tube and ineffectual efforts to vomit), is prone to tissue ischemia, necrosis and gastric perforation, and is considered an emergency surgery [2–4].

The chronic form of presentation is asymptomatic or oligosymptomatic, and may be responsible for uncharacteristic symptoms of abdominal discomfort and heart burn [4, 5] and often requires a new procedure to treat the condition, especially if the patient presents several comorbidities. Here, we present a different concept in GV treatment.

CASE REPORT

A 77-year-old female with past history of diabetes, mental disease and hiatal hernia complained of epigastric pain and abdominal cramps, more evident after meals and relieved by vomiting. Patient went to the emergency department due to vomiting associated with blood and halitosis. Analytical parameters (WBC, HGB, PCR, metabolic panel and liver function), showed no significant alterations.

Thoracic X-ray revealed an enlarged mediastinum due to herniation of the stomach. The stomach bubble (gastric fundus) could be seen in its usual position suggesting herniation of the gastric antrum (Figure 1). A computed tomography (CT) scan confirmed intrathoracic localization of the gastric antrum (Figure 2). The diagnosis is a hiatal hernia with gastric mesenterioaxial rotation type, as depicted in Figure 2, where the arrow indicates the duodenum near the hernia position. An upper gastrointestinal endoscopy showed gastric stasis. The exam was interrupted due to patient intolerance, suggestive of gastric torsion. The patient's symptoms were relieved by nasogastric intubation and analgesia. After six months of follow-up the patient still asymptomatic.

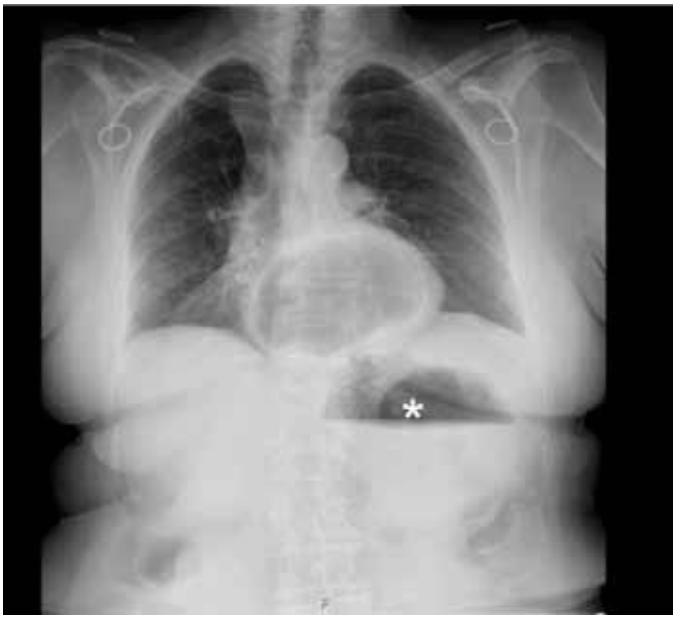


Figure 1: X-ray showing an enlargement of mediastinum due to a gastric antrum. Asterisk (*) indicates gastric fundus.

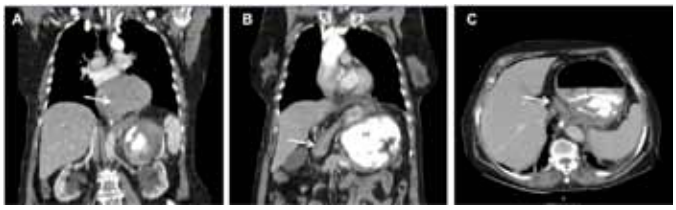


Figure 2: Computed tomography scan images (A) A coronal view of gastric antrum position in mediastinum (arrow), (B) A coronal view of duodenum position (arrow), and (C) A transversal view a first portion of duodenum towards in hiatal hernia (arrow).

DISCUSSION

Gastric volvulus is a complete obstruction of the gastric lumen by rotation, that can occur at any age, with equal frequency in men and women [6]. In 25% of patients, gastric rotation is primary or idiopathic and occurs because there is a lengthening of ligaments [5–7]. In 75% of patients, GV is associated with a pathological factor namely: hiatus hernia, diaphragmatic hernia resulting from trauma, herniation of the left diaphragm phrenic nerve injury, chronic pyloric obstruction with dilatation of the stomach or prior gastroesophageal surgery [6, 7]. In the presented case, the most probable etiology is laxity ligaments and hiatal hernia, although we cannot exclude other causes.

The most commonly used classification was proposed by Singleton and describes three types of GV, according to the rotational axis:

Type 1 or organoaxial is the most common (59%) and the rotation occurs about a line drawn from the pylorus to the esophagus gastric junction.

Type 2 or mesenteroaxial occurs in about 29% of patients and rotation turns on an axis that connects the greater curvature and the hepatic hilum.

As we can see in the given images of CT scan; Type 3 is a rare form (3%), which combines both types 1 and 2 [1–4].

Supplementary examination in these patients is important for diagnosis. In the analytical control, there may be a hyperamylasemia and elevated levels of lactate dehydrogenase and alkaline phosphatase. Gastrointestinal contrast studies, barium or gastrografin, have high sensitivity and specificity. The endoscopy usually shows a high deformation with gastric pylorus and difficult access, and in the most advanced stage of the disease, mucosal ulcerations. The CT scan has important diagnostic value in GV, as for example in this case, where it was performed in the acute phase and provided a rapid diagnostic. Moreover, this examination can detect the

presence of pneumatosis or pneumoperitoneum in case of necrosis and perforation [6].

Recent data suggest that routine elective repair of completely asymptomatic paraesophageal hernias may not be indicated. Surgical treatment of gastric voluvus includes reduction of the stomach and limited gastric resection in cases of gastric necrosis. The laparoscopic approach can be used in most of the cases, but conversion to open access should be considered for complex problems or for the safety of the patient [8, 9].

Large hiatal hernias with or without gastric volvulus can be repaired either transabdominally (open or laparoscopic) or via thoracotomy. However to date, there is no randomized trials directly comparing open transthoracic versus open transabdominal repair.

In the surgical approach, we need to take to consideration the following four hallmarks:

Hernia sac excision Sac dissection during paraesophageal hernia repair is thought to release the tethering of the esophagus, facilitating reduction of the hernia and the decrease of early recurrence, as well as protecting the esophagus from iatrogenic damage [10].

Reinforced repair Primary sutured crural repair has been the main option for many years, but follow-up has suggested very high recurrence rates (>42%) after laparoscopic paraesophageal hernia repair [11]. Several case series suggests benefit with mesh, however, there are a few which question the use of meshed repair [12].

Fundoplication The majority of reports in the recent literature describe the performance of a fundoplication as a step of the repair. In a case-controlled study, surgeons found increased dysphagia with fundoplication, and of reflux symptoms in the group without fundoplication, thus routine fundoplication should be avoided [13].

Gastropexy One of the first studies using anterior gastropexy to reduce the recurrence rate after laparoscopic hiatal hernia repair with gastric volvulus showed no recurrences up to two years of follow-up evaluation [14]. This finding has been supported by a recent study showing that the addition of an anterior gastropexy significantly reduced recurrent hernias. However, other reports found no significant difference in recurrence rate [15].

In this case, the placement of a nasogastric tube has the function of gastric decompression, and solved the gastric volvulus. A nasogastric decompression is possible only because the cardia mesenteroaxial VG is open. The endoscopic “des rotation” have satisfactory results but it is a temporary solution, being the definitive treatment the surgical approach. Urgent surgery in acute cases is fundamental and its delay increases mortality [4]. Contraindications for surgical treatment involve conditions or comorbidities in which the patient cannot tolerate general anesthesia. The surgeon should also use clinical judgment and make sure the patient conditions are optimal before the operation.

CONCLUSION

A nasogastric decompression is an option in chronic form of hiatal hernia associated to gastric volvulus.

****

Author Contributions

Ana Franky Carvalho – 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

Ana João Rodrigues – Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Pedro Leão – Acquisition of data, 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. Kulkarni K, Nagler J. Emergency endoscopic reduction of a gastric volvulus. Endoscopy 2007 Feb;39 Suppl 1:E173.
2. Coulier B, Broze B. Gastric volvulus through a Morgagni hernia: Multidetector computed tomography diagnosis. Emerg Radiol 2008 May;15(3):197–201.
3. Wasselle JA, Norman J. Acute gastric volvulus: pathogenesis, diagnosis, and treatment. Am J Gastroenterol 1993 Oct;88(10):1780–4.
4. Axon PR, Whatling PJ, Dwerryhouse S, Forrester-Wood CP. Strangulated iatrogenic diaphragmatic hernia: A late diagnosed complication. Eur J Cardiothorac Surg 1995;9(11):664–6.
5. Iannelli A, Fabiani P, Karimdjee BS, Habre J, Lopez S, Gugenheim J. Laparoscopic repair of intrathoracic mesenterioaxial volvulus of the stomach in an adult: Report of acase. Surg Today 2003;33(10):761–3.

6. Testini M, Vacca A, Lissidini G, Di Venere B, Gurrado A, Loizzi M. Acute intrathoracic gastric volvulus from a diaphragmatic hernia after left splenopancreatectomy: Report of a case. Surg Today 2006;36(11):981–4.
7. Alrabeeah A, Giacomantonio M, Gillis DA. Paraesophageal hernia after Nissen fundoplication: A real complication in pediatric patients. J Pediatr Surg 1988 Aug;23(8):766–8.
8. Katkhouda N, Mavor E, Achanta K, et al. Laparoscopic repair of chronic intrathoracic gastric volvulus. Surgery 2000 Nov;128(5):784–90.
9. Koger KE, Stone JM. Laparoscopic reduction of acute gastric volvulus. Am Surg 1993 May;59(5):325–8.
10. Watson DI, Davies N, Devitt PG, Jamieson GG. Importance of dissection of the hernial sac in laparoscopic surgery for large hiatal hernias. Arch Surg 1999 Oct;134(10):1069–73.
11. Wiechmann RJ, Ferguson MK, Naunheim KS, et al. Laparoscopic management of giant paraesophageal herniation. Ann Thorac Surg 2001 Apr;71(4):1080–6.
12. 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.
13. Morris-Stiff G, Hassn A. Laparoscopic paraoesophageal hernia repair: Fundoplication is not usually indicated. Hernia 2008 Jun;12(3):299–302.
14. Ponsky J, Rosen M, Fanning A, Malm J. Anterior gastropexy may reduce the recurrence rate after laparoscopic paraesophageal hernia repair. Surg Endosc 2003 Jul;17(7):1036–41.
15. Diaz S, Brunt LM, Klingensmith ME, Frisella PM, Soper NJ. Laparoscopic paraesophageal hernia repair, a challenging operation: medium-term outcome of 116 patients. J Gastrointest Surg 2003 Jan;7(1):59–66.

CASE REPORT PEER REVIEWED | OPEN ACCESS

ST-elevation myocardial infarction secondary to paradoxical coronary emboli in a patient with massive pulmonary embolism and essential thrombocytosis: A case report

Fahad S. Almehmadi, Albayda M. Mehdar, Kumar Sridhar, Patrick Teefy

ABSTRACT

Introduction: Essential thrombocytosis (ET) is a myeloproliferative disorder with higher incidence of thrombotic events. To our knowledge, we present the first case of ST-segment elevation myocardial infarction (STEMI) secondary to paradoxical right coronary artery (RCA) embolus through a patent foramen ovale (PFO) in a patient with essential thrombocytosis and pulmonary embolus. Case Report: A 67-year old female with a history of ET presented to the emergency room with dyspnea. Physical examination revealed an elevated JVP, an S1Q3T3 pattern on her presenting ECG, and an elevated D-dimer. V/Q scan showed a high probability for pulmonary embolism as well as unusual evidence of right-to-left cardiac shunting. After starting low molecular weight heparin, she developed new-onset chest pain and her ECG showed ST-elevation in the inferior leads. Emergency left and right heart catheterization showed an acutely occluded RCA with heavy thrombus burden. This was managed successfully with thrombus aspiration only. Massive bilateral pulmonary embolism was seen on thoracic computed tomography (CT) scan, which was managed by systemic thrombolysis. A Transesophageal echocardiogram was performed, which confirmed a patent PFO with right-to-left shunting. The patient was treated medically with dual antiplatelets, anticoagulation with heparin and hydroxyurea. Given the degree of thrombotic burden PFO closure was not performed and the patient was managed conservatively with lifelong anticoagulation. The patient has been followed closely, and three years post-event, she has done remarkably well on warfarin with no evidence of further thromboembolism. Conclusion: We describe the first case of paradoxical coronary artery embolism through a PFO in a patient with ET and massive PE. Our patient was managed conservatively on oral anticoagulation without further thromboembolic events at three years post-event.

Keywords: Essential thrombocytosis, Myocardial infarction, Paradoxical embolism, Patent foramen ovale, Pulmonary embolism

Fahad S. Almehmadi[1], Albayda M. Mehdar[2], Kumar Sridhar[3], Patrick Teefy[4]

Affiliations: [1]MBBS, FRCPC, Adult Cardiology Resident, Department of Medicine, University of Toronto, Toronto, Ontario, Canada, Adult Cardiology Scholar at King Saud Bin Abdulaziz University for Health Sciences, Jeddah, Saudi Arabia; [2]MBBS, Internal Medicine Resident, Department of Medicine, University of Toronto, Toronto, Ontario, Canada; [3]MD, FRCPC, Associate Professor, Department of Medicine, University of Western Ontario, London, Ontario, Canada; [4]MD, FRCPC, Associate Cardiology Professor, Department of Medicine, University of Western Ontario, London, Ontario, Canada.

Corresponding Author: Dr. Fahad Saleh Almehmadi, Adult Cardiology Program, University of Toronto, 2305-1055 Bay Street, Toronto, Ontario, M5S 3A3, Canada; Tel: 416-567-5516; Email: falmehma@gmail.com

Received: 13 October 2014
Accepted: 19 November 2014
Published: 01 March 2015

How to cite this article

Almehmadi FS, Mehdar AM, Sridhar K, Teefy P. ST-elevation myocardial infarction secondary to paradoxical coronary emboli in a patient with massive pulmonary embolism and essential thrombocytosis: A case report. Int J Case Rep Images 2015;6(3):149–155.

doi:10.5348/ijcri-201528-CR-10489

INTRODUCTION

Essential thrombocytosis (ET) is a myeloproliferative disorder characterized by pathological clonal proliferation of megakaryocytes and thrombocythemia [1]. Essential thrombocytosis can be complicated by both thrombotic and/or hemorrhagic sequelae. De Novo coronary thrombus is rare, but has been described in literature [2–11]. We present a patient with ET and an unusual presentation of inferior myocardial infarction secondary to paradoxical embolus through a patent foramen ovale (PFO) in the setting of acute pulmonary embolism.

CASE REPORT

A 67-year-old female presented to the emergency department with rapidly progressing dyspnea and epigastric pain. Her past medical history was significant for ET, hypertension, atrial fibrillation, and osteoporosis. Her ET was diagnosed six years ago based on persistent thrombocythemia (1100×10^9/dL). Her medications included aspirin 81 mg and hydroxyurea 1000/500 mg on alternate days. On presentation, she was tachycardic, tachypneic and hypoxic. Physical examination revealed an elevated JVP and bilateral lower limb edema. Initial blood work showed a white blood cell count 7.1×10^9/dL, hemoglobin 14.9 g/dL, platelet count 412×10^9/dL, D-dimer >4000, creatine kinase 59, and troponin-T 0.03. Presenting ECG showed a typical S1Q3T3 pattern and symmetric T-wave inversion in anterior precordial leads (Figure 1). Her chest X-ray was unremarkable. Acute pulmonary embolus was suspected and a V/Q scan revealed high probability of pulmonary embolism. V/Q scan unexpectedly revealed uptake of radiotracer in the kidneys, suspicious for a right-to-left cardiac shunt. (Figure 2) Transthoracic echocardiogram showed severe right ventricular dilation with septal shift and left-to-right intra-atrial shunting, suggestive of a PFO. The patient was started on subcutaneous low molecular weight heparin for a presumed pulmonary embolism.

She remained clinically stable for eight hours, and then suddenly developed worsening chest pain, hypotension, and worsening hypoxia. Repeat ECG revealed ST-segment elevation in leads II, III and aVF consistent with STEMI (Figure 3). She was immediately intubated, started on dopamine and taken for cardiac catheterization. Left and right heart catheterization showed normal left main, left anterior descending, and left circumflex arteries, and a completely occluded right coronary artery (RCA) (Figure 4). Thrombectomy to the RCA was performed with a 6 F Export® thrombectomy catheter with concomitant administration of intra-coronary abciximab. TIMI 3 flow in RCA was established though distal embolization to the posterior descending and posterior-lateral arteries was apparent (Figure 5) that subsequently cleared when additional unfractionated heparin was bloused (Figure 6). Right heart catheterization showed an elevated RV pressure of 70/18 mmHg. Oxygen saturation analysis of the right-sided chambers did not reveal a "step-up" to suggest a large left-to-right shunt. Moreover, trans-septal puncture and left sided chamber oxygen saturation measurements also did not reveal a significant step down form femoral artery's readings.

The patient was started on an abciximab infusion for 12 hours and admitted to the intensive care unit. She continued to deteriorate hemodynamically and recombinant tissue plasminogen activator (tPA) was given. Transesophageal echocardiography showed a markedly dilated and hypokinetic right ventricle, a redundant and hypermobile inter-atrial septum with an 8-mm connection suggestive of a PFO (Figure 7). Bubble study showed intense and immediate opacification of the left atrium, confirming right-to-left shunting (Figure 8). There was no evidence of left atrial or left ventricular thrombi. The computed tomography (CT) pulmonary angiography confirmed the diagnosis of massive bilateral pulmonary emboli. The patient did well post coronary thrombectomy and systemic thrombolysis with no further thrombotic sequelae or hemorrhagic complications. She was started on unfractionated heparin and transferred to the cardiology ward one week later. She was transitioned to warfarin with a target INR between 2 and 3, and

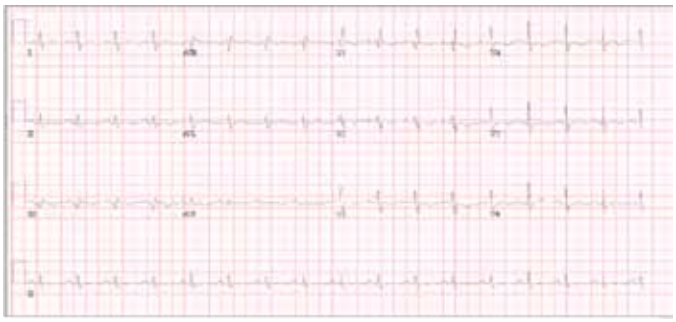


Figure 1: Electrocardiography at presentation, S1Q3T3 pattern and symmetric T wave inversion in anterior precordial leads indicating right ventricular strain.

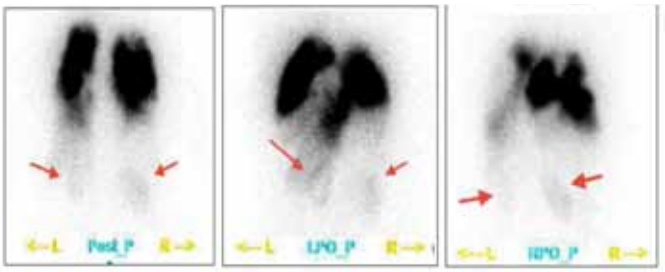


Figure 2: Perfusion images of V/Q scan showing high probability for pulmonary embolism, but also show an unexpected radiotracer uptake in kidneys.

also maintained on hydroxyurea 1000 mg/500 mg on alternate days. She continued to improve clinically and was eventually discharged home. The decision was not to close her PFO and resume therapeutic anticoagulation. The patient was done well on medical therapy, symptom free at three-year follow-up

DISCUSSION

Essential thrombocytosis (ET) is a rare myeloproliferative disorder characterized by megakaryocytic lineage expansion leading to sustained thrombocythemia and increased risk of bleeding and thrombosis. Essential thrombocytosis affects 0.5–2.5/100,000 patients with higher predisposition in females [1]. Thrombosis can involve any vascular bed and is associated with a higher morbidity and mortality than bleeding, which is typically limited to the skin and mucus membranes [1]. Deep venous thrombosis and pulmonary

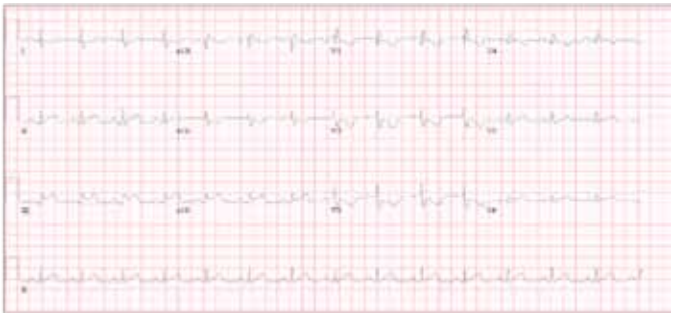


Figure 3: Electrocardiography after eight hours of admission showing ST-segment elevation in II, III and aVF indicating inferior injury with ST-segment depression in V2-V3 pointing to posterior extension.

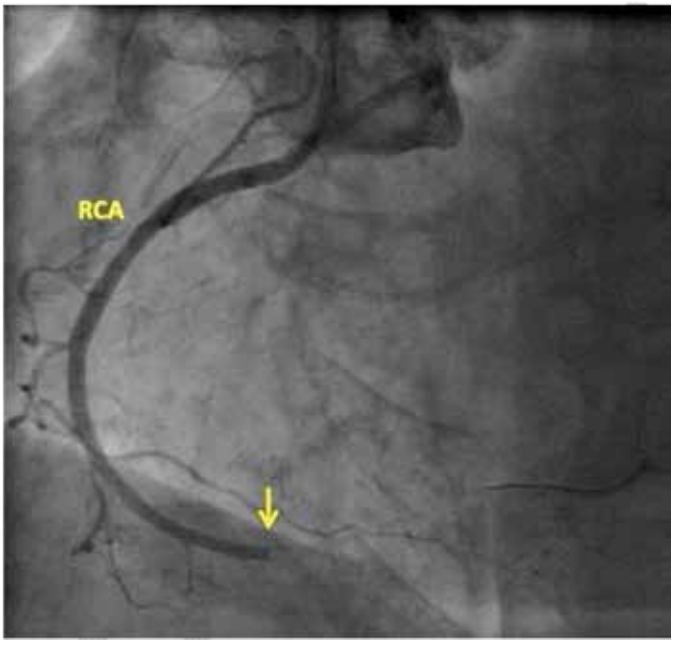


Figure 4: Left anterior oblique view of right coronary artery angiogram showing a complete occlusion at mid-segment with TIMI-0 flows.

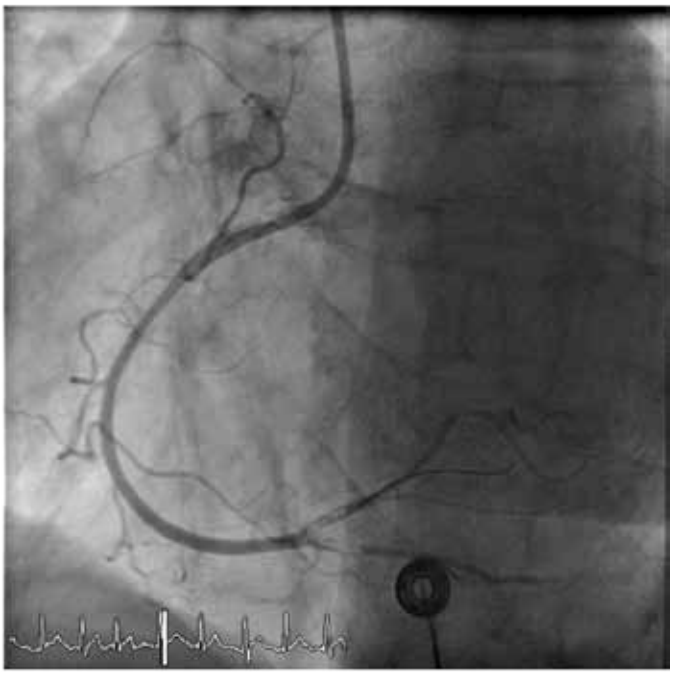


Figure 5: Left anterior oblique view of the right coronary artery angiogram post-thrombectomy with distal embolization to posterior lateral and posterior descending artery. Note the smooth lining within the right coronary artery.

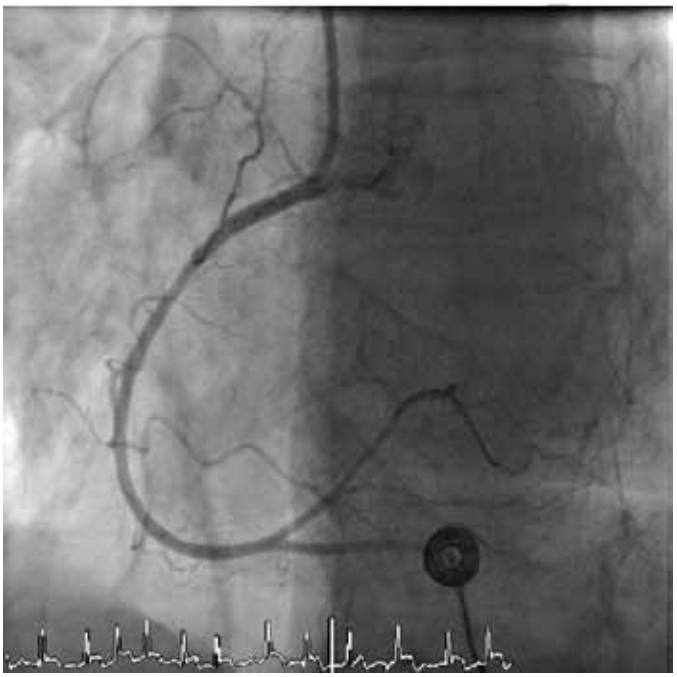


Figure 6: Left anterior oblique view of the right coronary artery after thrombectomy, IIb/IIIa and heparin showing no residual clot in right coronary artery, posterior descending artery or posterior-lateral artery.

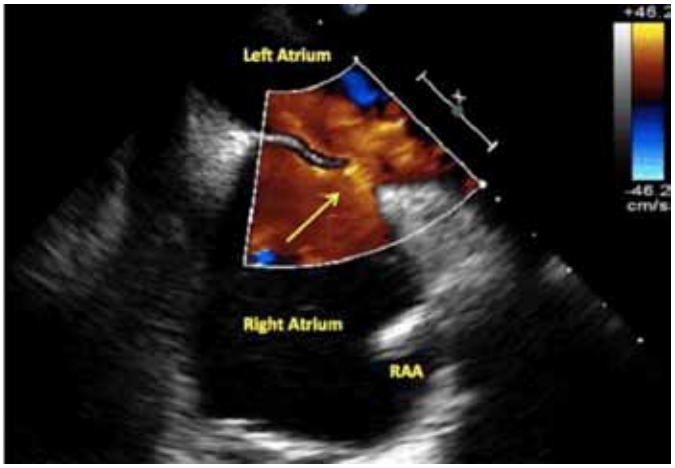


Figure 7: Transesophageal echo bi-caval view with color flow Doppler showing right to left shunt.

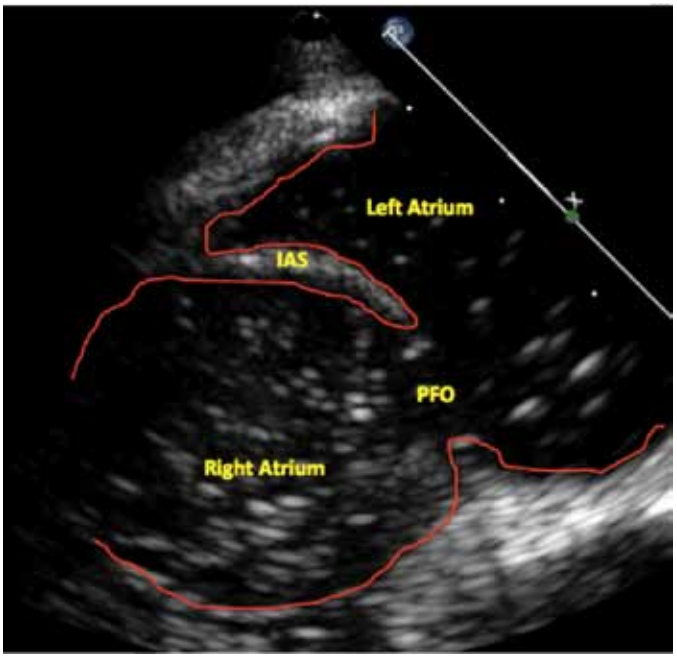


Figure 8: Transesophageal echo bi-caval view with bubble study showing immediate left atrial opacification through the patent foramen ovale.

embolism are the most common thrombotic manifestation [12]. The incidence of ET related thrombotic events is 15–26.4% and in western studies [3]. Factors predisposing to thrombosis are age >60, previous thrombotic event, higher cardiac risk factors, and leukocytosis >15,000 [1, 13]. Platelet counts do not predict thrombotic events, as 10–20% of severe thrombotic complications occur in patients with platelets counts of $<60x10^9/L$ [14, 15]. Our case also represents an example of life-threatening thrombosis with normal platelets counts.

Typical ECG changes in setting of pulmonary embolism include sinus tachycardia, S1Q3/S1Q3T3 pattern, right axis deviation, transient complete or incomplete right bundle branch block, and T-wave inversions in the right pericardial lead. All of these findings, except sinus tachycardia were seen on our first ECG. The patients ECG also had symmetrical T-Wave inversion in leads V1–V4, which is a marker for worse prognosis in hospitalized patients with pulmonary embolism [16]. ST-segment elevation has been described in the setting of pulmonary embolism [17, 18]. It is presumed to be secondary to pressure overload and subendocardial ischemia involving the right ventricle, but paradoxical embolism with subsequent coronary occlusion has also been postulated [17, 19]. STEMI is a rare but well described complication of ET both as an initial manifestation and during follow-up. Approximately, 20 cases of STEMI occurring in the setting of ET have been described. Left main and left anterior descending (LAD) occlusions represent the majority of cases [8, 11] and only five cases have right coronary artery (RCA) involvement [5–7, 20, 21], and no isolated circumflex involvement has been described. To our knowledge, our case is the first published report of RCA occlusion secondary to paradoxical coronary embolism in a patient with ET and massive bilateral pulmonary embolism.

As per criteria suggested by Johnson et al., a paradoxical embolism (PDE) can be presumed when there is evidence of arterial embolization in the absence of a source in the left heart, or when there is a venous source of embolism and an abnormal communication between the venous and the arterial circulation is demonstrated [22]. All three criteria are applicable to our case [22].

Most cases of paradoxical embolism are associated with PFO, while ASD, pulmonary AV malformation, or VSD have been described less frequently [23]. Paradoxical embolism to the coronary tree is rare and constitutes 5–10% of all paradoxical emboli [22]. Paradoxical emboli are further supported if pressure gradients support right-to-left shunting. Pulmonary embolism is associated with 60% of the cases of paradoxical embolism in patients with a PFO and right-to-left shunting [23].

We decided to treat the coronary emboli with manual aspiration and intracoronary abciximab without stenting given the absence of residual disease post-aspiration. Manual thrombectomy has been shown to be beneficial in patients with STEMI in the setting of primary PCI [24]. Murhtay et al. described a successful aspiration of LAD emboli using Export® catheter with eptifibatide infusion in treating PDE resulting from an underlying DVT [22] Kujime et al. described successful use of Thrombuster® aspiration catheter in treating paradoxical RCA clot [25]. Wilson et al. successfully used Export® catheter in conjunction with heparin and eptifibatide in treating an occluded obtuse marginal coronary embolus [26]. Glycoprotein IIb/IIIa infusion post-catheterization has been used in conjunction with thrombectomy to treat high thrombus burden in ET related coronary emboli. Murthay et al. described the use of eptifibatide infusion for 18 hours post-procedure in a similar case [22]. Michaels et al. described the utility of abciximab as an effective sole therapy for decreasing the thrombus burden. In a 50-year-old male with ET and STEMI, angiogram showed extensive clot to left coronary system and total occlusion

of RCA. He was successfully treated with thrombectomy to the RCA alone with abciximab infusion for 24 hours [6].

We decided not to close the patient's PFO. The thrombotic potential of the closure device may have negated any potential benefit from closure. Three relatively recent randomized controlled trails, CLOSURE I [27], PC [28], and PROSPECT [29], failed to prove any benefit of percutaneous closure over medical therapy in patients with paradoxical embolism secondary to PFO. Also a recent meta-analysis by Ntaios et al. showed the same results [30]. Our patient is unique, as her paradoxical embolic event was a STEMI. She has remained stable with rigorous maintenance of therapeutic anticoagulation with warfarin.

CONCLUSION

Essential thrombocytosis is a disorder of dysfunctional platelet overproduction leading a high risk of catastrophic thrombotic events even with normal platelets count. This case shows a rare presentation of paradoxical coronary embolism through a patent foramen ovale in patients with massive pulmonary embolism. Coronary embolism itself is rare, and should always be considered in venous thromboembolism patients with cardiac deterioration. There is no clear treatment consensus in such patients, but combination therapy of thrombectomy and a GPIIb/IIIa inhibitor in the acute setting with chronic anticoagulation was successfully employed in our case. Patent foramen ovale closure may be considered to prevent further embolism, however, our patient did well conservatively, with anticoagulation only.

****

Author Contributions

Fahad S. Almehmadi – Conception and design, Drafting the article, Critical revision of the article, Final approval of the version to be published
Albayda M. Mehdar – Conception and design, Drafting the article, Critical revision of the article, Final approval of the version to be published
Kumar Sridhar – Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Patrick Teefy – Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of 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.

REFERENCES

1. Briere JB. Essential thrombocythemia. Orphanet J Rare Dis 2007 Jan 8;2:3.
2. Bildirici U, Celikyurt U, Ural E. Essential thrombocythemia: A case of acute st-segment elevation myocardial infarction in a young female. Clin Cardiol 2009 Feb;32(2):104–5.
3. Cheng CW, Hung MJ. Coronary spasm-related acute myocardial infarction in a patient with essential thrombocythemia. World J Cardiol 2011 Aug 26;3(8):278–80.
4. Daya SK, Gowda RM, Landis WA, Khan IA. Essential thrombocythemia-related acute st-segment elevation myocardial infarction. A case report and literature review. Angiology 2004 May-Jun;55(3):319–23.
5. Hamada Y, Matsuda Y, Fujii B, et al. Multiple coronary thrombosis in a patient with thrombocytosis. Clin Cardiol 1989 Dec;12(12):723–4.
6. Michaels AD, Whisenant B, MacGregor JS. Multivessel coronary thrombosis treated with abciximab (reopro) in a patient with essential thrombocythemia. Clin Cardiol 1998 Feb;21(2):134–8.
7. Mizuta E, Takeda S, Sasaki N, et al. Acute myocardial infarction in a patient with essential thrombocythemia: Successful treatment with percutaneous transluminal coronary recanalization. Circ J 2005 Aug;69(8):1000–2.
8. Ozben B, Ekmekci A, Bugra Z, Umman S, Meric M. Multiple coronary thrombosis and stent implantation to the subtotally occluded right renal artery in a patient with essential thrombocytosis: A case report with review. J Thromb Thrombolysis 2006 Aug;22(1):79–84.
9. Singla A, Jagasia D, Garg M, Lowry PA, Stapleton D. Acute st-segment elevation myocardial infarction: A rare initial presentation of previously undiagnosed essentialthrombocythemia. Platelets 2012;23(6):463–6.
10. Turgut T, Harjai KJ, Edupuganti R, et al. Acute coronary occlusion and in-stent thrombosis in a patient with essential thrombocythemia. Cathet Cardiovasc Diagn 1998 Dec;45(4):428–33.
11. Watanabe T, Fujinaga H, Ikeda Y, et al. Acute myocardial infarction in a patient with essential thrombocythemia who underwent successful stenting--a case report. Angiology 2005 Nov-Dec;56(6):771–4.
12. Mitus AJ, Schafer AI. Thrombocytosis and thrombocythemia. Hematol Oncol Clin North Am 1990 Feb;4(1):157–78.
13. Wolanskyj AP, Schwager SM, McClure RF, Larson DR, Tefferi A. Essential thrombocythemia beyond the first decade: Life expectancy, long-term complication

rates, and prognostic factors. Mayo Clin Proc 2006 Feb;81(2):159–66.
14. Colombi M, Radaelli F, Zocchi L, Maiolo AT. Thrombotic and hemorrhagic complications in essential thrombocythemia. A retrospective study of 103 patients. Cancer 1991 Jun 1;67(11):2926–30.
15. Regev A, Stark P, Blickstein D, Lahav M. Thrombotic complications in essential thrombocythemia with relatively low platelet counts. Am J Hematol 1997 Nov;56(3):168–72.
16. Falterman TJ, Martinez JA, Daberkow D, Weiss LD. Pulmonary embolism with st segment elevation in leads v1 to v4: Case report and review of the literature regardingelectrocardiographic changes in acute pulmonary embolism. J Emerg Med 2001 Oct;21(3):255–61.
17. Cheng TO. Mechanism of st-elevation in precordial leads v(1)-v(4) in acute pulmonary embolism. Int J Cardiol 2009 Aug 21;136(3):251–2.
18. Goslar T, Podbregar M. Acute ecg st-segment elevation mimicking myocardial infarction in a patient with pulmonary embolism. Cardiovasc Ultrasound 2010 Nov 24;8:50.
19. Uchida S, Yamamoto M, Masaoka Y, Mikouchi H, Nishizaki Y. A case of acute pulmonary embolism and acute myocardial infarction with suspected paradoxical embolism after laparoscopic surgery. Heart Vessels 1999;14(4):197–200.
20. Koh KK, Cho SK, Kim SS, Oh BH, Lee YW. Coronary vasospasm, multiple coronary thrombosis, unstable angina and essential thrombocytosis. Int J Cardiol 1993 Sep;41(2):168–70.
21. Saffitz JE, Phillips ER, Temesy-Armos PN, Roberts WC. Thrombocytosis and fatal coronary heart disease. Am J Cardiol 1983 Sep 1;52(5):651–2.
22. Murthy A, Shea M, Karnati PK, El-Hajjar M. Rare case of paradoxical embolism causing myocardial infarction: Successfully aborted by aspiration alone. J Cardiol 2009 Dec;54(3):503–6.
23. Ahmed S, Sadiq A, Siddiqui AK, Borgen E, Mattana J. Paradoxical arterial emboli causing acute limb ischemia in a patient with essential thrombocytosis. Am J Med Sci 2003 Sep;326(3):156–8.
24. Burzotta F, De Vita M, Gu YL, et al. Clinical impact of thrombectomy in acute st-elevation myocardial infarction: An individual patient-data pooled analysis of 11 trials. Eur Heart J 2009 Sep;30(18):2193–203.
25. Kujime S, Hara H, Enomoto Y, et al. A case of paradoxical embolic st-segment elevation myocardial infarction triggered by sleep apnea. Intern Med 2012;51(14):1851–5.
26. Wilson AM, Ardehali R, Brinton TJ, Yeung AC, Vagelos R. Successful removal of a paradoxical coronary embolus using an aspiration catheter. Nat Clin Pract Cardiovasc Med 2006 Nov;3(11):633–6.
27. Furlan AJ, Reisman M, Massaro J, et al. Closure or medical therapy for cryptogenic stroke with patent foramen ovale. N Engl J Med 2012 Mar 15;366(11):991–9.
28. Meier B, Kalesan B, Mattle HP, et al. Percutaneous closure of patent foramen ovale in cryptogenic embolism. N Engl J Med 2013 Mar 21;368(12):1083–91.
29. Carroll JD, Saver JL, Thaler DE, et al. Closure of patent foramen ovale versus medical therapy after cryptogenic stroke. N Engl J Med 2013 Mar 21;368(12):1092–100.
30. Ntaios G, Papavasileiou V, Makaritsis K, Michel P. PFO closure vs. Medical therapy in cryptogenic stroke or transient ischemic attack: A systematic review and meta-analysis. Int J Cardiol 2013 Oct 30;169(2):101–5.

ABOUT THE AUTHORS

Article citation: Almehmadi FS, Mehdar AM, Sridhar K, Teefy P. ST-elevation myocardial infarction secondary to paradoxical coronary emboli in a patient with massive pulmonary embolism and essential thrombocytosis: A case report. Int J Case Rep Images 2015;6(3):149–155.

Fahad S. Almehmadi is a second year Adult Cardiology Resident at University of Toronto in Canada. He earned his medical degree with distinction from Umm Alqura University in Makkah, Saudi Arabia. He intends to pursue an academic career in cardiology and cardiac electrophysiology.

Albayda M. Mehdar is a second year Internal Medicine Resident at University of Toronto in Canada. She earned her medical degree with distinction from King Abdulaziz University in Jeddah, Saudi Arabia.

Kumar Sridhar is Associate Professor at Western University – Department of Medicine and University Hospital and Interventional Cardiologist and Director of CCU at London Health Sciences Centre. Dr. Sridhar completed his medical training at Western University, London, Ontario and his interventional cardiology fellowship at Western University and Thomas Jefferson University Hospital in Philadelphia, Pennsylvania. He has a number of publications in peer-reviewed journals, book chapters and research interests specific to novel treatment coronary interventions and treatment strategies.

Patrick Teefy is Director of the Cardiac Catheterization Laboratories at London Health Sciences Centre, London, Ontario Canada and Associate Professor, University of Western Ontario. His research interests include interventional cardiology and trans-catheter aortic valve implantation.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Short left main coronary artery causing dynamic left ventricular outflow tract obstruction and new onset left bundle branch block

Geoffrey Chibuzor Nwuruku, Godsent Chichebem Isiguzo, Joel Tamayo Brooks, Ernest Madu

ABSTRACT

Introduction: Left bundle branch block can be seen in conditions like aortic stenosis, extensive coronary artery disease, primary disease of the cardiac electrical conduction system, dilated cardiomyopathy, Lyme disease; it is also associated with short left main coronary artery and dynamic left ventricular outflow tract obstruction. While the former is caused by the shearing force on septal branches of the left anterior descending artery, the latter is related to either Venturi effect in hypertrophic cardiomyopathy or effect of systolic anterior motion caused by abnormal geometric relationship of papillary muscle and the mitral apparatus. Case Report: A 50-year-old male, former smoker with a history of dyslipidemia, presented with shortness of breath and exertional chest pain. After clinic review, he was thought to have stable angina, electrocardiogram, and echocardiography were normal and lifestyle modification was advised. Patient had some improvement, but represented seven months later with worsening of symptoms. Repeat electrocardiograph showed a new onset left bundle branch block, with short left main coronary artery on coronary angiogram and dynamic left ventricular outflow tract obstruction in stress echocardiography, with a gradient of 40 mmHg. Conclusion: Short left main coronary artery is a rare cause of left bundle branch block, and it should be considered when evaluating patients with new onset left bundle branch block without hypertrophic cardiomyopathy.

Keywords: Erectile dysfunction, Hypertrophic cardiomyopathy, Left bundle branch block (LBBB), Left main coronary artery (LMCA), Left ventricular outflow tract (LVOT), Lyme disease, Microvascular dysfunction (MVD), Septal dyssynchrony, Short left main coronary

Geoffrey Chibuzor Nwuruku[1], Godsent Chichebem Isiguzo[2], Joel Tamayo Brooks[3], Ernest Madu[4]

Affiliations: [1]MD, Registered Cardiologist and Family Physician, DOCS VIP Clinic, Enugu, Nigeria; [2]MBBS, FWACP, Consultant Cardiologist, DOCS VIP Clinic, Enugu, Nigeria; [3]MD, Interventional Cardiologist, Heart institute of Caribbean (HIC), Kingston, Jamaica; [4]MD, FACP, FACC, FRCP Edin, Professor of Cardiology, Heart institute of Caribbean (HIC), Kingston, Jamaica, and Director, DOCS VIP Clinic Enugu, Nigeria.

Corresponding Author: Dr. Godsent Chichebem Isiguzo, Cardiologist, DOCS VIP Clinic Enugu Nigeria, Consultant Physician/Cardiologist, Federal Teaching Hospital, PMB 102, Abakaliki, Ebonyi State, Nigeria; Ph: +2348035419835

Email: isiguzogodsent@yahoo.com

Received: 02 December 2014
Accepted: 16 January 2015
Published: 01 March 2015

How to cite this article

Nwuruku GC, Isiguzo GC, Brooks JT, Madu E. Short left main coronary artery causing dynamic left ventricular outflow tract obstruction and new onset left bundle branch block. Int J Case Rep Images 2015;6(3):156–160.

doi:10.5348/ijcri-201529-CR-10490

INTRODUCTION

Although left bundle branch block (LBBB) is a common finding in conditions such as aortic stenosis, extensive coronary artery disease, primary disease of the cardiac electrical conduction system, dilated cardiomyopathy, Lyme disease; it can also be associated with short left main coronary artery (LMCA) and a dynamic left ventricular

outflow tract (LVOT) obstruction [1]. While the former is caused by the shearing force on the septal branches of the left anterior descending artery, the latter is related to these major possibilities among others: the Venturi effect in hypertrophic cardiomyopathy (HCM) as well as systolic anterior motion of the mitral valve (SAM) generated largely by drag effect, that is hydrodynamic pushing force of flow directly on the leaflets, and the SAM caused by abnormal geometric relationship of papillary muscle and the mitral apparatus. This abnormal geometry is in part as a result of non-coordinated contraction (mechanical dyssynchrony) of interventricular septum and left ventricular posterior or posterolateral walls which lead to the displacement of the papillary muscles and the chordae tendineae (chordae tendineae SAM).

CASE REPORT

A 50-year-old male with a history of dyslipidemia and erectile dysfunction, stopped smoking 10 years ago, not known to be hypertensive or have diabetes mellitus and no significant family history. He presented with shortness of breath and exertional chest pain at variable threshold. Physical examination was negative while blood workup showed total cholesterol (223 mg/dL), LDL (165 mg/dL) and the remainder of the blood test results were normal. Various other tests conducted including ECG (Figure 1A), ECHO/Doppler and Stress ECHO were not significant while 24-hour Holter monitoring revealed sinus arrhythmias with a solitary ventricular ectopic beat and rare supraventricular ectopic beat with a couplet. A treatment plan that included lifestyle modification was instituted for stable angina. The patient noted improvement but less than a year later the same symptoms recurred and even became more severe. He presented to the clinic again and a repeat ECG revealed a new onset BBB (Figure 1B) that was not seen in the previous rest ECG before stress test (Figure 1A). He was suspected to have a progressive ischemia, and repeat echocardiography showed septal dyssynchrony with good left ventricular systolic function. A coronary angiogram was then done, showing a short left main coronary artery (Figure 2), while a stress ECHO revealed a significant dynamic LVOT obstruction (chordae tendineae SAM) with a gradient of 40.9 mmHg (Figure 3) at the end of a third dose with symptoms that prompted the termination of the test. He was discharged on beta blocker (bisoprolol 5 mg daily) management and has reported great improvement in his functional capacity.

DISCUSSION

This is a case of a 50-year-old male patient with a new onset LBBB associated with systolic anterior motion of the mitral valve (chordae tendineae SAM). This finding triggered a LVOT obstruction with a gradient of 40.9 mmHg on a stress echo. The LBBB was spontaneous and thought to have resulted from progressive ischemia as previous ECG and resting ECG before stress test did not reveal this. The echocardiographic findings were related to shortness of breath and angina (on a mild activity) noticed on the patient that was also reproduced during stress test. Studies have shown longstanding LVOT obstruction (basal gradient, ≥30 mmHg) as a strong determinant of HCM-related progressive heart failure symptoms and cardiovascular death but however, showed only a weak relationship evident between outflow obstruction and specifically the risk for sudden cardiac death (usually in patients without significant heart failure symptoms) [2]. We did not find any study that demonstrated this relationship in a patient with SAM caused purely by abnormal geometry of the mitral valve apparatus. A study by Rodriguez Rodrigo et al. demonstrates microvascular dysfunction (MVD) as the cause of deterioration of left ventricular function in patients with isolated LBBB [3]. The study only considered the functional influence of LBBB on the coronary arteries; in contrast our report shows the prognostic influence of the coronary arteries anatomy in the pathogenesis of a spontaneous LBBB of which MVD plays a vital role.

Short LMCA has been shown to be a prelude to development of coronary artery disease, and a cause of LBBB, with the later known as a marker of slowly progressing ischemic or non-ischemic cardiac disease and increase cardiovascular disease related death risk, and lower survival rate [4], hence the need for an exhaustive evaluation. In the index patient, finding of new onset

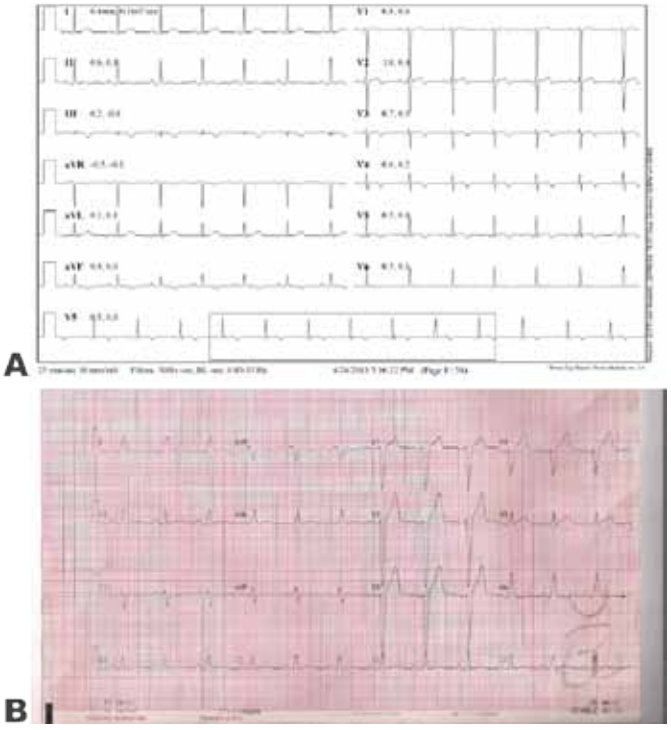


Figure 1: (A) Electrocardiograph of the patient at initial presentation (normal), and (B) After onset of new symptoms showing left bundle branch block.

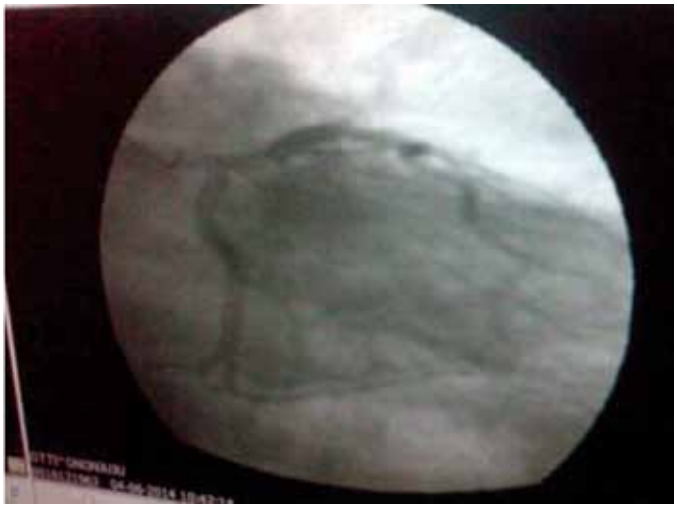


Figure 2: Coronary angiogram showing short left main coronary artery.

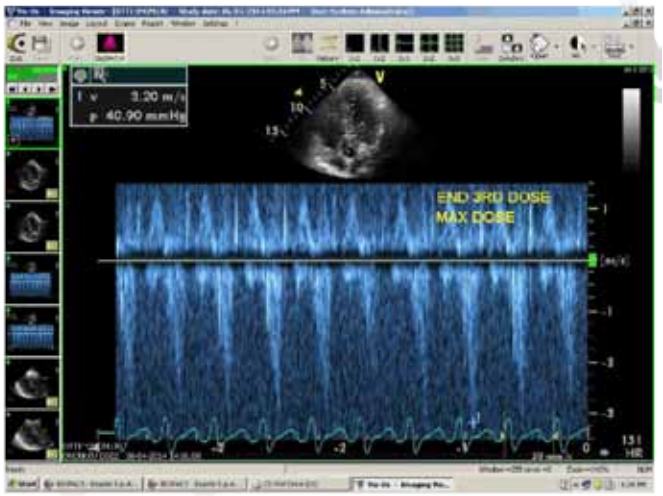


Figure 3: Stress echocardiogram showing a dynamic left ventricular outflow tract obstruction with a gradient of 40.9 mmHg.

LBBB, without any apparent trigger associated with progressive symptoms was a source of concern and lead to our further evaluation with a resting echocardiogram which revealed a septal dyssynchrony while a stress echo showed SAM of the mitral valve with LVOT obstruction demonstrated by a gradient of 40.9 mmHg. He was thereafter studied on coronary angiogram and a short LMCA was observed with no significant coronary artery obstruction.

There are divergent results on the role of short LMCA in the pathogenesis of LBBB, most of which are not recent. A study by Lewis et al. in 1969 found that the length of the left main coronary artery (LMCA) was less than 6 mm in all but one of 12 patients with LBBB and was longer than 7 mm in a control group of 25 patients [1]. This data suggest that the cardiac diseases associated with LBBB were not etiologically important or that they were important only when the LMCA was short. We posited that the association between a short LMCA and LBBB could be explained by greater shearing forces imposed during systole in the short arteries (LMCA). This, in turn might compromise flow through the early septal branches of the left coronary system and thus produce ischemia and fibrosis of the left bundle branch. Ischemia as a possible pathogenetic mechanism producing fibrosis, when it affects conducting system, has been found to be the histologic abnormalities in patients with LBBB [5]. This could explain the new onset conducting system abnormality noticed in this patient after one year of a normal ECG.

In a study based on pathological observations by Gazetopoulos et al. suggested that short LMCA should be considered as a congenital factor predisposing to the development of atherosclerotic coronary artery disease, confirming their earlier finding that length of the LMCA is an anatomic factor that may influence the rate of development of atheromatosis on its branches [6]. But in contrast to the findings, De Mots et al. showed that the length of LMCA in their 13 patients with LBBB was not significantly different compared to the 78 patients in the control group [7].

The LBBB leads to electrical conduction disorder which by the way of mechanical dyssynchrony causes abnormalities in the movement of the mitral apparatus including anterior and inward or central displacement of the papillary muscle and leaflet elongation that can ultimately lead to SAM and left ventricular outflow tract obstruction in patients without HCM.

Several explanations have been adduced for the occurrence of SAM in patients with hypertrophic cardiomyopathy (HCM), notable among which are Venturi effect and anatomic difference in position of papillary muscles and valve leaflets [8, 9]. More recent studies have also attributed the dominant force in the anterior displaced mitral valve leaflets as drag force that is in proportion to velocity of flow in LVOT and to the angle between the anterior mitral leaflet and the direction of flow in the LVOT [10]. Many other factors may play roles in causing patients without HCM to present with LVOT obstruction as seen in the index. These include primary abnormalities of the mitral valve apparatus, like anterior and inward or central displacement of the papillary muscles, leaflet elongation, geometric disarray of the left ventricle caused by LBBB. Short LMCA may also cause progressive microvascular disease, and its clinical consequence depends on the level of obstruction at the LVOT, dyssynchrony impact of LBBB and dynamic state of the patient.

CONCLUSION

Though there are divergent reports and paucity of recent studies regarding the role of a short left main coronary artery in causing left bundle branch block (LBBB), it has been shown that isolated LBBB with microvascular dysfunction is associated with worse

outcome. Therefore, there is need for physicians to evaluate for causes of new onset LBBB in their patients as early discovery and prompt intervention may alter the cause of potential cardiovascular disease mortality and morbidity as seen in the index patient.

****

Author Contributions

Geoffery Chibuzor Nwuruku – Substantial contribution to conception design, Acquisition of data, Drafting of article, Revising it critically for important intellectual content, Final approval of version to be published
Godsent Chichebem Isiguzo – Substantial contribution to conception design, Analysis and interpretation of data, Drafting of article, Revising it critically for important intellectual content, Final approval of version to be published
Joel Tamayo Isiguzo – Substantial contribution to conception design, Acquisition of data, Revising critically for important intellectual content, Final approval of version to be published
Madu Ernest – Substantial contribution to conception design, Acquisition of data, Revising it critically for important intellectual content, Final approval of version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCE

1. Lewis CM, Dagenais GR, Friesinger GC, Ross RS. Coronary Arteriographic Appearances in Patients with Left Bundle-Branch. Circulation 1970 Feb;41(2):299–307.
2. Maron BJ, Maron MS, Wigle ED, Braunwald E. The 50-year history, controversy, and clinical implications of left ventricular outflow tract obstruction in hypertrophic cardiomyopathy from idiopathic hypertrophic subaortic stenosis to hypertrophic cardiomyopathy: from idiopathic hypertrophic subaortic stenosis to hypertrophic cardiomyopathy. J Am Coll Cardiol 2009 Jul 14;54(3):191–200.
3. Rodrigo RFJ, Peralta JM, Arias EV, Barrero AA, Lascorz TSA, Pereira EP, Chaverri AR. Prognostic implication of the coronary microvascular dysfunction in patientswith isolated left bundle branch block. World J Card Dis 2014;4:61–9.
4. Eriksson P, Wilhelmsen L, Rosengren A. Bundle-branch block in middle-aged men: risk of complications and death over 28 years. The Primary Prevention Study in Göteborg, Sweden. Eur Heart J 2005 Nov;26(21):2300–6.
5. Davies M, Harris A. Pathological basis of primary heart block. Br Heart J 1969 Mar;31(2):219–6.
6. Gazetopoulos N, Ioannidis PJ, Karydis C, Lolas C, Kiriakou K, Tountas C. Short left coronary artery trunk as a risk factor in the development of coronary atherosclerosis Pathological study. Br Heart J 1976 Nov;38(11):1160–5.
7. De Mots H, Rösch J, Rahimtoola SH. Coronary Artery Anatomy in Left Bundle Branch Block. Circulation 1973 Sep;48(3):605–8.
8. Lemke R, Kaltenbach M. Systolic anterior movement of the mitral valve and the venturi effect: An in vitro study. Z Kardiol 1987;76 Suppl 3:87–90.
9. Chun EJ, Choi SI, Jin KN, et al. Hypertrophic cardiomyopathy: Assessment with magnetic resonance imaging and multidetector CT. Radiographics 2010 Sep;30(5):1309–28.
10. Sherrid MV, Gunsburg DZ, Moldenhauer S, Pearle G. Systolic anterior motion begins at low left ventricular outflow tract velocity in obstructive hypertrophic cardiomyopathy. J Am Coll Cardiol 2000 Oct;36(4):1344–54.

ABOUT THE AUTHORS

Article citation: Nwuruku GC, Isiguzo GC, Brooks JT, Madu E. Short left main coronary artery causing dynamic left ventricular outflow tract obstruction and new onset left bundle branch block. Int J Case Rep Images 2015;6(3):156–160.

Geoffrey Chibuzor Nwuruku is Registered Cardiologist and family physician in DOCS VIP Clinic, Heart Centre, Enugu, Enugu State, Nigeria. He earned undergraduate degree, Doctor in Medicine from Latin American School of Medicine, Havana, Cuba and postgraduate degree, Specialist in Cardiology from 'Hermanos Ameijeiras' Specialist Hospital Department of Cardiology and Cardiovascular Surgery, Havana, Cuba. He has published one research paper in national academic journal. This research interests include diastolic heart failure, non dipping pattern of blood pressure and target organ damage, the relationship between clinical and echo findings in functional capacity. He intends to pursue a research fellowship with any applicable research institute of good repute in future.
Email: jeffrey_2@live.co.uk

Godsent Chichebem Isiguzo is Consultant Physician, Cardiologist at Medicine department, Federal teaching hospital, Abakaliki, Ebonyi state, Nigeria and Cardiologist at DOCS VIP Clinic, Heart Centre, Enugu, Enugu State, Nigeria. He earned the undergraduate degree (MBBS) from College of Medicine, Abia Stae University, Nigeria and postgraduate degree (FWACP) from West African college of Physician, following Postgraduate training at Jos University teaching hospital, Jos, Plateau state Nigeria. His fellowship thesis was on Pulmonary hypertension and HIV-related cardiac dysfunction. He has published 11 research papers in national and international academic journals. His research interests include: HIV-related heart disease, Pregnancy related heart disease, hypertension, cardiomyopathies and cardiac imaging). He intends to pursue Postdoctoral fellowship in echocardiography and Preventive cardiology in future.
Email: isiguzogodsent@yahoo.com

Joel Tamayo Brooks is Registered Consultant Cardiologist, interventional cardiologist and family physician at the Heart Institute of the Caribbean, Kingston, Jamaica and DOCS VIP Clinic, Heart Centre, Enugu, Nigeria. He earned undergraduate degree, Doctor in Medicine from University of Medicine, Havana, Cuba and postgraduate degree, Specialist in Cardiology from 'Hermanos Ameijeiras' Specialist Hospital Department of Cardiology and Cardiovascular Surgery, Havana, Cuba. Interventional Cardiolgy fellowship, University Complex Hospital of Vigo, Spain. He has published 12 research papers in national and international academic journals and the interventional cardiologist handbook. His research interests include all the interesting topics on the interventional cardiologist field. He intends to pursue a research fellowship with any applicable research institute of good repute in future.
Email: blackhope7869@yahoo.com

Ernest Madu is Professor of Cardiovascular Medicine and Imaging Technology and Chairman of the Heart Institute of the Caribbean. He has published extensively in cardiovascular medicine. His current research interests are in innovative health systems development and technology applications in emerging nations. He has given keynote lectures in many countries and is widely sought after as a global leader in innovative and disruptive healthcare applications.
Email: emadu@caribbeanheart.com

CASE REPORT PEER REVIEWED | OPEN ACCESS

Successful treatment of a patient with obesity, type 2 diabetes and hypertension with the paleolithic ketogenic diet

Csaba Tóth, Zsófia Clemens

ABSTRACT

Introduction: Metabolic syndrome is a major public health problem affecting at least 20% of the world's adult population. Components of the metabolic syndrome include obesity, impaired glucose metabolism, hypertension and altered lipid profile. Currently, medical treatment relies on drugs. A major problem is that patients with long-standing disease are excessively medicated because of an increase in the number of symptoms over time. A few clinical studies indicate that low-carbohydrate diets, including the paleolithic as well as the ketogenic diet, may be beneficial in the treatment of conditions associated with the metabolic syndrome. Case Report: Herein, we present a case of patient with metabolic syndrome successfully treated with the paleolithic ketogenic diet. While on the diet the patient was able to discontinue eight medicines, lost weight, showed a continuous improvement in glucose parameters and her blood pressure normalized. Currently, the patient is on the paleolithic ketogenic diet for 22 months, free of symptoms and side effects. Conclusion: We conclude that the paleolithic ketogenic diet was safe, feasible and effective in the treatment of this patient with metabolic syndrome.

Keywords: Diabetes, Hypertension, Metabolic syndrome, Obesity, Paleolithic diet, Paleolithic ketogenic diet

Csaba Tóth[1], Zsófia Clemens[2]

Affiliations: [1]MD, Medical Director, Paleomedicina Hungary Ltd. Evolutionary Medicine Working Group, Budapest, Hungary; [2]PhD, Senior Researcher, Neurological Department, University of Pécs, Pécs, Hungary, Paleomedicina Hungary Ltd. Evolutionary Medicine Working Group, Budapest, Hungary.

Corresponding Author: Zsófia Clemens, Department of Neurology, University of Pécs, Rét u. 2, H-7623, Pécs, Hungary; Ph: 003672535900; Email: clemenszsofia@gmail.com

Received: 11 December 2014
Accepted: 13 January 2015
Published: 1 March 2015

How to cite this article

Tóth C, Clemens Z. Successful treatment of a patient with obesity, type 2 diabetes and hypertension with the paleolithic ketogenic diet. Int J Case Rep Images 2015;6(3):161–167.

doi:10.5348/ijcri-201530-CR-10491

INTRODUCTION

Obesity, type 2 diabetes and hypertension represent a major health problem. These conditions are components of the metabolic syndrome affecting every fifth adult worldwide [1]. Although it is now acknowledged that type 2 diabetes and hypertension mostly result from lifestyle factors but medical treatment continues to rely on drugs [2]. Symptoms of the metabolic syndrome typically show up in mid-life but the number of co-morbidities are increasing through later years. Typically, symptoms are controlled with an increasing number of drugs. In parallel side effects are also increasing and are usually controlled with additional medications. It is a vicious circle. Currently, a major proportion of elderly in the western world is overmedicated [3]. In sharp contrast with this diseases of civilization are virtually absent in contemporary hunter-gatherer societies [4]. It is suggested that chronic diseases of civilization result from an evolutionary mismatch between our ancient and current diet [5]. It was also suggested that a return to an evolutionary adapted diet may be beneficial for

health [5]. The paleolithic diet has previously been shown to confer metabolic benefits in healthy as well as in patients with metabolic syndrome [6–10]. Voegtlin, first proponent of the human evolutionary diet, suggested an animal fat-meat based diet as being evolutionary adapted [11]. Recently, we reported successful treatment of patients with epilepsy [12] and type 1 diabetes [13] with the paleolithic ketogenic diet. The diet we refer to as the paleolithic ketogenic diet is close to the meat-fat based diet originally proposed by Voegtlin [11]. Herein, we report on a patient with obesity, type 2 diabetes and hypertension whose excessive medication could be discontinued and clinical parameters associated with the metabolic syndrome markedly improved.

CASE REPORT

Previous medical history

The patient's previous medical history included gallbladder surgery in 1987 due to choledocholithiasis and chronic cholecystitis. Hyperglycemia and impaired glucose tolerance was first demonstrated on 24 February 2004. Due to bloody stool Weber test was performed on 25 August 2006 which showed positivity. Therefore, colonoscopy was performed on 19 September 2006 which indicated a 2-cm polyp in the sigmoid colon. Irrigoscopy performed on 25 September 2006 confirmed the above mentioned polyp and also indicated dilation of the colon as well as sigmoid diverticulosis. The polyp was removed on 6 November 2006. Histopathology from this specimen showed tubulovillous adenocarcinoma in polyp (Grade 1). Resection margins were negative. Gastroscopy performed on 21 September 2006 indicated reflux esophagitis and erosive gastritis. Gastroscopy follow-up, three years later, on 26 January 2009 indicated reflux esophagitis but no evidence of gastritis. A next gastroscopy follow-up on 26 January 2011, showed reflux esophagitis and a 5-mm gastric polyp which was removed. Histopathology from this sample showed no abnormalities. A subsequent gastroscopy performed on 14 March 2013 showed reflux esophagitis. Colonoscopy follow-up on 26 January 2007 demonstrated dilation of colon. Subsequent colonoscopy examinations on 27 August 2007, 25 August 2008, 31 August 2009 and 15 April 2013 were able to examine the colon until the transversus lienalis and showed no alterations. A computed tomography (CT) colonography on 27 April 2011 was negative too.

On 10 December 2009, she experienced pressing pain behind the sternum. Echocardiography on 14 December 2009 showed impaired left ventricular diastolic function as well as aortic and mitral insufficiency.

A routine ophthalmological examination on 01 June 2006 showed crossing phenomenon and hyperemic macula, ocular signs of hypertonia and diabetes. Follow-up ophthalmology examination on 20 May 2009 and on 21 May 2010 showed angiopathy while follow-up examinations on 18 May 2012 and 30 July 2013 indicated angiopathy as well as retinopathy.

Medications

Due to elevated blood glucose parameters and high blood pressure from 01 March 2005 onwards the patient was treated with acarbose, ramipril and hydrochlorothiazide. On 02 November 2006 pantoprazole was added because of reflux esophagitis revealed by gastroscopy. Due to increases in blood glucose from 23 August 2007, the patient was also taking metformin. On 14 December 2009 bisoprolol was added because of high blood pressure. Following the cardiological event acetylsalicylic acid was prescribed on 15 April 2010. From 28 October 2010, the patient was prescribed amlodipine due to high blood pressure. The number and the names (active substance) of the medications taken through the course of the disease and the associated 90-min glucose are indicated in Figure 1.

Paleolithic ketogenic diet

We first met the 65-year-old female on 30 January 2013. She was overweight (BMI 37.1, height 160 cm, weight 95 kg), had high blood sugar and frequent high blood pressure spikes despite antidiabetic and antihypertensive medication. At this time her systolic blood pressure was between 130 and 160 mmHg and diastolic blood pressure between 70 and 85 mmHg. Her fasting glucose level was between 144 and 162 mg/dL. Altogether she was taking eight medicines. The patient was motivated in weight loss and in the reduction of her medication. She was advised to start the paleolithic ketogenic diet. She was suggested a diet consisting of at least 70% animal-based food with a fat : protein ratio of at least 2:1. Fat and red meats were preferred over lean meats. Offals meat were encouraged to ensure adequate intake of vitamins. No more than 30% of the diet was suggested as plant-derived food including root vegetables and small amounts of fruit. Dairy, cereals, grains, legumes, solanaceous vegetables, plant oils (including coconut oil), artificial sweeteners and foods with additives were not allowed. In addition to the paleolithic ketogenic diet, she was taking 2000 IU of vitamin D3 for four months then it was stopped. No other vitamin or mineral supplements were used. Typical foods the patient was eating include broth, stew, fried bacon, beef steak, stewed calf liver, braised pork marrow, greaves. Vegetables consumed as garnish were limited and typically included root vegetables, onion and cabbage.

Upon diet commencement (on the third day of the diet) all medications were stopped promptly except for bisoprolol which was discontinued within two weeks. The patient was controlled tightly during the first weeks of the diet. In case of high blood pressure, she was advised to take captopril, a short-acting antihypertensive. Laboratory workup was performed regularly (eight times during 22 months on the diet) in order to control adherence to the diet and to give feedback to the patient (Table 1). All urinary analyses were positive for ketones. While on the paleolithic ketogenic diet blood glucose

Table 1: Laboratory parameters between 2004 and 2014. The red line indicates onset of the paleolithic ketogenic diet. Note that in spite of the withdrawal of eight medications her blood glucose parameters and triglyceride decreased. Dashes indicate that a given parameter was not measured.

		Glucose (mg/dL)	90-min glucose (mg/dL)	HgA1c (%)	Triglyceride (mmol/L)	Cholesterol (mmol/L)	HDL (mmol/L)	LDL (mmol/L)	Uric acid (mmol/L)	ESR (mm/h)	Urinary ketone	Number of medications
2004	24 Feb	133	165	–	–	–	–	–	–	–	–	0
	16 Jun	114	–	5.7	–	4.07	–	–	–	–	negative	0
	30 Sep	126	–	–	1.16	6.47	–	–	209	12	–	0
2005	1 Mar	137	154	6	1.48	5.63	–	3.88	159	19	negative	0
	28 Jul	126	126	6.2	1.25	4.08	1.2	2.31	268	37	negative	3
2006	11 Jul	139	143	6.3	0.77	5.22	1.3	3.5	179	17	positive	3
	02 Nov	149	92	6.2	1.72	5.42	1.08	3.56	241	17	negative	3
2007	13 Feb	130	117	6.12	1.4	6.38	1.62	4.12	277		negative	4
	17 May	126	–	5.6	1.18	5.56	0.95	4.07	324	23	negative	4
	23 Aug	150	125	–	1.48	6.02	1.15	4.2	258	32	negative	4
	29 Nov	116	81	5.03	1.64	5.61	1.03	–	–	–	–	5
2008	11 Mar	129	120	5.6	2.38	5.95	1.23	–	284	21	negative	5
	24 Jun	128	102	6.2	2.37	6.9	1.73	4.09	268	25	negative	5
	09 Oct	106	–	6	2.06	6.1	1.55	3.61	222	34	–	5
2009	15 Jan	141	121	6.3	3.6	6.1	–	3.06	293	35	negative	5
	09 Apr	105	110	–	1.56	5.1	1.1	3.29	348	30	negative	5
	09 Jul	105	90	6	1.97	5.6	–	3.51	–	24	negative	5
	13 Oct	106	99	5.5	1.76	5.2	1.51	2.89	–	29	negative	5
2010	14 Jan	115	103	6.3	1.78	5.2	1.23	3.16	–	19	negative	6
	15 Apr	110	97	6.3	1.48	5	–	3.15	249	15	negative	6
	27 Jul	112	97	6.76	1.34	5.6	–	3.54	207	24	negative	7
	28 Oct	119	115	6	1.62	6.1	1.16	4.2	–	–	–	7
2011	01 Feb	133	146	6.6	2.31	6.4	1.26	4.09	378	20	negative	8
	05 May	114	117	6.2	2.53	5.1	1.14	2.81	–	–	–	8
	16 Aug	132	146	6.7	2.01	5.8	1.26	3.63	305	26	negative	8
	17 Nov	124	119	6.9	1.99	5.9	1.29	3.71	–	–	negative	8
2012	21 Feb	151	173	7.2	2.12	5.3	1.03	–	419	45	negative	8
	12 Mar	123	–	–	–	–	–	–	–	25	–	8
	22 May	124	164	7	1.76	5.6	1.19	3.61	300	23	negative	8
	06 Sep	150	173	7.1	2.88	6.7	1.28	4.11	–	–	–	8
	18 Dec	159	137	–	1.49	6	1.2	4.12	217	21	negative	8
2013	26 Mar	128	114	6.4	1.14	5.3	1.29	3.49	–	–	–	0
	13 Jun	139	123	6.8	0.77	4.5	1.22	2.93	282	8	positive	0
	10 Sep	139	123	6.9	1.09	4.8	1.32	2.98	238	18	positive	0
	09 Oct	110	–	–	–	–	–	–	–	–	–	0
	19 Dec	108	114	7	1	4.51	1.52	2.54	274	20	positive	0
2014	05 Mar	118	–	–	0.69	5.12	–	–	297	–	positive	0
	10 Jun	121	108	6.8	0.96	4.8	1.23	3.13	248	15	positive	0
	04 Dec	108	99	6.5	1.6	5.7	1.19	3.78	274	9	positive	0

Abbreviations: HgA1c glycated hemoglobin, HDL high density lipoprotein, LDL low density lipoprotein, ESR erythrocyte sedimentation rate

level, 90-min glucose on the glucose tolerance test, HgA1c and total cholesterol levels decreased as compared to previous measurements. Triglyceride levels also dropped markedly as well as inflammatory markers including erythrocyte sedimentation rate. A statistical analysis (t-test) comparing laboratory measures during the two years of the paleolithic ketogenic diet and during the previous nine years on a normal diet revealed a significant decrease for HgA1c (p=0.02), cholesterol (p=0.01) and triglyceride (p=0.003). Typically, the patient had two meals a day. She tracked glucose levels daily both preprandially and postprandially before and after the first meal of the day. The patient reported to strictly adhere to the diet insofar not consuming non-paleolithic food at all. At the same time she admitted having some difficulties with fruit restriction and reported that her spikes in blood sugar (Figure 2) and blood pressure to be associated with these events of excess fruit intake. Nevertheless home monitoring of glucose showed a clear decreasing trend both preprandially and postprandially between February 2013 and June 2014 (Figure 2). Then due to the normalization of blood glucose she measured blood glucose levels only occasionally. While on the diet she also reported a decreasing tendency in the frequency of high blood pressure spikes. At the time of writing this case report, she had no high blood pressure spike for six months. Despite no vitamin D supplementation, her winter time vitamin D level (on 05 March 2014) was in the normal range (85 nmol/L). The patient did not perform exercise while on the paleolithic diet.

Currently, she is on the diet for 22 months. While on the diet her weight changed from 95 kg to 81 kg and she is still losing weight. Her BMI changed from 37.1 to 31.6. She reported increased physical fitness and to be free of symptoms. The patient gave written informed consent for publication of her case.

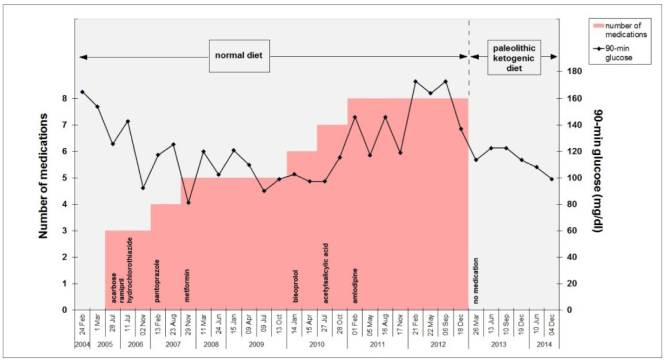


Figure 1: Time course of 90-min glucose level in the glucose tolerance test and the number as well as the names of the medications (active substance) between 2004 and 2014. Note that 90-min glucose levels first tended to decrease with the use of oral antidiabetics. Then from 2011 90-min glucose levels increased again. We attribute decreased glucose tolerance to the use of bisoprolol and amlodipine drugs known for their diabetogenic effect. From January 2013 the patient shifted toward the paleolithic ketogenic diet and was able to discontinue all eight medications. In parallel 90-min glucose levels returned to normal.

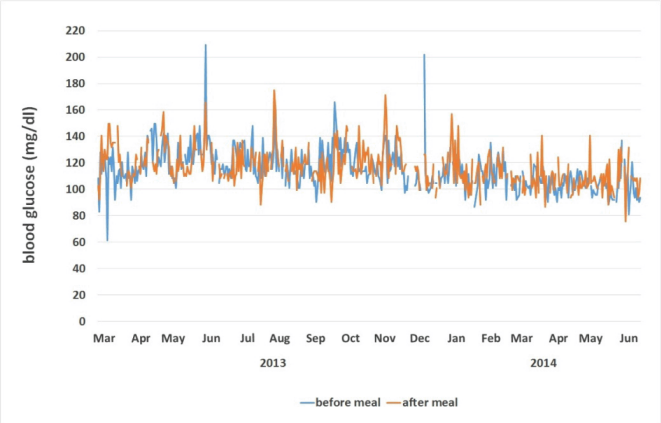


Figure 2: Glucose levels preprandially and postprandially while on the paleolithic ketogenic diet between February 2013 and June 2014. Due to normalized blood glucose levels later on the patient measured blood glucose only occasionally.

DISCUSSION

Recent clinical studies show that low-carbohydrate diets including the paleolithic diet are beneficial in conditions associated with the metabolic syndrome [6–10]. In the current medical practice, patients with metabolic syndrome are generally treated with numerous drugs. Yet studies available on the paleolithic diet does not give clues how to deal with preexisting medications when shifting towards the paleolithic diet. Our experience indicate that upon the shift towards the paleolithic ketogenic diet most drugs become unnecessary and should be discontinued [14].

Herein, we analyzed past medical history of a patient with metabolic syndrome to reveal how her medication might have contributed to the worsening of her disease and how disease was influenced by the paleolithic ketogenic diet.

This case represents a typical disease career of a patient with metabolic syndrome. Her medical history included elevated glucose parameters and hypertension initially controlled with oral antidiabetics and antihypertensives in 2005. Then with the emergence of new diagnostic findings pantoprazole was added. Two years later metformin was added to support glucose metabolism. Nevertheless glucose control deteriorated again in 2011 following the addition of three new drugs. These included bisoprolol, acetylsalicylic acid and amlodipine. Bisoprolol and amlodipine, which belong to beta-blocker and calcium channel blocker type antihypertensives, respectively, are known for their effect to adversely affect glucose metabolism [15, 16]. Glucose parameters further deteriorated in 2012.

Current guidelines first advise lifestyle changes and exercise to treat the metabolic syndrome and to induce

weight loss [1]. Yet these dietary advices usually remain without effect [17]. Also, overweight people are unable to exert considerable physical activity. Therefore, patients are prescribed medications. A main problem is that in current medical practice each component of the metabolic syndrome is treated separately even though targeting one symptom by a drug may worsen another symptom also associated with the metabolic syndrome. Therefore, the number of drugs as well as side effects are increasing.

Our patient was overweight and was taking eight medicines. When shifting towards the paleolithic ketogenic diet she was able to discontinue all medications. Her weight begin to decrease along with improving glucose parameters and lowered blood pressure. Our experience with patients on the paleolithic ketogenic diet indicate that the use of antihypertensive drugs hinder the normalization of glucose levels and weight loss [14]. Also, antihypertensive drugs become unnecessary since the paleolithic ketogenic diet efficiently lowers high blood pressure. In this case, antihypertensive drugs could be discontinued because the patient had no atherosclerosis. In those cases, with atherosclerosis antihypertensives may be discontinued only within a longer time period. The blood pressure lowering effect of the paleolithic ketogenic diet is due to the fact that the paleolithic ketogenic diet limits those food components which result in elevated blood pressure. These components mainly include fruits and foods with added fructose. Drop in blood pressure was also reported in previous studies with the paleolithic diet [6, 8]. Our experience with patients with hypertension indicate that as compared to the paleolithic diet which does not limit fruits and vegetables, the paleolithic ketogenic diet more efficiently normalize blood pressure. Given that in the paleolithic ketogenic diet carbohydrate intake is strongly limited less insulin is required for normoglycemia and therefore oral antidiabetics become unnecessary.

While on the paleolithic ketogenic diet home monitoring of glucose in our patient showed a decreasing tendency both preprandially and postprandially. Laboratory measurements also showed a decreasing tendency in glucose parameters and triglyceride normalized too. Total cholesterol and LDL cholesterol tended to decrease while HDL cholesterol remained relatively unchanged. Uric acid remained in the normal range while on the paleolithic ketogenic diet. These laboratory parameters are similar to those in our two previous cases on the paleolithic ketogenic diet [12, 13]. All six urinary tests were positive for ketones indicating a good adherence to the diet. The patient admitted to occasionally exceed the advised limit for fruit. She also linked excessed fruit intake to high blood pressure spikes which is in accordance with literature data showing high blood pressure to be associated with fructose [18]. Nevertheless our patient reported a decreasing tendency of blood pressure across the 22 months.

Those patients who underwent gallbladder surgery are advised against eating fatty foods and the ketogenic diet too. Our patient, however, reported no gastrointestinal side effects while on the paleolithic ketogenic diet. Of note, her winter-time vitamin D level was normal in spite of no vitamin D supplementation indicating that a regular intake of offal, animal fat and meat may ensure normal vitamin D status. Also, no signs of vitamin or mineral deficiency emerged despite of the lack of supplementation.

Currently, the patient is on the paleolithic ketogenic diet for 22 months. No side effects emerged and she is free of symptoms. She is resoluted to continue the diet.

CONCLUSION

The paleolithic ketogenic diet proved to be a safe, feasible and effective therapy in this patient with metabolic syndrome. Medications could be discontinued and components of the metabolic syndrome improved continuously. We used the paleolithic ketogenic diet in a patient without a gallbladder indicating that, contrary to the widely held notion, this organ is not a prerequisite for maintaining a diet rich in animal fat. Neither vitamin nor other supplements was used indicating the effectiveness of the paleolithic ketogenic diet as a sole therapy.

****

Author Contributions

Csaba Tóth – 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

Zsófia Clemens – 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. International Diabetes Federation: The IDF consensus worldwide definition of the metabolic syndrome.

Available at: http://www.idf.org/metabolic-syndrome Accessed December 8, 2014.
2. Perez-Pastor EM, Metcalf BS, Hosking J, Jeffery AN, Voss LD, Wilkin TJ. Assortative weight gain in mother-daughter and father-son pairs: An emerging source of childhood obesity. Longitudinal study of trios (EarlyBird 43). Int J Obes (Lond) 2009 Jul;33(7):727–35.
3. Hofer-Dückelmann C. Gender and polypharmacotherapy in the elderly: A clinical challange. In: Sex and Gender Differences in Pharmacology (Ed: Regitz-Zagrosek V) 2002. pp. 169–82.
4. Lindeberg S. Food and western disease: Health and nutrition from an evolutionary perspective. Chichester: Wiley-Blackwell 2009.
5. Cordain L. The paleo diet: Lose weight and get healthy by eating the food you were designed to eat. New York: Wiley 2002.
6. Österdahl M, Kocturk T, Koochek A, Wändell PE. Effects of a short-term intervention with a paleolithic diet in healthy volunteers. Eur J Clin Nutr 2008 May;62(5):682–5.
7. Jönsson T, Granfeldt Y, Ahrén B, et al. Beneficial effects of a Paleolithic diet on cardiovascular risk factors in type 2 diabetes: A randomized cross-over pilot study. Cardiovasc Diabetol 2009 Jul 16;8:35.
8. Frassetto LA, Schloetter M, Mietus-Synder M, Morris RC Jr, Sebastian A. Metabolic and physiologic improvements from consuming a paleolithic, hunter-gatherer type diet. Eur J Clin Nutr 2009 Aug;63(8):947–55.
9. Mellberg C, Sandberg S, Ryberg M, et al. Long-term effects of a Palaeolithic-type diet in obese postmenopausal women: A 2-year randomized trial. Eur J Clin Nutr 2014 Mar;68(3):350–7.
10. Boers I, Muskiet FA, Berkelaar E, et al. Favourable effects of consuming a Palaeolithic-type diet on characteristics of the metabolic syndrome: A randomized controlled pilot-study. Lipids Health Dis 2014 Oct 11;13:160.
11. Voegtlin WL. The stone age diet: Based on in-depth studies of human ecology and the diet of man. New York: Vantage Press 1975.
12. Clemens Z, Kelemen A, Fogarasi A, Tóth C. Childhood Absence Epilepsy Successfully Treated with the Paleolithic Ketogenic Diet. Neurology and Therapy 2013;2(1-2):71–6.
13. Tóth C, Clemens Z. Type 1 diabetes mellitus successfully managed with the paleolithic ketogenic diet. Int J Case Rep Images 2014;5(10):699–703.
14. Tóth C. Paleolithic medicine (Paleolit orvoslás). Budapest: Jaffa 2012.
15. Blackburn DF, Wilson TW. Antihypertensive medications and blood sugar: Theories and implications. Can J Cardiol 2006 Mar 1;22(3):229–33.
16. Levine M, Boyer EW, Pozner CN, et al. Assessment of hyperglycemia after calcium channel blocker overdoses involving diltiazem or verapamil. Crit Care Med 2007 Sep;35(9):2071–5.
17. Accurso A, Bernstein RK, Dahlqvist A, et al. Dietary carbohydrate restriction in type 2 diabetes mellitus and metabolic syndrome: Time for a critical appraisal. Nutr Metab (Lond) 2008 Apr 8;5:9.
18. Madero M, Perez-Pozo SE, Jalal D, Johnson RJ, Sánchez-Lozada LG. Dietary fructose and hypertension. Curr Hypertens Rep 2011 Feb;13(1):29–35.

ABOUT THE AUTHORS

Article citation: Tóth C, Clemens Z. Successful treatment of a patient with obesity, type 2 diabetes and hypertension with the paleolithic ketogenic diet. Int J Case Rep Images 2015;6(3):161–167.

Csaba Tóth is physician from Hungary with 20 years experience in intensive care medicine, internal medicine and family medicine. He is using the paleolithic-ketogenic nutrition in the treatment of chronic medical illnesses including diabetes, cancer, autoimmune diseases and epilepsy for five years. He is operating private practice in Budapest and in a few other cities in Hungary. In 2013, he organized a course on evolutionary medicine in the mandatory training of GPs at the University of Szeged, Hungary. With an evolutionary medical attitude he strives for the full recovery of his patients.
E-mail: csaba@paleomedicina.com

Zsófia Clemens is biologist and clinical researcher specialized in nutrition, nutritional therapy and brain research. She earned her PhD in electroencephalograpy of sleep and epilepsy from Semmelweis University, Budapest, Hungary in 2005. Currently, she is Senior Research Fellow at the Neurological Department, University of Pécs, Hungary and is also affiliated with the Evolutionary Medicine Working Group of Paleomedicina Hungary Ltd. In international academic journals, she has published 30 research articles with more than 500 citations. E-mail: clemenszsofia@gmail.com

CASE REPORT PEER REVIEWED | OPEN ACCESS

Massive hemoperitoneum from a ruptured corpus luteum cyst masquerading as biliary colic

Justin B. Belsky, Jumana F. Nagarwala, Glenn F. Tokarski

ABSTRACT

Introduction: Corpus luteum cysts are functional cysts that secrete progesterone in preparation for pregnancy. If conception does not occur, they typically dissipate, however, they may collect with fluid or blood and form a cyst that can rupture. Although cyst rupture is generally benign, causing mild pain to the patient, it can result in massive hemoperitoneum requiring emergent surgical intervention. Case Report: A 23-year old female presented to the emergency department after experiencing several hours of right upper quadrant abdominal pain. Emergency department evaluation included normal liver function studies and a negative urine and serum pregnancy test. Abdominal ultrasound revealed a large amount of intraperitoneal fluid but no biliary disease was identified. While in the emergency department, she developed signs of acute hypovolemic shock. Serial hemoglobin revealed a four-gram drop after arrival in emergency department. She underwent emergent exploratory laparoscopy where massive hemoperitoneum (750 cm³ of blood) was identified. A ruptured corpus luteum cyst was identified as the source of acute blood loss. Conclusion: We present the first known case of a ruptured corpus luteum cyst resulting in massive hemoperitoneum masquerading as biliary colic. It is paramount that clinicians consider a ruptured ovarian cyst in females with abdominal pain, regardless of the location of pain.

Keywords: Corpus luteum cyst, Ectopic pregnancy, Hemoperitoneum, Right upper quadrant pain

How to cite this article

Belsky JB, Nagarwala JF, Tokarski GF. Massive hemoperitoneum from a ruptured corpus luteum cyst masquerading as biliary colic. Int J Case Rep Images 2015;6(3):168–172.

doi:10.5348/ijcri-201531-CR-10492

Justin B. Belsky[1], Jumana F. Nagarwala[2], Glenn F. Tokarski[3]

Affiliations: [1]MD, Assistant Instructor, Department of Emergency Medicine, Massachusetts General Hospital, Harvard Medical School, Boston, Massachusetts, United States of America; [2]MD, Senior Staff Physician, Department of Emergency Medicine, Henry Ford Hospital, Detroit, Michigan, United States of America; [3]MD, Department of Emergency Medicine, Henry Ford Hospital, Detroit, Michigan, United States of America.

Corresponding Author: Justin Brett Belsky, 55 Fruit Street, Zero Emerson Place, Suite 3B, Boston, Massachusetts 02114, United States of America; Ph: 248-860-5854; Fax: 617-724-0917; Email: jbelsky@partners.org

Received: 22 December 2014
Accepted: 19 January 2015
Published: 01 March 2015

INTRODUCTION

The corpus luteum is a temporary hormone secreting remnant of a mature ovarian follicle after it ruptures to release an ovum into the fallopian tube. Its main function is to establish and maintain early pregnancy by secreting progesterone. If fertilization occurs the corpus luteum involutes at approximately 8–10 weeks of gestation and the placenta produces progesterone for the remainder of pregnancy. If fertilization does not occur, the corpus luteum involutes several days after ovulation (now referred to as the corpus albicans), shrinks and stops producing progesterone resulting in endometrial sloughing and menstruation. However, the corpus luteum may fill with blood or other fluids forming a cyst and rupture [1].

Abdominal pain caused by a ruptured corpus luteum cyst is a common complaint seen in the emergency department in a woman of childbearing age. They are seldom of any pathological consequence and are usually self-limited to pain. However, in rare cases they can lead to massive hemoperitoneum requiring surgical management.

CASE REPORT

A 23-year old female presented to the emergency department with abdominal pain. The patient reported colicky, non-radiating right upper quadrant (RUQ) pain with onset while eating approximately 15 hours prior to admission. Over the next several hours, she experienced several similar episodes of pain of variable intensity that were also associated with eating. Two episodes of non-bloody emesis occurred during this time but she denied other gastrointestinal symptoms. Her last menstrual period was nine days prior and normal in timing and duration. She was sexually active and did not use birth control. She had sexual intercourse less than 30 minutes prior to her first episode of pain. She denied vaginal bleeding and had no known gynecologic problems. She denied urinary symptoms and reported no previous abdominal surgeries.

Vital signs upon arrival revealed a temperature of 36.9°C, blood pressure 96/64 mmHg, heart rate 99 bpm, respiratory rate 16 bpm, and SaO2 100% on room air. Her abdominal examination was remarkable for diffuse tenderness throughout the entire abdomen but worse in the RUQ. Voluntary guarding with palpation was noted in the RUQ area but no peritoneal signs were present. Murphy's sign was negative and the patient did not have any masses or hepatosplenomegaly. The patient denied costovertebral tenderness. Her skin was pale without evidence of jaundice.

Pertinent laboratory results included white blood cell count 16.5 K/uL (normal 3.8–10.6 K/uL) with absolute neutrophil count 14.51 K/uL (normal 1.0–7.70 K/uL), hemoglobin 10.3 g/dL (normal 12.0–15.0 g/dL). Serum electrolytes, liver function tests, lipase and urinalysis were normal. Urine pregnancy test was negative and serum beta-HCG was <10 IU (non-pregnant <10 IU). Abdominal ultrasound revealed no gallstones or biliary dilatation, however, a large amount of intra-peritoneal fluid was observed. Transvaginal ultrasound revealed a complex mass in the left ovary and a large amount of intra-peritoneal fluid was again described, consistent with an ectopic pregnancy (Figure 1).

The negative pregnancy markers were not consistent with the ultrasound findings of a ruptured ectopic pregnancy and the history, examination and normal liver function tests not consistent with ascites so the studies were re-examined with an independent ultrasound radiologist. A complex left adnexal mass and large amount of peritoneal fluid were again identified. Computed tomography (CT) scan of the abdomen/pelvis was recommended to better delineate possible intra-abdominal and/or pelvic pathology. This study revealed a complex left ovarian mass and a large amount of intra-peritoneal fluid with a radiodensity of blood – findings again suggestive of a ruptured ectopic pregnancy (despite the negative urine and serum pregnancy tests) (Figure 2).

An emergent OB-GYN consultation was obtained. After examining the patient and reviewing the laboratory, ultrasound and CT studies the consultant diagnosed a possible ruptured ovarian cyst and recommended admission to the observation unit for symptom control, repeat examinations and hemoglobin monitoring.

Shortly, after OB-GYN evaluation the patient again complained of severe RUQ pain. Repeat vital signs revealed a heart rate 131 bpm and blood pressure 114/57 mmHg. Examination demonstrated diffuse abdominal pain worse in the RUQ and rebound tenderness was now present. Volume resuscitation was initiated and the OB-

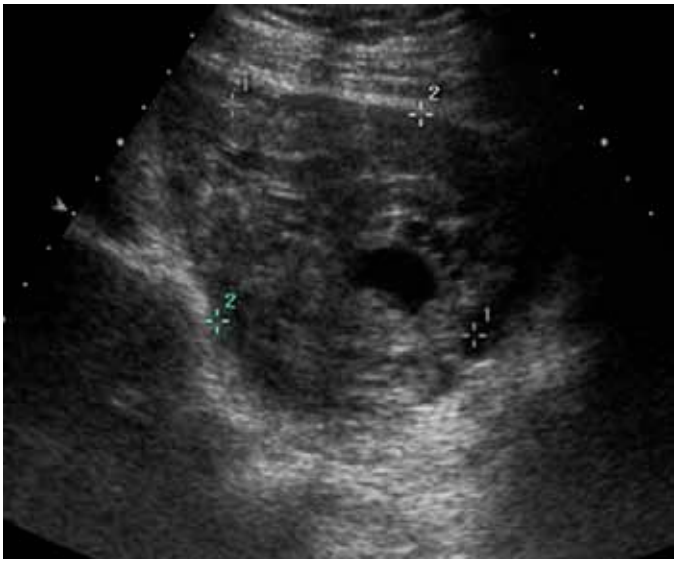


Figure 1: Transvaginal ultrasound of left adnexa. Adjacent to the left ovary there is a 8.9x5.6x6 cm complex mass which contains a 2.2–2.2 mm anechoic structure.

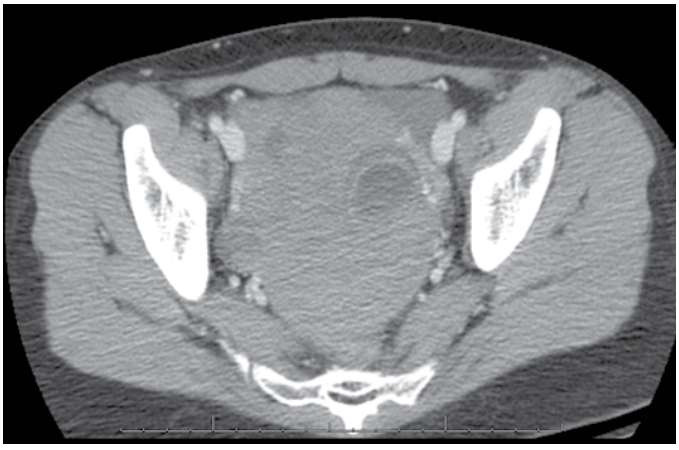


Figure 2: Computed tomography scan with contrast of the pelvis: A 3-cm cystic focus present in the left adnexa with surrounding hyperdense material most consistent with acute blood products.

GYN consultant recalled. Repeat hemoglobin was found to be 8.4 g/dL (down from 10.3 g/dL 5 hours prior). The patient was taken for emergent exploratory laparoscopy.

Upon exploration of the abdomen, 750 cm^3 of intra-abdominal blood was identified. The liver and abdominal organs were normal. A ruptured corpus luteum cyst was identified in the left ovary and was felt to be the source of the intra-abdominal hemorrhage. An intraoperative hemoglobin was found to be 6.0 g/dL so blood was transfused. Postoperative hemoglobin levels remained stable and pain improved. The patient was discharged on the second postoperative day. When seen in the OB-GYN clinic two weeks later she was symptom-free and her hemoglobin level was similar to the day of hospital discharge.

DISCUSSION

During the follicular phase of a normal menstrual cycle a single ovarian follicle is stimulated to grow by follicle stimulating hormone and luteinizing hormone (LH). A mid-cycle surge of LH stimulates the mature follicle to rupture and release the ovum into the fallopian tube. The remnants of the ruptured ovarian follicle is now referred to as the corpus luteum. The corpus luteum functions during the luteal phase of the menstrual cycle by synthesizing and releasing multiple hormones of which progesterone is the most important (responsible for the maintenance of the highly vascular and glandular endometrium). If fertilization occurs, the corpus luteum involutes at approximately 8–10 weeks of gestation and the placenta produces progesterone for the remainder of pregnancy. If fertilization does not occur, the corpus luteum involutes several days after ovulation (now referred to as the corpus albicans), shrinks and stops producing progesterone resulting in endometrial sloughing and menstruation.

The corpus luteum is a highly vascular structure. The rate of blood flow to the corpus luteum exceeds any other adult organ (per unit of tissue); the increased blood flow is needed to deliver substrates for hormone production and to nurture the rapidly dividing luteal cells [2]. The highly vascular nature of the corpus luteum is reflected by its oxygen consumption which is estimated to be 2–6 times that of the liver, kidney and heart (per unit of tissue) [1, 2]. If pregnancy occurs, the corpus luteum grows to 3–5 cm in size. Marked angiogenesis occurs in the corpus luteum of pregnancy; 50–70% of all cells in the mature corpus luteum are endothelial cells or microvascular pericytes and capillary lumina account for 22% of the size of the corpus luteum.

Ovarian cysts occur commonly in menstruating women ages 18–35 years and are commonly identified by ultrasound. The minimum size to be considered a cyst is 2.5–3.0 cm [3]. Two types of functional ovarian cysts are described. Follicular cysts arise from ovarian follicles that do not mature into the primary follicle. They are of variable size, thin walled and usually contain clear cystic fluid. Pain may result from stretching of the ovarian capsule, bleeding into the cyst or spontaneous rupture and local peritoneal irritation. Most follicular cysts will spontaneously regress and medical intervention is usually limited to symptomatic control.

Corpus luteal cysts arise due to the rapid growth and high vascularity of the corpus luteum resulting in intra-luteal hemorrhage and the formation of a hemorrhagic cyst. Ongoing bleeding into the cyst can result in rapid enlargement, spontaneous rupture and leakage of blood into the peritoneal cavity. Abdominal and/or pelvic pain may occur with any of these complications.

Hemoperitoneum resulting from ruptured corpus luteum cyst have been described since the early 1900s and is frequently misdiagnosed as ruptured ectopic pregnancy, acute appendicitis, ovarian torsion and endometriosis [4, 5]. The incidence of this complication is not reported; practicing gynecologists relate this as rare but recognized. More recent case reports of massive hemoperitoneum from a ruptured corpus luteum cyst are associated with systemic anticoagulation, coagulation disorders, von Willebrand disease, and sickle cell anemia [6–9]. Symptoms presenting following sexual intercourse, such as those described by our patient, are also described in literature [10, 11]. It is possible that the changes in intraluminal pressure created during sexual intercourse was a catalyst for cyst rupture.

Corpus luteal cysts are commonly identified as incidental structures on pelvic ultrasound studies. Corpus luteum cyst rupture with intra-abdominal hemorrhage may appear ultrasonographically identical to a ruptured ectopic pregnancy as in our case a negative serum pregnancy test may be a discerning feature [11, 12]. Computed tomography scan of the abdomen/pelvis is of limited value and usually supports the diagnosis of ruptured ectopic pregnancy–again a negative serum pregnancy test should suggest an alternate diagnosis [13].

Therapy for ruptured corpus luteal cysts with intra-abdominal hemorrhage must be tailored to the patient. Observation, pain control and serial hemoglobin monitoring may be appropriate for select cases but signs of blood loss or hypovolemic shock should prompt immediate surgical intervention.

Our patient presented with symptoms and findings suggestive of acute biliary colic but testing revealed no biliary disease. Gynecologic causes of RUQ pain include perihepatitis and adhesions between the liver and abdominal wall due to gonococcal and chlamydial salpingitis (Fitzhugh–Curtis syndrome). Other common differentials for a young female with abdominal pain include, gastritis, peptic ulcer disease, gastroesophageal reflux disease, ectopic pregnancy, pyelonephritis, pancreatitis, and ovarian related conditions such as pelvic inflammatory disease, ovarian torsion, and cyst rupture.

CONCLUSION

Massive hemoperitoneum resulting from a ruptured corpus luteum cyst is rare, but potentially life-threatening if not diagnosed and treated emergently. We could identify no other reports of hemoperitoneum from a ruptured corpus luteum cyst with a similar presentation. We hypothesize that our patient's right upper quadrant pain was secondary to accumulation of blood into Morison's pouch. Abdominal pain in a young female is a frequent complaint in the emergency department and massive hemoperitoneum from a ruptured corpus luteum cyst should be in the differential diagnosis for all females presenting with abdominal pain, regardless of the location.

****

Author Contributions

Justin Belsky – Substantial contributions to conception and design, Analysis and interpretation of data; Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published.
Jumana Nagarwala – Substantial contributions to conception and design, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published.
Glenn Tokarski – Substantial contributions to conception and design, 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. Niswender GD, Juengel JL, Silva PJ, Rollyson MK, McIntush EW. Mechanisms controlling the function and life span of the corpus luteum. Physiol Rev 2000 Jan;80(1):1–29.
2. Swann RT, Bruce NW. Oxygen consumption, carbon dioxide production and progestagen secretion in the intact ovary of the Day-16 pregnant rat. J Reprod Fertil 1987 Jul;80(2):599–605.
3. Potter AW, Chandrasekhar CA. US and CT Evaluation of Acute Pelvic Pain of Gynecologic origin in Nonpregnant Premenopausal Patients. Radiographics 2008 Oct;28(6):1645–59.
4. Waters H. Hemorrhage into an ovarian cyst simulating ectopic pregnancy. JAMA 1918;70(5):295–6.
5. Phaneuf L. Intraperitoneal hemorrhage from ruptured ovarian cyst. JAMA 1924;83(9):658–2.
6. Hackethal A, Ionesi-Pasacica J, Kreis D, Litzlbauer D, Tinneberg HR, Oehmke F. Feasibility of laparoscopic management of acute haemoperitoneum secondary to ruptured ovarian cysts in a haemodynamically unstable patient. Minim Invasive Ther Allied Technol 2011 Jan;20(1):46–9.
7. Gupta N, Dadhwal V, Deka D, Jain SK, Mittal S. Corpus luteum hemorrhage: Rare complications of congenital and acquired coagulation abnormalities. J Obstet Gynaecol Res 2007 Jun;33(3):376–80.
8. Terzic M, Likic I, Pilic I, Bila J, Knezevic N. Conservative management of massive hematoperitoneum caused by ovulation in a patient with severe form of von Willebrand disease–-a case report. Clin Exp Obstet Gynecol 2012;39(4):537–40.
9. Andikyan V, Ronald J, Bowers C. Massive hemoperitoneum secondary to ruptured corpus luteum cyst of pregnancy in 17-year old female with hemoglobin SC disease. The Internet Journal of Gynecology and Obstetrics 2010;12(2). Available at: https://ispub.com/IJGO/12/2/3876. Accessed December 15, 2014.
10. Ho WK, Wang YF, Wu HH, Tsai HD, Chen TH, Chen M. Ruptured Corpus Luteum With Hemoperitoneum: Case Characteristics And Demographic Changes Over Time. Taiwan J Obstet Gynecol 2009 Jun;48(2):108–2.
11. Hallatt JG, Steele CH Jr, Snyder M. Ruptured corpus luteum with hemoperitoneum: A study of 173 surgical cases. Am J Obstet Gynecol 1984 May 1;149(1):5–9.
12. Yoffe N, Bronshtein M, Brandes J, Blumenfeld Z. Hemorrhagic ovarian cyst detection by transvaginal sonography: The great imitator. Gynecol Endocrinol 1991 Jun;5(2):123–9.
13. Swire MN, Castro-Aragon I, Levine D. Various sonographic appearances of the hemorrhagic corpus luteum cyst. Ultrasound Q 2004 Jun;20(2):45–58.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Internal herniation beneath mesodiverticular band producing ambiguous picture in intestinal obstruction: A management dilemma

Rakesh Chauhan, Rajesh Chaudhary, R. Dayashankar

ABSTRACT

Introduction: Mesodiverticular band is a rare cause of obstruction due to Meckel's diverticulum complication. However, exact incidence could not be found in literature. Obstruction as a complication of the Meckel's diverticulum is the most common presentation in adults. Case Report: We present here a case of intestinal obstruction by mesodiverticular band in a 13-year-old child with an ambiguous picture by patient clinically settling with conservative treatment but radiologically air fluid levels still persisting. Conclusion: Mesodiverticular band cannot be diagnosed preoperatively and because of the loosely entrapped loop, obstruction is not complete. So it produces difficult situation for the surgeon to take decision in favor of surgery.

Keywords: Intestinal obstruction, Management dilemma, Meckel's diverticulum, Mesodiverticular band

How to cite this article

Chauhan R, Chaudhary R, Dayashankar R. Internal herniation beneath mesodiverticular band producing ambiguous picture in intestinal obstruction: A management dilemma. Int J Case Rep Images 2015;6(3):173–177.

Rakesh Chauhan[1], Rajesh Chaudhary[2], R. Dayashankar[2]

Affiliations: [1]MBBS, MS, Assistant Professor Surgery Dr. RP Govt. Medical College Kangra at Tanda, HP, India; [2]MBBS, Junior Resident, Dr. RP Govt. Medical College Kangra at Tanda, Himachal Pradesh, India.

Corresponding Author: Dr. Rakesh Chauhan, MS, Assistant Professor Surgery, Dr. RP Govt. Medical College Kangra at Tanda, Himachal Pradesh, India; Email: drrakeshchauhan66@gmail.com

Received: 22 November 2014
Accepted: 02 January 2015
Published: 01 March 2015

doi:10.5348/ijcri-201532-CR-10493

INTRODUCTION

Intestinal obstruction because of entrapment of small gut loop between mesodiverticular band and mesentery is a rare cause of obstruction due to Meckel's diverticulum complication. It cannot be diagnosed preoperatively and because of the loosely entrapped loop obstruction is not complete. So it produces difficult situation for the surgeon to take decision in favor of surgery [1]. We report a case of 13-year-old boy presenting with intestinal obstruction due to mesodiverticular band which presented an ambiguous picture difficult to take decision for surgery.

CASE REPORT

A 13-year-old boy from poor socio-economic family background presented to the emergency outpatient department in the night with a history of colicky pain in umbilical region associated with bilious vomiting and constipation for the last two days. There was a history of ingestion of some dry fruits and half cooked food before the appearance of symptoms. There was no history suggestive of worm infestation. On examination, the patient was afebrile and pulse was 80/min. Blood pressure was normal and there was no dehydration. Abdomen was mildly distended and soft, but bowel sounds were not present. Hernia sites were normal and on digital rectal examination (DRE) there was slight ballooning with hard fecal matter impacted. Biochemistry and hematology was normal especially the TLC which was 5700/cc. Urine output was adequate. Ryle's tube aspirate was about 1 L. Chest X-ray was normal and standing abdominal X-ray showed multiple air fluid levels in the small gut and there was no gas in the large gut (Figure 1). Peritoneal aspiration showed straw colored fluid. Diagnosis of acute intestinal obstruction was made with possibility of ileocecal tuberculosis or bolus obstruction.

So conservative trial was given with proctoclysis enema. Patient passed multiple hard stools with passage of the flatus. With this pain subsided altogether and distention was decreased. Next day air fluid levels persisted in the abdominal X-ray. However, patient was pain free and Ryle's tube aspirate was decreased to 200 mL/d. Patient gave the history of passage of flatus in between. Sluggish bowel sounds were also heard. On second day of admission the patient was again pain free, feeling hungry, pulse was 78/min, abdomen was soft, bowel sounds were very sluggish and rectum was collapsed but air fluid levels persisted in the X-ray (Figure 2). Therefore, decision for laparotomy was taken. Patient was taken up for surgery with prior anesthetic clearance with one unit of whole blood arranged. On exploration mesodiverticular band extending from the tip of Meckel's diverticulum to the right side of the mesentery was found forming a tunnel of about 2–3 cm through which small gut loops were loosely herniated without forming complete obstruction or constriction band in its wall (Figure 3). Proximal segment was dilated and there was about 200 cm^3 straw colored fluid in the cavity. Diverticulum was about 7 cm with wide base (Figure 4). Excision of band and diverticulum with ileo-ileal anastomosis was done. Patient recovered well without any complication and discharged on eighth day of surgery. Specimen was sent for histopathological examination. Histopathological examination report confirms Meckel's diverticulum with a fibrous band attached to its tip.

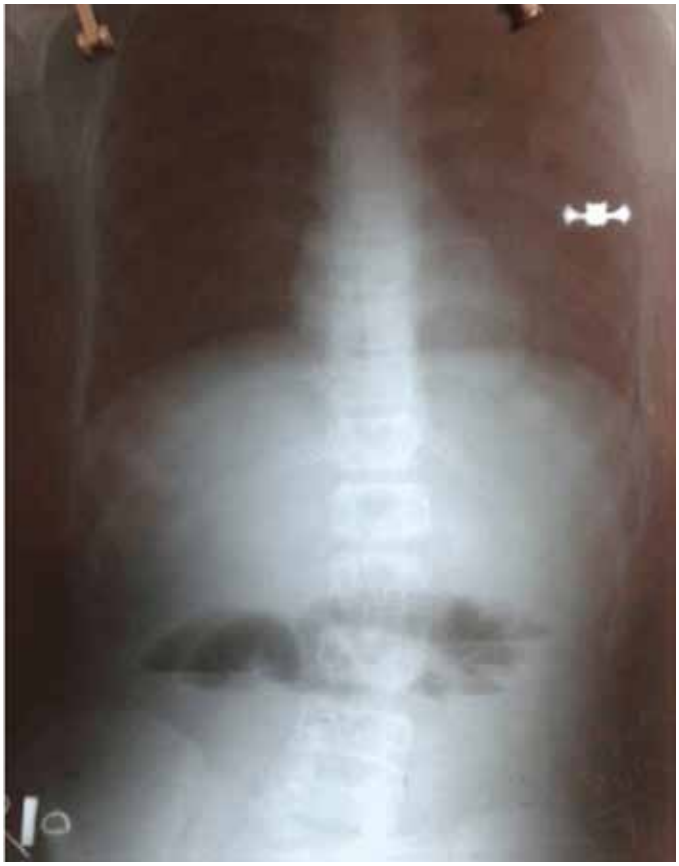


Figure 1: X-ray of abdomen standing at the time of admission.

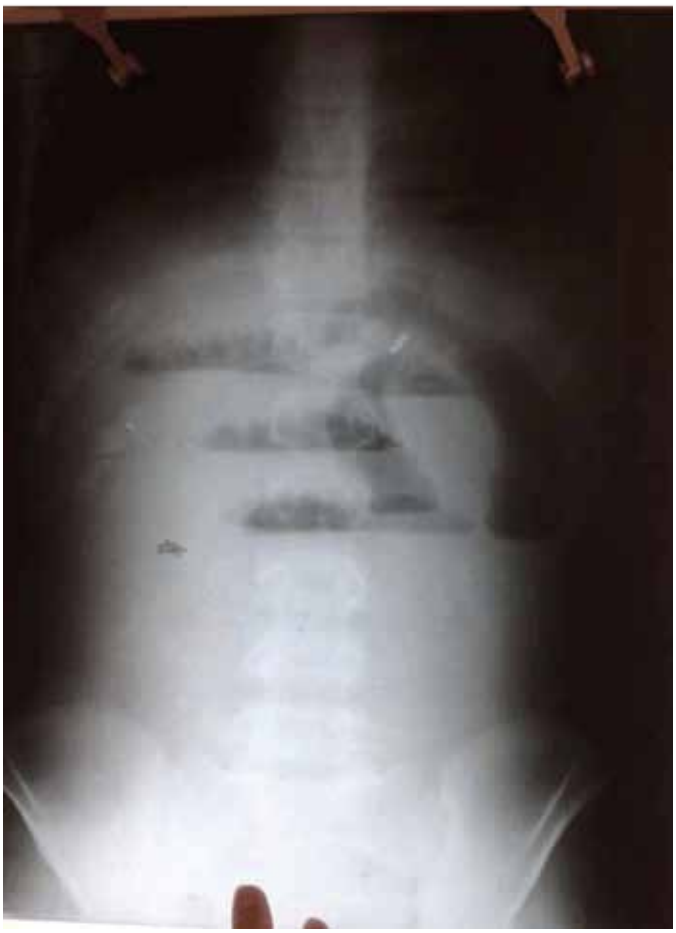


Figure 2: X-ray of abdomen standing after 36 hours of conservative management.

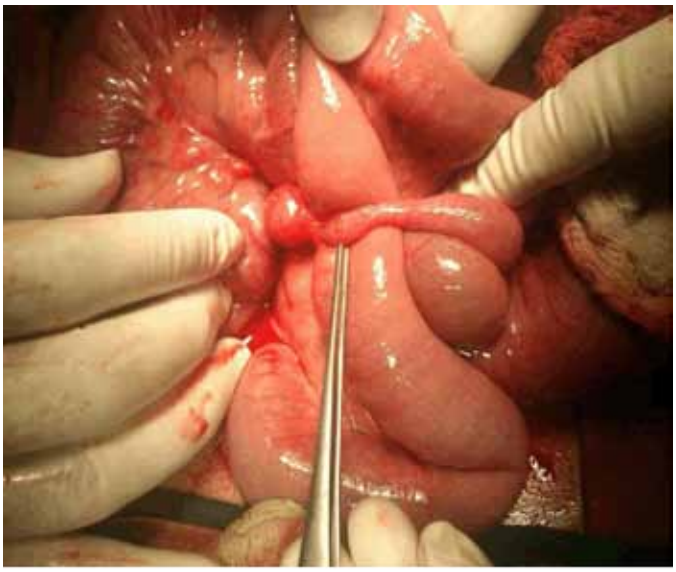


Figure 3: The loop of the small gut entrapped between the mesodiverticular band and the mesentery, which is viable.

DISCUSSION

Meckel's diverticulum, a true diverticulum, is the most common congenital abnormality of the gut. First reported

in 1598 by Hildanus, it was described in anatomical detail by Johann Meckel in 1809 whose name it carries [2]. It is a remnant of intestinal end of omphalomesenteric duct which normally gets obliterated during eighth week of gestation. It is present in 2% of the population, however, about 96% of the cases are asymptomatic in which it is incidentally detected on surgery, radiographic imaging, endoscopically for other purposes or on autopsy [3]. Only in 4% of the cases symptoms arise as a result of complications related to diverticulum. More than 50% of the symptomatic patients are less than two years of age [4]. The complications related to the Meckel's diverticulum include bleeding, intestinal obstruction, infection, perforation, neoplasms. The most common symptom of complicated Meckel's diverticulum is intestinal obstruction. The reasons for obstruction are volvulus around the band between diverticulum and umbilicus, intussusception of the inverted diverticulum acting as a lead point, stricture, entrapment of gut loops under the mesodiverticular band and litter's hernia [5]. Mesodiverticular band is a rare intraoperative finding leading to entrapment of the gut loop between it and mesentery. Mesodiverticular band cannot be diagnosed preoperatively by any investigation and is an incidental finding during surgery only. It is a flimsy mesentery carrying independent blood supply to the diverticulum through persistent vitelline vessels and runs between the tip of diverticulum and the mesentery. Through the space between the two (mesentery and Meckel's diverticulum carrying band) internal herniation of the small gut loops take place [6]. Investigations for Meckel's diverticulum depends upon the complication it produces. For the bleeding which is the most common complication in the children sodium 99Tc pertechnetate scan is investigation of the choice with accuracy of more than 90% [7]. In adults accuracy of scan is less than 50% because of less prevalence of the gastric mucosa. In adults, accuracy can be increased by pentagastrin, cimetidine or glucagon [8]. Angiography can also be used to detect the bleeding. Obstruction as a result of the intussusception can be detected with the help of the ultrasonography or computed tomography scan or barium enema [7, 8]. But in most of the cases of obstruction, cause is confirmed on laparotomy. This patient also presented to the emergency department with a history of colicky pain abdomen, vomiting and not passing stool and flatus for the last two days. Vitals were normal but standing X-ray of abdomen showed multiple air fluid levels in small gut. Abdomen was mildly distended but soft, however, shifting dullness was present. Paracentesis showed straw colored fluid. RT aspiration showed bilious fluid about one liter on presentation. There was no history of contact with tuberculosis patient or intake of ATT but ileocecal tuberculosis was kept one of the possibilities. With conservative treatment patient settled clinically but not radiologically and on exploratory laparotomy internal herniation of small gut through a mesodiverticular band was found. This picture leading to the incomplete obstruction makes the decision difficult if to carry on conservative treatment or do emergency laparotomy. But since the air fluid levels were not disappearing despite the improvement in the clinical status we decided in the favor of laparotomy and found mesodiverticular band. Emptied sigmoid colon with enema reduced the pressure and allowed luminal contents to pass through herniated gut leading to relief of symptoms but loop was still entrapped under the mesodiverticular band not allowing to disappearance of the air fluid levels. It projected ambiguous picture leading to difficult situation for the surgeon to take decision for surgery.

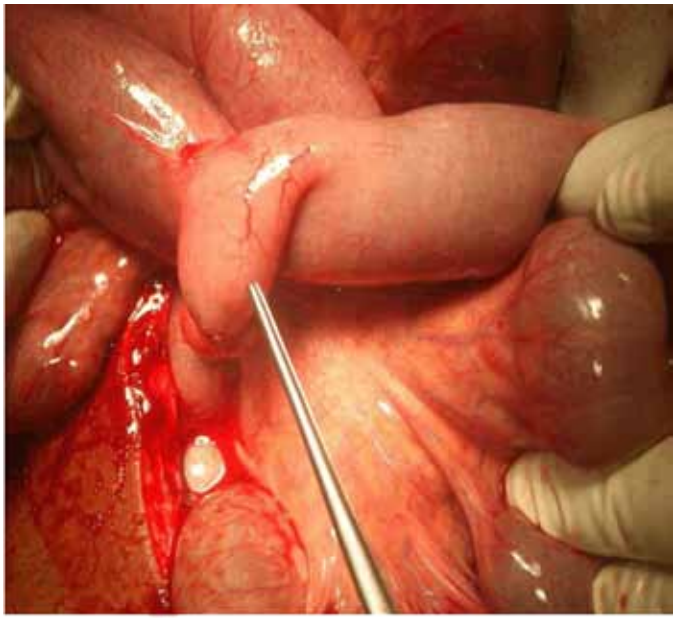


Figure 4: Herniated loop has been retrieved and showing diverticulum with band attached to mesentery.

CONCLUSION

Mesodiverticular band is a rare cause of intestinal obstruction due to loose entrapment of small gut loops in a tunnel produced by it and mesentery. So it does not produce complete obstruction. On conservative treatment patient gives an improving clinical picture but radiologically obstruction is not settled. Therefore, surgeon faces a dilemma weather to operate or to continue manage the patient conservatively. We faced this challenge and decided to operate the patient.

****

Author Contributions

Rakesh Chauhan – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, drafting the article, Critical revision of the article and final approval of the version to be published

Rajesh Chaudhary – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, drafting the article, Critical revision of the article and final approval of the version to be published
Dayashankar – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, drafting the article, Critical revision of the article and 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. Park JJ, Wolff BG, Tollefson MK. Meckel diverticulum: the Mayo Clinic experience with 1476 patients (1950-2002). Ann Surg. Mar 2005;241(3):529-33.
2. Opitz JM, Schultka R, Gobbel L. Meckel on developmental pathology. Am J Med Genet A. Jan 15 2006;140(2):115-28.
3. Anderson DJ. Carcinoid tumor in Meckel's diverticulum: laparoscopic treatment and review of the literature. J Am Osteopath Assoc. Jul 2000;100(7):432-4. [Medline].
4. Ruscher KA, Fisher JN, Hughes CD, Neff S, Lerer TJ, Hight DW. National trends in the surgical management of Meckel's diverticulum. J Pediatr Surg. May 2011;46(5):893-6.
5. Elsayes KM, Menias CO, Harvin HJ, Francis IR. Imaging manifestations of Meckel's diverticulum. AJR Am J Roentgenol. Jul 2007;189(1):81-8. [Medline].
6. Chieh-Wen Lin, Internal Herniation of Ileum Caused by Band of sMeckel's Diverticulum. J Med Sci 2011;31(2):079-080.
7. Connolly SA, Drubach LA, Connolly LP. Meckel's diverticulitis: diagnosis with computed tomography and Tc-99m pertechnetate scintigraphy. Clin Nucl Med. Dec 2004;29(12):823-4.
8. Rerksuppaphol S, Hutson JM, Oliver MR. Ranitidine-enhanced 99mtechnetium pertechnetate imaging in children improves the sensitivity of identifying heterotopic gastric mucosa in Meckel's diverticulum. Pediatr Surg Int. May 2004;20(5):323-5. [Medline].

ABOUT THE AUTHORS

Article citation: Chauhan R, Chaudhary R, Dayashankar R. Internal herniation beneath mesodiverticular band producing ambiguous picture in intestinal obstruction: A management dilemma Int J Case Rep Images 2015;6(3):173–177.

Rakesh Chauhan is Assistant Professor Department of Surgery at Dr. Rajendera Prasad Medical College, Kangra at Tanda, Himachal Pradesh, India. He earned undergraduate (MBBS) and postgraduate (MS General Surgery) degrees from Indira Gandhi Medical College, Shimla, Himachal Pradesh, India. His research work include general surgery, gastrointestinal surgery, oncosurgery and hospital administration.
Email: drrakeshchauhan@gmail.com

Rajesh Chaudhary is Junior Resident in Department of Surgery at Dr. Rajendera Prasad Medical College, Kangra at Tanda, Himachal Pradesh, India. He earned undergraduate degree from Indira Gandhi Medical College, Shimla, Himachal Pradesh, India.

R. Dayashankar is Junior Resident in Department of Surgery at Dr. Rajendera Prasad Medical College, Kangra at Tanda, Himachal Pradesh, India.

CLINICAL IMAGES PEER REVIEWED | OPEN ACCESS

An unusual presentation of primary B-cell lymphoma

Salahaddin Ubaid, Iman Ali, Mark Whitsey

CASE REPORT

A young female presented to the accident and emergency department with incidental finding of a chest wall swelling. She was completely asymptomatic, and otherwise fit and well. She was a non-smoker and was not on prescribed medications. There was no family history of serious illnesses. Clinically, she looked well and not distressed. Systemic examination revealed a firm non-mobile painless skin mass in the upper right parasternal chest area measuring about 2x4 cm.

The rest of physical examination was unremarkable. A plain chest X-ray showed widened mediastinum, (Figure 1A). Computed tomography (CT) scan of thorax revealed an approximately 9x4 cm mass with irregular outline in the anterior mediastinum invading the vascular structures of the mediastinum and the anterior chest wall. It also showed a right-sided pleural effusion (Figure 1B). There was no evidence of neck lymphadenopathy, metastasis, interstitial changes or other lung lesions and the rest of the organs were normal. The superior vena cava was patent but flattened by the mass. Pericardial effusion was noted. Incisional biopsy of the mass and subsequent histological examination confirmed the diagnosis of high grade primary mediastinal B-cell lymphoma. She was started on urgent chemotherapy in the form of rituximab with prednisolone, fluconazole, and allopurinol. A repeat CT scan of thorax after six cycles of chemotherapy treatment revealed no convincing evidence of residual or recurrent lymphoma. The patient is in remission and under regular follow-up.

Salahaddin Ubaid[1], Iman Ali[2], Mark Whitsey[3]

Affiliations: [1]ST5 Cardiology New Cross Hospital, Wolverhampton, West Midlands, UK; [2]CMT2, Medicine, New Cross Hospital, Wolverhampton, West Midlands, UK. [3]CMT1, Medicine, New Cross Hospital, Wolverhampton, West Midlands, UK.

Corresponding Author: Salahaddin Ubaid, 7 Barbel Drive, Wolverhampton, West Midlands, United Kingdom, WV10 0TQ; Ph: 00447894856699; Email: saladdinak@yahoo.com

Received: 04 February 2012
Accepted: 04 April 2012
Published: 01 March 2015

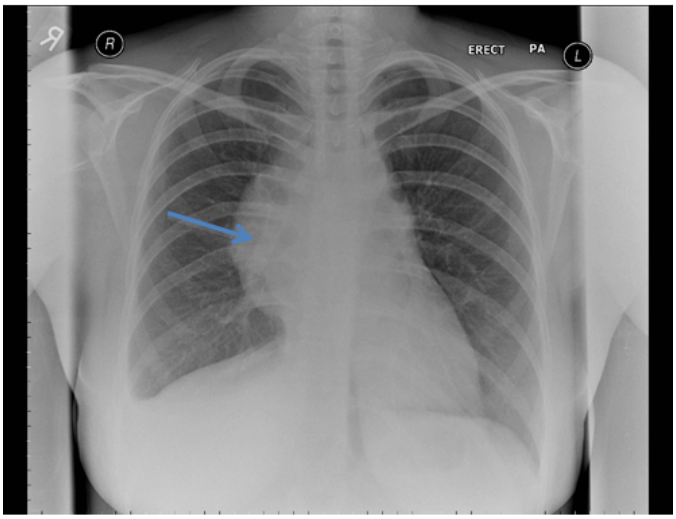


Figure 1A: A chest X-ray showing widened mediastinum.

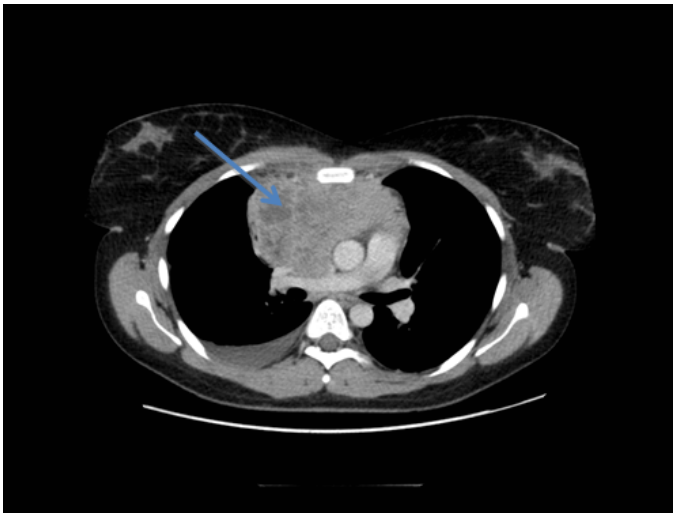


Figure 1B: Computed tomography scan of thorax showing anterior mediastinal mass with right-sided pleural effusion.

DISCUSSION

Primary mediastinal B-cell lymphoma (PMBL) comprises 7% diffuse large B-Cell lymphomas (2.4% of all non-Hodgkin lymphomas). The PMBL is a rapidly growing mediastinal tumor which arises in the thymus and is sometimes palpable in the supraclavicular area. Superior vena cava syndrome is common. Other presentations include phrenic nerve palsy, dysphagia and hoarseness. Shortness of breath can be due to pleural effusion or massive mediastinal mass. Our patient presented with incidental finding of a skin mass and subsequent investigations revealed the diagnosis of anterior mediastinal primary B-cell lymphoma that has invaded the anterior chest wall.

In this type of lymphomas, there is a female predominance and a median age at diagnosis in the third to fourth decade [1]. Patients present with a locally invasive anterior mediastinal mass originating in the thymus, with frequent airway compromise and superior vena cava syndrome [2]. Physical examination may show evidence of superior vena cava obstruction in the form of congested upper limbs and face. A mass could be palpable in the neck. Pleural effusion can be elicited clinically by dullness at lung bases with crepitations. The diagnosis of primary mediastinal large B-cell lymphoma is based on an evaluation of the tumor morphology and immunophenotyping interpreted in the context of the clinical presentation.

CONCLUSION

Patients with primary mediastinal B-cell lymphoma are often symptomatic due to compression of the superior vena cava or invasion of the lungs, pleura and even the pericardium. This case demonstrates how a young female patient was asymptomatic in spite of having advanced disease. The superior vena cava was flattened by the intrathoracic mass and there was a right-sided pleural effusion and pericardial effusion. The mass also invaded the anterior chest wall. In spite of all that, the patient presented with an incidental finding of a palpable skin mass, which is unusual of this type of tumor. It is important to realize that patients with primary B-cell lymphoma can have advanced disease without having typical symptoms at presentations.

How to cite this article

Ubaid S, Ali I, Whitsey M. An unusual presentation of primary B-cell lymphoma. Int J Case Rep Images 2015;6(3):178–180.

doi:10.5348/ijcri-201508-CL-10063

****

Author Contributions

Salahaddin Ubaid – 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

Iman Ali – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Mark Whitsey – Substantial contributions to conception and design, Acquisition of data, 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. Nguyen LN, Ha CS, Hess M, et al. The outcome of combined-modality treatments for stage I and II primary large B-cell lymphoma of the mediastinum. Int J Radiat Oncol Biol Phys 2000 Jul 15;47(5):1281–5.
2. Van Besien K, Kelta M, Bahaguna P. Primary mediastinal B-cell lymphoma: A review of pathology and management. J Clin Oncol 2001 Mar 15;19(6):1855–64.

CLINICAL IMAGES PEER REVIEWED | OPEN ACCESS

Usefulness of three-dimensional computed tomography reconstruction of incisional hernia for planning laparoscopic hernia repair

Hiroto Kayashima, Takashi Maeda, Noboru Harada, Teruyoshi Ishida

CASE REPORT

We report on two incisional hernia's cases in which preoperative three-dimensional computed tomography (3D-CT) reconstruction images were very useful for planning their laparoscopic surgeries. Patient 1 was a 78-year-old male who had been performed partial hepatectomy for hepatocellular carcinoma with J-shaped incision two years before and developed a midline incisional hernia. The preoperative 3D-CT using SYNAPSE VINCENT (Fuji Photo Film Co. Ltd., Tokyo, Japan) showed the defect measuring 4.3×2.9 cm (Figure 1A). The laparoscopic incisional hernia repair was performed. Intraoperative hernia size was 4.3 × 2.8 cm. We repaired the defect with 12.0×12.0 cm Parietex™ Optimized Composite Mesh (Nippon Covidien Inc., Tokyo, Japan). The patient tolerated the procedure well with no complication. Patient 2 was a 64-year-old male who had received central bisegmentectomy for hepatocellular carcinoma two years before and presented with a large incisional hernia of the lateral aspect of the right subcostal incision, and the content of hernia was almost intestines. The hernia size was 14.8×12.9 cm measuring by the preoperative 3D-CT (Figure 1B) and the actual intraoperative hernia size was 15.0×13.0 cm. We performed a laparoscopic incisional hernia repair with 20.0×15.0 cm Parietex™ Optimized Composite Mesh and the patient was discharged without any complication and hernia recurrence is not occurred at this time.

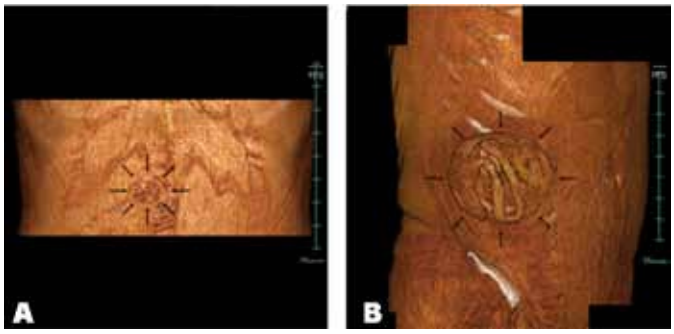


Figure 1: The three-dimensional computed tomography reconstruction images of the incisional hernias. (A) The image showing the midline incisional hernia of Patient 1 (Black arrows). (B) The image showing the right subcostal incisional hernia of Patient 2 (Black arrows). The content was almost intestines.

DISCUSSION

Incisional hernia is a frequent complication of laparotomy that occurs in up to 11% of surgical abdominal wounds and in up to 20% of patients who develop postoperative wound infections [1]. Since the first report in 1993, laparoscopic incisional hernia repair has led to improved results with a low recurrence rate of 4.2%, with an impressive conversion rate of only 2.4%, and enterotomy rate of 1.8% [2]. Recent report about the comparison between laparoscopic and open incisional hernia repair demonstrated that the incidences of wound infection (2.8% versus 16.2%) and the rates of wound drainage (2.6% versus 67.0%) were significantly lower in the laparoscopic group and there were no significant differences in the incidences of hernia recurrence, postoperative seroma, hematoma, bowel obstruction, bleeding, and reoperation [3]. Laparoscopic approach of incisional hernia repair is based on two technical principles: a hernia gate that is not closed and the proper sized mesh placed intraperitoneally. Therefore, accurate

Hiroto Kayashima[1], Takashi Maeda[1], Noboru Harada[1], Teruyoshi Ishida[1]

Affiliations: [1]MD, Ph.D, Department of Surgery, Hiroshima Red Cross Hospital and Atomic Bomb Hospital, 1-9-6 Senda-cho, Naka-ku, Hiroshima 730-8619, Japan.

Corresponding Author: Hiroto Kayashima, MD, PhD, Department of Surgery, Hiroshima Red Cross Hospital and Atomic Bomb Hospital, 1-9-6 Senda-cho, Naka-ku, Hiroshima 730-8619, Japan; Ph: +81-082-241-3111, Fax: +81-082-246-0676; Email: hkaya@surg2.med.kyushu-u.ac.jp

Received: 30 October 2014
Accepted: 12 December 2014
Published: 01 March 2015

measurement of hernia size is important for preoperative choice of patch size. Some papers reported that 3D-CT reconstruction could identify abdominal wall defects and hernia contents more clearly compared with plain CT scans, and that diagnosis of hernia was easy based on 3D-CT [4]. Recent 3D-CT reconstruction software represents a marked improvement and is capable of displaying 3D-CT images within a few minutes using a single click. In fact, the 3D-CT images of our two cases could clearly display abdominal wall defects and directly obtain hernia sizes in a short time. Briefly, 3D-CT reconstruction images are more useful to display abdominal wall defects, to know if the hernia gate is not closed, and to obtain hernia size than ordinary CT. There is a general consensus on the patch size that extends 3 cm or 5 cm beyond the edges of hernia, therefore, we chose 12.0×12.0 cm mesh for Patient 1. In subcostal incisional hernias, it had been reported that the close proximity to the xiphoid process and the costochondral structures hindered adequate coverage of the defect, and the role of laparoscopic approach was still controversial because of its low prevalence, however, recent reports described that the laparoscopic approach was a safe and effective treatment for non-midline incisional hernias [5]. Therefore, the large right subcostal incisional hernia of Patient 2 was repaired by laparoscopic approach using 20.0×15.0 cm mesh. Although the patient was discharged without any complications, there is a need for a careful follow-up.

CONCLUSION

Three-dimensional computed tomography reconstruction of incisional hernia can be displayed within a few minutes and calculate the hernia size with considerable accuracy. It is of great use for planning the laparoscopic approach for incisional hernia repair.

How to cite this article

Kayashima H, Maeda T, Harada N, Ishida T. Usefulness of three-dimensional computed tomography reconstruction of incisional hernia for planning laparoscopic hernia repair. Int J Case Rep Images 2015;6(3):181–183.

doi:10.5348/ijcri-201509-CL-10064

****

Author Contributions

Hiroto Kayashima – 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

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

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

Teruyoshi Ishida – 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. Mudge M, Hughes LE. Incisional hernia: A 10 year prospective study of incidence and attitudes. Br J Surg 1985 Jan;72(1):70–1.
2. LeBlanc KA. Incisional hernia repair: Laparoscopic techniques. World J Surg 2005 Aug;29(8):1073–9.
3. Zhang Y, Zhou H, Chai Y, Cao C, Jin K, Hu Z. Laparoscopic versus open incisional and ventral hernia repair: A systematic review and meta-analysis. World J Surg 2014 Sep;38(9):2233–40.
4. Yao S, Li JY, Liu FD, Pei LJ. Significance of measurements of herniary area and volume and abdominal cavity volume in the treatment of incisional hernia: Application of CT 3D reconstruction in 17 cases. Comput Aided Surg 2012;17(1):40–5.
5. Moreno-Egea A, Carrillo-Alcaraz A. Management of non-midline incisional hernia by the laparoscopic approach: Results of a long-term follow-up prospective study. Surg Endosc 2012 Apr;26(4):1069–78.

CLINICAL IMAGES PEER REVIEWED | OPEN ACCESS

A rare cause of acute pancreatitis: Groove pancreatitis

Yusuf Kayar, Mehmet Yigit, Iskender Ekinci, Kenan Ahmet Turkdogan

CASE REPORT

A 44-year-old female was admitted to emergency department with abdominal pain that starts 2 days ago and gradually increases after meals. She has no history of alcohol consumption but she smoked one pack of cigarettes daily for twenty years. Epigastric tenderness was revealed on physical examination. Laboratory examination showed leukocytosis, elevated serum levels of amylase and lipase (20.0x10^3/µL, 697 IU/l and 893 IU/l, respectively). A contrast enhanced computerized tomography scan of the abdomen showed swelling of the pancreatic uncinate, thickening of the distal segment of the second part and proximal segment of third part of the duodenum (Figure 1). Peripancreatic fluid was also seen. Upper gastrointestinal endoscopy revealed an edematous, reddish raised duodenal mucosa and stenosis of the descending part of the duodenum. Histological examination of biopsy obtained from duodenal mucosa revealed hyperplastic Brunner's glands and malignancies were excluded. Thickening of the duodenal wall, a pancreatic head compatible with pancreatitis and peripancreatic adenopathies were observed on endoscopic ultrasound examination (Figure 2). These findings appeared consistent with the diagnosis of groove pancreatitis. The patient was treated conservatively with fluid replacement (200 cc/h ringer lactate), analgesics

Yusuf Kayar[1], Mehmet Yigit[2], Iskender Ekinci[3], Kenan Ahmet Turkdogan[2]

Affiliations: [1]Department of Gastroenterology, Bezmialem Vakif University, Istanbul, Turkey; [2]Department of Emergency Medicine, Bezmialem Vakif University, Istanbul, Turkey; [3]Department of Internal Medicine, Bezmialem Vakif University, Istanbul, Turkey.

Corresponding Author: Mehmet Yigit, Department of Emergency Medicine, Bezmialem Vakif University, Istanbul, Turkey; GSM: +90 505 747 21 63; Email: yigitacil@gmail.com

Received: 04 November 2014
Accepted: 06 December 2014
Published: 01 March 2015

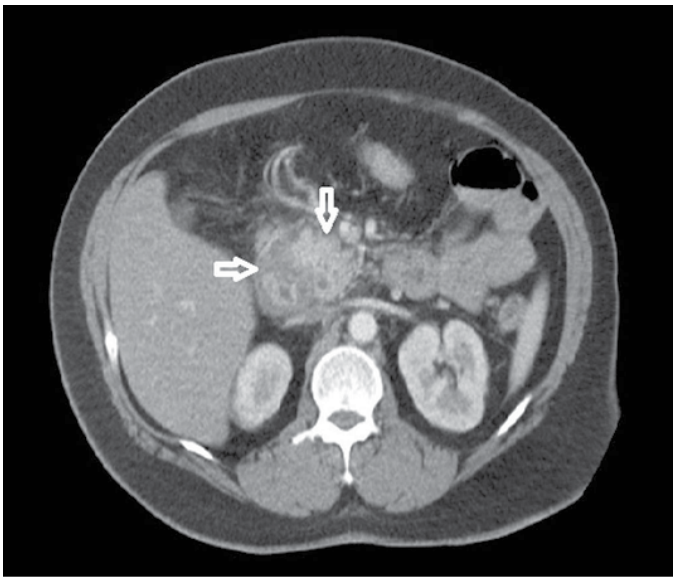


Figure 1: A contrast enhanced computed tomography scan of the abdomen showing swelling of the pancreatic uncinate, thickening of the distal segment of the second part and proximal segment of third part of the duodenum.

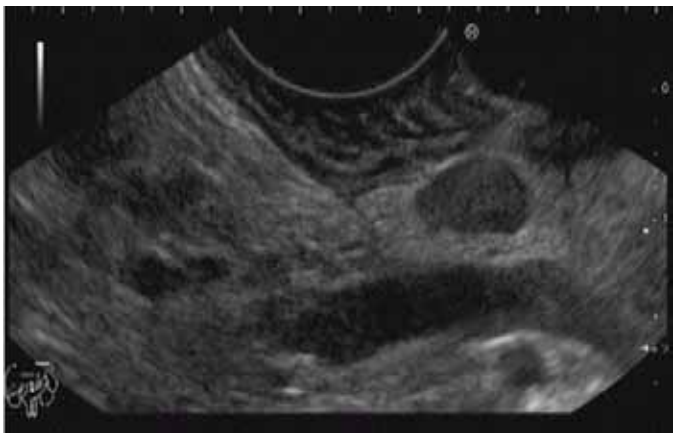


Figure 2: Thickening of the duodenal wall, a pancreatic head compatible with pancreatitis and peripancreatic adenopathies were observed on endoscopic ultrasound examination.

(0.5 mg fentanyl 2x1) and proton pump inhibitors (40 mg pantoprazole 1x1). On the second day leukocyte, amylase and lipase levels of the patients started to decrease ($13.4x10^3/\mu L$, 286 IU/l and lipase is 334 IU/l, rerspectively) were decrease. On third and fourth day, all laboratory examinations were in normal range. On day fourth the patient was totally healed and discharged with recommendations.

DISCUSSION

Groove pancreatitis is a segmental form of chronic pancreatitis that affects the 'groove area' which is defined as the area between the head of pancreas, duodenum and common bile duct and it is often diagnosed in 40 to 50-year-old alcoholic men [1]. But the incidence of groove pancreatitis in younger individuals and women is considerably lower [2]. Brunner's gland hyperplasia gives rise to the stasis of the pancreatic juice in the dorsal pancreas additionally the viscosity changes of the pancreatic fluid due to the excessive alcohol consumption and/or smoking. These changes lead to pancreatitis in the groove area [1]. Also the cause of this condition can be a history of gastrectomy, a gastroduodenal ulcer, biliary diseases and the presence of anatomic abnormalities which causing minor papilla dysfunction [3]. Subsequent clinic manifestations may occur in groove pancreatitis; abdominal pain, postprandial vomiting, weight loss, nausea and vomiting and jaundice. The stenosis of the second part of the duodenum due to thickening and scarring of the duodenal wall and cystic changes in the thickened duodenal wall are the pathological findings of the groove pancreatitis. On microscopic examination, thickened submucosa and muscle layers secondary to fibrosis and Brunner's gland hyperplasia are seen [3]. Upper gastrointestinal endoscopy reveals stenosis secondary to edema and an inflamed and polypoid appearance in the descending part of the duodenum. The radiological images generally show a mass between the pancreatic head and duodenum. Endoscopic ultrasonography shows smooth tubular stenosis of the common bile duct without abnormality of the main pancreatic duct [4]. Although these findings suggest groove pancreatitis, a biopsy should be done to rule out malignancy. Conservative therapy including the cessation of smoking / alcohol consumption, recovery of pancreatic function and analgesics or surgical treatment comprising pancreaticoduodenectomy and pylorus-preserving pancreaticoduodenectomy can be preferred in the treatment of the groove pancreatitis but surgery is often recommended to exclude malignancy [3].

CONCLUSION

Groove pancreatitis is a rare cause of acute pancreatitis and it should be kept in mind because it can mimic the pancreatic adenocarcinoma. Clinical signs may improve with conservative treatment but a surgical procedure should be preferred with suspicion of malignancy.

How to cite this article

Kayar Y, Yigit M, Ekinci I, Turkdogan KA. A rare cause of acute pancreatitis: Groove pancreatitis. Int J Case Rep Images 2015;6(3):184–186.

doi:10.5348/ijcri-201510-CL-10065

****

Acknowledgements
We would like to acknowledge the patient who kindly allowed us to publish her history.

Author Contributions
Yusuf Kayar – Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Mehmet Yigit – Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published
Iskender Ekinci – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Kenan Ahmet Turkdogan – Acquisition of data, 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. Stolte M, Weiss W, Volkholz H, Rosch W. A special form of segmental pancreatitis: "groove pancreatitis". Hepatogastroenterology 1982 Oct;29(5):198–208.

2. Triantopoulou C, Dervenis C, Giannakou N, Papailiou J, Prassopoulos P. Groove pancreatitis: A diagnostic challenge. Eur Radiol 2009 Jul;19(7):1736–43.
3. Tezuka T, Makino T, Hirai I, Kimura W. Groove pancreatitis. Dig Surg 2010;27(2):149–52.
4. Balakrishnan V, Chatni S, Radhakrishnan L, Narayanan VA, Nair P. Groove pancreatitis: A Case Report and Review of Literature. JOP 2007 Sep 7;8(5):592–7.

ABOUT THE AUTHORS

Article citation: Kayar Y, Yigit M, Ekinci I, Turkdogan KA. A rare cause of acute pancreatitis: Groove pancreatitis. Int J Case Rep Images 2015;6(3):184–186.

Yusuf Kayar is Internal Medicine specialist at Department of Gastroenterology, Bezmialem Vakif University, Istanbul, Turkey. His research interests include hepatobiliary system, and gastroscopy, pancreatitis. He has published 10 research papers in national and international academic journals.

Mehmet Yigit is Emergency Medicine specialist at Department of Emergency Medicine, Bezmialem Vakif University, Istanbul, Turkey. His research interests include trauma, pancreatitis, stroke and acute coronary syndrome. He has published 23 research papers in national and international academic journals.

Iskender Ekinci is assistant at Department of Internal Medicine, Bezmialem Vakif University, Istanbul, Turkey. His research interests include acute renal failure and chronic renal failure, gastroenterology. He has published three research papers in national and international academic journals.

Kenan Ahmet Turkdogan is Emergency Medicine specialist at Department of Emergency Medicine, Bezmialem Vakif University, Istanbul, Turkey. His research interests include trauma, crimean-congo hemorrhagic fever, acute coronary syndrome and toxicology. He has published 38 research papers in national and international academic journals.

CLINICAL IMAGES PEER REVIEWED | OPEN ACCESS

The lung riddle: A Blesovsky's syndrome case

Aviral Vij, Anshu Singh

CASE REPORT

A 49-year-old male presented with worsening shortness of breath and reduction in exercise tolerance over eight months. Review of systems was negative. Past history was significant for untreated hypertension, a 40 pack-year smoking history and work in construction, where he was exposed to asbestos and silica for over 20 years without use of masks. Vital signs were relevant for blood pressure of 159/94 mmHg. Physical examination revealed reduced breath sounds in the left infra axillary area. Rest of the examination was unremarkable. Chest X-ray showed a small, well-circumscribed pleural-based density in the left lower zone. A subsequent computed tomography (CT) scan of the chest showed a rounded, well-defined, 14x17 mm pleural based mass with pleural thickening in the posterior segment of the left lower lung which was identified as 'rounded atelectasis'. At sixth month follow-up, the CT scan findings remained unchanged and his symptoms had improved with treatment of underlying chronic obstructive pulmonary disease with salmeterol and tiotropium inhalers.

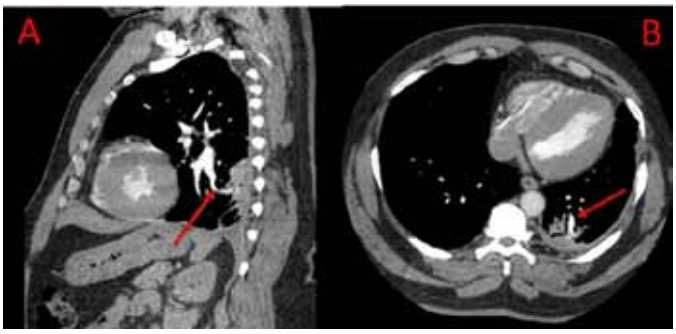


Figure 1: Computed tomography scan of the chest with contrast (at the time of presentation). (A): Typical "Cornet tail" sign with bronchovascular bundies extending from hilum to the area of Rounded atelectasis (red arrow), and (B): Air bronchogram in the center of the area of Rounded atelectasis (red arrow).

DISCUSSION

Rounded atelectasis (RA) is a benign process, incidentally found on follow-up X-ray of patients with known occupational exposure or on routine testing. It has been associated with several etiologies [1] like exposure to mineral dust of asbestos (most common cause) and silica, exudative pleural effusions like tuberculosis and para-pneumonic effusions, infections like *Legionella pneumonia* and histoplasmosis and also heart failure. Rounded atelectasis is a radiological diagnosis and characteristic findings on CT scan include [2]:

(i) Rounded pleural-based mass, 4-7 cm in diameter, which forms an acute angle with the pleura and is never completely surrounded by the lung.

(ii) Pleural thickening.

(iii) Bronchovascular markings extending from the hilum to the "mass" with characteristic "Cranial tilting" and the "Comet Tail sign [3]", which is highly specific and often considered pathognomic of RA.

(iv) Air bronchogram, usually in the central part of the mass.

As in our patient, RA can be diagnosed with characteristic CT scan. Atypical radiographic presentations may require additional testing such as MRI scan, PET scan or invasive procedures such as VATS or thoracotomy. Absence of lymphadenopathy with minimal to no pleural effusion and metabolically inactive mass seen on PET scan can help differentiate Rounded atelectasis from the most common differential which is malignancy.

Aviral Vij[1], Anshu Singh[2]

Affiliations: [1]MBBS, MD, Internal Medicine Resident at John H. Stroger Hospital of Cook County, Chicago, IL; [2]MBBS, MD, Attending Physician and Firm Chief at John H. Stroger Hospital of Cook County, Chicago, IL.

Corresponding Author: Aviral Vij, MBBS, MD, Internal Medicine Resident at John H. Stroger Hospital of Cook County, Chicago, IL; Email: aviral.vij@gmail.com

Received: 10 November 2014
Accepted: 17 December 2014
Published: 01 March 2015

RA by itself is benign and need not be followed, however, such patients often are exposed to asbestos and should be followed-up considering the malignant potential of asbestosis. Awareness of this radiographic entity can help avoid invasive procedures in patients with typical CT scan findings.

There is no specific treatment for rounded atelectasis and management is usually symptom guided. Surgical resection of the mass is rarely indicated, when there is significant compromise of lung function or if there is high suspicion of malignancy which could not be ruled out by other diagnostic modalities.

CONCLUSION

Rounded atelectasis is a rare but increasingly recognized entity due to enhanced screening especially in patients with exposure to asbestosis. Radiographic features are often typical but in the presence of atypical features or in equivocal cases, malignancy must be ruled out.

How to cite this article

Vij A, Singh A. The lung riddle: A Blesovsky's syndrome case. Int J Case Rep Images 2015;6(3):187–188.

doi:10.5348/ijcri-201511-CL-10066

****

Author Contributions

Aviral Vij – 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

Anshu Singh – Substantial contributions to conception and design, Acquisition 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. Stathopoulos GT, Karamessini MT, Sotiriadi AE, Pastromas VG. Rounded atelectasis of the lung. Respir Med 2005 May;99(5):615–23.
2. Doyle TC, Lawler GA. CT features of rounded atelectasis of the lung. AJR Am J Roentgenol 1984 Aug;143(2):225–8.
3. Partap VA. The Comet tail sign. Radiology 1999 Nov;213(2):553–4.

CLINICAL IMAGES PEER REVIEWED | OPEN ACCESS

Radiculomegaly of permanent canines and first premolars: Report of two cases in conjunction with oculo-facio-cardio-dental syndrome

Masayasu Iwase, Hiroaki Nishijima, Gen Kondo, Michiko Ito

CASE REPORT

Case 1: An 18-year-old Japanese female was referred to us for the improvement of malocclusion (Figure 1A). The patient was born at 38 weeks by cesarean section; her birth weight was 2,990 g and she was 48 cm in length. At birth, she was noted to have multiple congenital anomalies, including bilateral cataracts, atrial septal defect, long narrow face, high nasal bridge, broad nasal tip with separated cartilages, and a long philtrum. Bilateral cataracts were surgically removed at 1 year. She, however, suffered resultant secondary glaucoma and her visual impairment still remained. At the age of 1 year 8 months, she received an operation for cleft palate. At the age of 18 years 6 months, an operation was undertaken for atrial septal defect. Facial findings showed a somewhat asymmetric face, bilateral strabismus, high nasal bridge, and broadness at the tip of the nose at the initial consultation. Her intelligence was not tested, but may have exceeded the average level. The intraoral aspects showed malocclusion, multiple dental caries, delayed eruption of permanent teeth, high palate, and narrow arched maxilla. On a panoramic radiograph, tooth length and radiculomegaly in the lower canines and first premolars had significantly increased (Figure 1B). The cephalogram analytical data showed marked craniofacial dysmorphology, characterized by an extremely decreased anterior and posterior cranial base and decreased midfacial depth with a short midfacial height. The mandible was characterized by a remarkably large gonial angle and small lower facial depth. Both the anterior maxillary and mandibular heights had increased. Malocclusion was characterized by a bimaxillary skeletal discrepancy with maxillary undergrowth and oligodontia. We planned to undertake orthognathic surgery and prosthodontic treatment for the improvement of the craniofacial dysmorphology and skeletal class III malocclusion. Subsequently, at the age of 19 years 10 months, she underwent sagittal split ramus osteotomy (Figure 1C). Furthermore, she received prosthodontic treatment because of impairment of tooth form.

Masayasu Iwase[1], Hiroaki Nishijima[2], Gen Kondo[2], Michiko Ito[3]

Affiliations: [1]Department of Dentistry and Oral Surgery, Hakujikai Memorial General Hospital, 5-11-1 Shikahama, Adachi-ku, Tokyo, Tokyo, 123-0864, Japan; [2]Department of Dentistry and Oral Surgery, Jinkokai Hospital, 3-8-11, Nakamachi, Atsugi, Kanagawa, 243-0018, Japan; [3]Department of Oral and Maxillofacial Surgery, School of Dental Medicine, Tsurumi University, 2-1-3, Tsurumi, Tsurumi-ku, Yokohama, Kanagawa, 230-8501, Japan.

Corresponding Author: Masayasu Iwase, Department of Dentistry and Oral Surgery, Hakujikai Memorial General Hospital, 5-11-1 Shikahama, Adachi-ku, Tokyo 123-0864, Japan; Mob: 81338991311, Fax: 81338552851; Email: iwase@dent.showa-u.ac.jp

Received: 20 December 2014
Accepted: 19 January 2015
Published: 01 March 2015

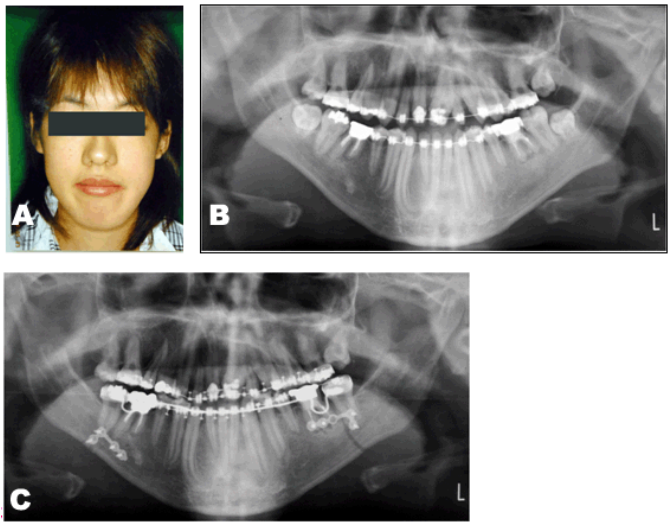


Figure 1: (A) Photograph of patient 1 at the age of 18 years, (B) Panoramic radiograph of patient 1 at the age of 18 years, The lower canines and first premolars conspicuously showed increased tooth length and radiculomegaly, (C) Panoramic radiograph of patient 1 at the age of 19 years after sagittal split ramus osteotomy. Mandibular prognathism was improved by the orthognathic surgery.

Case 2: A 6-year-old Japanese girl was referred to us for crossbite of the incisors. After a normal pregnancy, she was born at term with a birth weight of 3,040 g and length of 47 cm. At birth, the patient was noted to have multiple congenital anomalies. She had bilateral congenital cataracts and secondary glaucoma. A congenital heart defect was excluded by an echocardiographic examination and a ventricular septal defect has been monitored by a cardiologist since she was 12 years of age, with no further cardiac symptoms. Surgical correction for the cleft palate was performed at the age of 1 year 4 months. Her stature was small, below average for her age. The patient had a narrow face, high nasal bridge, separated cartilages of the nasal tip, broad chin, and mild ptosis at the initial consultation. Her intellectual development was mildly retarded. Delayed dental eruptions were evident. Agenesis of the permanent incisor teeth of the mandible was found by a panoramic radiograph at the age of six years (data not shown). In addition, conspicuously wide open root apices and root dilacerations were noticed in the teeth that later developed canine radiculomegaly (Figure 2). At the age of six years, she started orthodontic treatment and is at present continuing the treatment. Similar to Case 1, we intend to perform orthognathic surgery for the improvement of her jaw deformity in the near future.

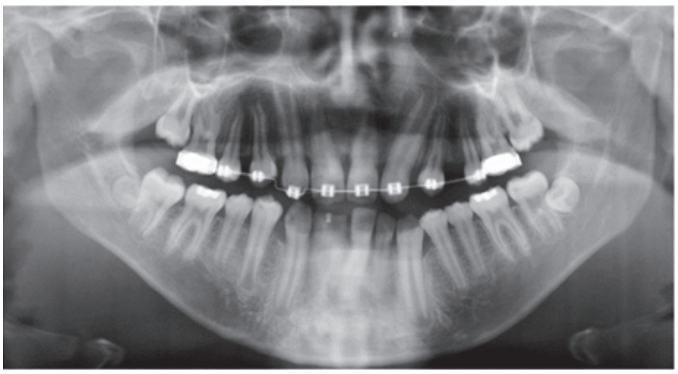


Figure 2: Panoramic radiograph of Patient 2 at the age of 14 years. Conspicuously wide open root apices and root dilacerations were noticed in the teeth that developed canine radiculomegaly.

DISCUSSION

Gorlin et al. have referred to a combination of congenital cataracts, facial characteristic appearance, cardiac anomalies, and typical dental findings as oculo-facio-cardio-dental (OFCD) syndrome [1]. A previous study reported that OFCD syndrome relates to an X-linked dominant trait [1]. X-linked dominant conditions with male hemizygosity can become lethal [2]. In this case report, although we could not obtain inherent evidence, the presented patients with OFCD syndrome were also females.

Most affected patients have hallmark dental anomalies, including radiculomegaly with prolonged dental roots and widely open apices, most typically in the canine roots [3–5]. Additional dental anomalies include delayed eruption of both deciduous and permanent teeth, persistence of deciduous teeth, and oligodontia. Previous reports have pointed out that the most important criteria leading to the diagnosis of OFCD syndrome are dental abnormalities, in particular extreme elongation of the cuspids' roots [1]. Radiculomegaly of incisors, canines, and premolars is a typical finding of OFCD [3–5]. Even in adult patients, the affected teeth show vast extension of the root and open apices [1]. Although the average tooth lengths of the normal mandibular canine and first premolar in Japanese are about 24 mm and 21 mm, respectively [4], the lengths of these abnormal teeth are more than 50 mm in the presented cases. Diagnosis can only be confirmed between 15 and 20 years of age, when the radiculomegaly becomes evident [3]. Histological study of canines accompanied with radiculomegaly has shown dentine formation disorder and thin enamel [6]. Ng et al. clearly found that OFCD syndrome is caused by mutations in the BCL6 co-repressor (BCOR) gene [7]. A recent study has suggested that mutation of periodontal ligament cells in the BCOR may contribute to hypersensitive root formation [8]. Although ocular, facial, and cardiac findings are found at an early age, the dental findings cannot be expected to appear simultaneously. Therefore, it is difficult for clinicians to confirm the diagnosis of OFCD syndrome until the development of dental findings, including radiculomegaly, delayed dentition, oligodontia, root dilacerations, and open root apex.

It is troublesome for OFCD syndrome patients to undergo proper dental treatment in endodontic treatment, tooth extraction, and orthodontic treatment [4, 5]. We only limited the orthodontic treatment to the improvement of malocclusion in these presented patients; we performed orthognathic surgery and prosthodontic treatment.

CONCLUSION

We report two new cases of female patients with radiculomegaly of canines and premolars associated with oculo-facio-cardio-dental (OFCD) syndrome.

How to cite this article

Iwase M, Nishijima H, Kondo G, Ito M. Radiculomegaly of permanent canines and first premolars: Report of two cases in conjunction with oculo-facio-cardio-dental syndrome. Int J Case Rep Images 2015;6(3):189–192.

doi:10.5348/ijcri-201512-CL-10067

****

Author Contributions

Masayasu Iwase – 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
Hiroaki Nishijima – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Gen Kondo – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Michiko Ito – 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.

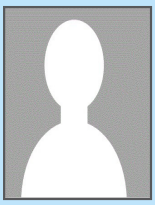
Copyright

REFERENCES

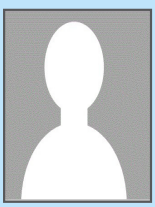
1. Gorlin RJ, Marashi AH, Obwegeser HL. Oculo-facio-cardio-dental (OFCD) syndrome. Am J Med Genet 1996 May 3;63(1):290–2.
2. Wieacker P, Zimmer J, Ropers HH. X inactivation patterns in two syndromes with probable X-linked dominant, male lethal inheritance. Clin Genet 1985 Sep;28(3):238–42.
3. Barthelemy I, Samuels L, Kahn DM, Schendel SA. Oculo-facio-cardio-dental syndrome: Two new cases. J Oral Maxillofac Surg 2001 Aug;59(8):921–5.
4. Kawamoto T, Motohashi N, Ohyama K. A case of oculo-facio-cardio-dental syndrome with integrated orthodontic-prosthdontic treatment. Cleft Palate Craniofac J 2004 Jan;41(1):84–94.
5. Maden M, Savgat A, Görgül G. Radiculomegaly of permanent canines: Report of endodontic treatment in OFCD syndrome. Int Endod J 2010 Dec;43(12):1152–61.
6. Larhant M, Sourice S, Grimaud F, et al. Giant canine with dentine anomalies in oculo-facio-cardio-dental syndrome. J Craniomaxillofac Surg 2014 Jun;42(4):321–4.
7. Ng D, Thakker N, Corcoran CM, et al. Oculofaciocardiodental and Lenz microphthalmia syndromes result from distinct classes of mutations in BCOR. Nat Genet 2004 Apr;36(4):411–6.
8. Surapornsawasd T, Ogawa T, Tsuji M, Moriyama K. Oculofaciocardiodental syndrome: novel BCOR mutations and expression in dental cells. J Hum Genet 2014 Jun;59(6):314–20.

ABOUT THE AUTHORS

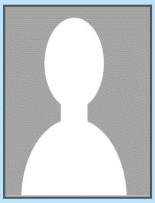
Article citation: Iwase M, Nishijima H, Kondo G, Ito M. Radiculomegaly of permanent canines and first premolars: Report of two cases in conjunction with Oculo-Facio-Cardio-Dental syndrome. Int J Case Rep Images 2015;6(3):189–192.



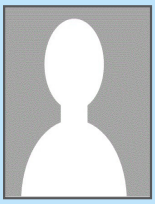
Masayasu Iwase is Chief in Department of Dentistry and Oral Surgery at Hakujikai Memorial General Hospital, Tokyo, Japan. He earned graduate degree (DDS) from Showa University, Tokyo, Japan and postgraduate degree form (PhD) from Showa University, Tokyo, Japan. He studied oral infection at Department of Pathology, School of Dental Medicine, University of Pennsylvania, PA, USA.
He has published more than 90 research papers in national and international academic journals and authored more than 10 books. His research interests include head and neck cancer, orthognathic surgery and oral infection.



Hiroaki Nishijima is Clinical Assistant in Department of Dentistry and Oral Surgery at Jinkokai Hospital, Atsugi, Japan. He earned graduate degree (DMD) from Tsurumi University, Yokohama, Japan. His research interests include maxillofacial surgery.



Gen Kondo is Chief in Department of Dentistry and Oral Surgery, Atsugi, Japan. He eared graduate degree (DDS) from Oou University, Kooriyama, Japan and postgraduate degree (PhD) from Showa University, Tokyo, Japan. He has published 20 research papers in national and international academic journals. His research interests include oral cancer and dental implant.



Michiko Ito is Clinical Fellow at Department of Oral and Maxillofacial Surgery, School of Dental Medicine, Tsurumi University, Yokohama, Japan. She earned graduate degree (DMD) from Tsurumi University, Yokohama, Japan. Her research interests include maxillofacial surgery.