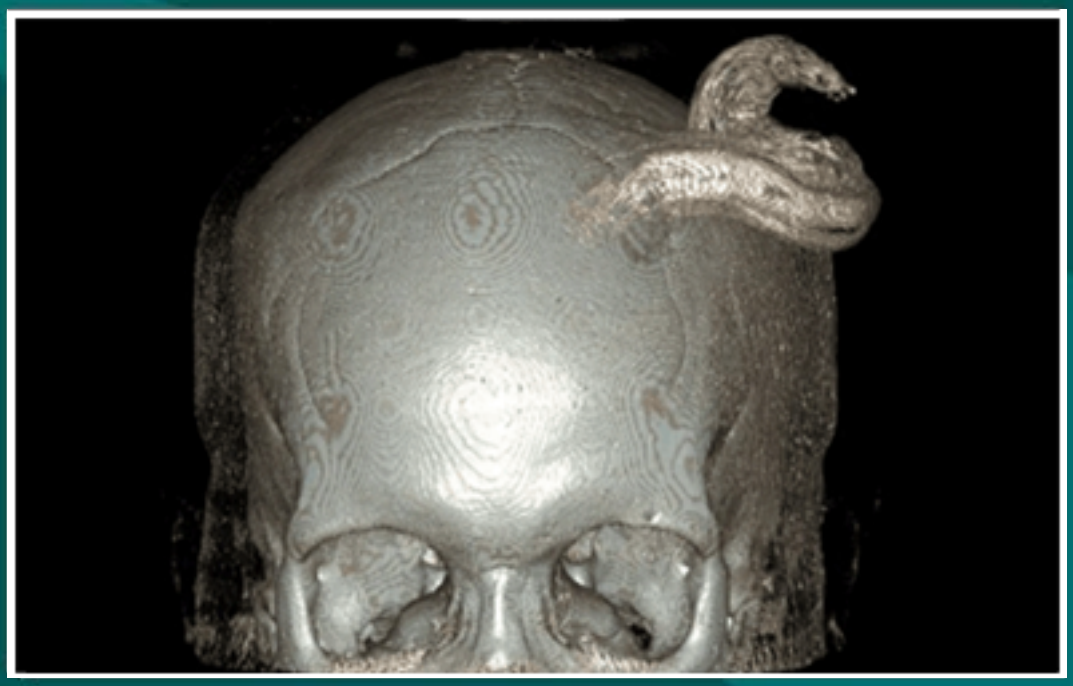
May 2016; Volume 7; Number 5

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

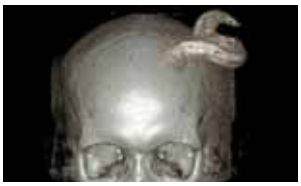
Guidelines for Authors

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. 7, No. 5 (May 2016)

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 SERIES

PEER REVIEWED | OPEN ACCESS

Migration of endoluminal gastroesophageal stents: A case series

Kent C. Sasse, David L. Warner, Jared Brandt, Ellen Ackerman

ABSTRACT

Introduction: Microperforation of the stomach following bariatric surgical procedures is often treated with endoluminal stent placement. Endoluminal stents migrate out of the desired position in a high frequency of cases. Case Series: This paper reports a series of five cases in which endoluminal stents migrated antegrade into unfavorable positions. One of the cases resulted in the stent migrating into the jejunum where it resulted in a jejunal perforation requiring surgery and bowel resection. After endoscopic repositioning of the stents, endoluminal suture fixation resulted in stabilization of the stents, prevented further migration, and facilitated clinical resolution of gastric fistula. No complications of the endoluminal suture fixation to the esophageal wall occurred, and all patients recovered fully. Conclusion: This paper presents five cases of migrated gastroesophageal stents that were successfully secured with endoluminal sutures without complications. Endoluminal suturing may be a technically straightforward and useful solution to the migration of gastroesophageal stents.

Keywords: Minimally invasive surgery, Microleak, Stent migration, Sleeve gastrectomy

Kent C. Sasse[1], David L. Warner[1], Jared Brandt[1], Ellen Ackerman[1]

Affiliations: [1]University of Nevada School of Medicine.

Corresponding Author: David Warner, BS, University of Nevada School of Medicine, 75 Pringle Way, Suite 804, Reno, NV 89502; Email: dwarner@medicine.nevada.edu

Received: 03 November 2015
Accepted: 19 February 2016
Published: 01 May 2016

How to cite this article

Sasse KC, Warner DL, Brandt J, Ackerman E. Migration of endoluminal gastroesophageal stents: A case series. Int J Case Rep Imag 2016;7(5):287–291.

Article ID: Z01201605CR10068KS

****

doi:10.5348/ijcri-201607-CS-10068

INTRODUCTION

Gastric microperforation following bariatric surgical procedures is often treated with endoluminal esophagogastric stent placement [1–4]. Stent migration is a common occurrence, reportedly occurring in 30–50% of stent placements following sleeve gastrectomy micro leaks [2, 5, 6]. In most cases, antegrade stent migration is treated with repeat endoscopy and repositioning of the stent [5]. In rare cases, however, stent migration may result in intestinal injury or perforation and require surgery. Methods of stent fixation within the lumen have been proposed and include use of clips and the use of endoluminal suturing devices [7–10]. We present five cases of stent migration including one resulting in jejunal perforation, and we report our experience with the use of endoluminal suturing to secure endoluminal stents and prevent migration.

CASE SERIES

Case 1

A 56-year-old male with morbid obesity who underwent a laparoscopic sleeve gastrectomy in Mexico

two weeks prior to presentation at our center with sepsis due to a leak at the proximal staple line. He underwent a laparoscopic washout and drainage procedure followed by endoscopic placement of a fully covered endoluminal esophagogastric stent (fully-covered 150x23 mm WallFlex, Boston Scientific). His sepsis resolved, and improved clinically, tolerating clear liquids orally while being primarily nourished with total parenteral nutrition. After 15 days after of stent placement he developed abdominal pain and vomiting. Computed tomography scan demonstrated the stent had migrated beyond the pylorus to the jejunum (Figure 1). He developed increased pain and fever and underwent laparotomy at which time he was found to have a perforation of the jejunum due to the migrated stent. He underwent a resection of a segment of jejunum as well as removal of the stent. An attempt at suture closure and omental patching of the proximal gastric fistula was performed.

In the days following his recovery from laparotomy, the gastric fistula was evident from the drain output. He underwent repeat endoscopy and placement of a new endoluminal stent (fully-covered 150x23 mm WallFlex, Boston Scientific), this time secured to the esophageal wall with two endoluminal sutures through the proximal stent flange and the esophageal wall (absorbable suture Apollo Overstitch). For the next 12 weeks, the stent remained in position with the patient tolerating oral liquids and nourished with TPN. Repeat endoscopy was then performed and the stent was removed. The patient was able to advance to a regular diet, and no further gastric fistula recurred.

Case 2

A 46-year-old female with a history of morbid obesity and diabetes who underwent a laparoscopic sleeve gastrectomy procedure complicated by a delayed microperforation from the proximal gastric staple line and a subdiaphragmatic abscess. She underwent laparoscopic drainage and endoscopic stent placement (fully-covered 150x23 mm WallFlex, Boston Scientific). The stent resulted in the elimination of gastric fistula, but three times in the ensuing six weeks, the stent migrated antegrade to the pylorus, producing pain and vomiting (Figure 2). In each case, the stent was endoscopically repositioned. The final procedure included endoluminal fixation of the stent by suturing it to the esophageal wall with two sutures to the proximal stent flange (Apollo OverStitch). After the endoluminal suture fixation, the stent remained in position and was removed 10 weeks later, resulting in successful resolution of the gastric leak.

Case 3

A 38-year-old female with morbid obesity and hypertension who underwent a laparoscopic sleeve gastrectomy procedure and presented three weeks later with fever and left sided chest pain. She was found to have a gastric microperforation with fluid above and below the diaphragm. At surgery, a laparoscopic washout and drain placement were performed with tube thoracostomy and endoscopic placement of a fully covered stent (150 x 20 mm Evolution, Cook Medical). She quickly improved clinically and went home with a drain and stent in position, on TPN only to return three weeks postop with epigastric pain and retching due to migration of the stent. The perigastric drain turned cloudy and purulent and increased in volume, indicating a return of the gastric fistula. Repeat endoscopy was performed to reposition the stent, and it was secured with endoluminal sutures to the esophageal wall (Figure 3). No further migration occurred. The stent was removed after 12 weeks and the leak resolved.

Case 4

A 44-year-old female developed a leak from the proximal stomach 4 weeks after undergoing sleeve gastrectomy. She was treated with laparoscopic washout and drain placement followed by endoscopic stent placement. The stent migrated antegrade within two weeks and was replaced by two stents positioned in tandem (150x23 mm and 120x23 mm WallFlex, Boston Scientific). Three weeks later ,the stents migrated and were replaced by a single stent, secured with two endoluminal sutures to the esophageal wall (Figure 4). The stent remained in place until it was removed 12 weeks later.

Case 5 is that of a 49-year-old female with a history of gastric banding and recurrent obesity. She underwent concomitant removal of the band and conversion to sleeve gastrectomy. Two weeks postoperatively, she presented with sepsis and evidence of a leak from the proximal staple line. Open surgical washout and drain placement was performed, and attempted primary closure of the fistula was unsuccessful. Endoscopic stent placement was performed (fully-covered 150x23 mm Wallflex, Boston Scientific), resulting in resolution of the gastric fistula, and the patient improved clinically. After three weeks the stent migrated distally and required repeat endoscopic positioning, this time secured with endoluminal suturing using two absorbable sutures through the proximal stent wall and the esophageal wall (Apollo OverStitch). No further stent migration occurred, and the patient eventually fully recovered.

DISCUSSION

Endoluminal stenting has proven to be an effective mode of treatment for esophageal and gastric leaks, perforations, structures, and fistulae [1–4]. With the growth of sleeve gastrectomy, there has been a parallel growth in delayed microperforation, the most serious and frequent complication of sleeve gastrectomy. Endoluminal stent migration, while normally fairly innocuous does cause vomiting and epigastric pain for the patient. It also requires an additional procedure for repositioning of the

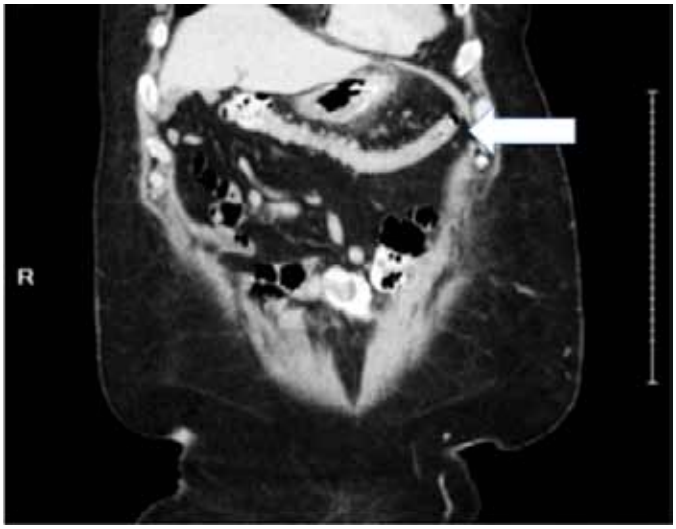


Figure 1: Computed tomography scan demonstrating migration of stent to the jejunum.

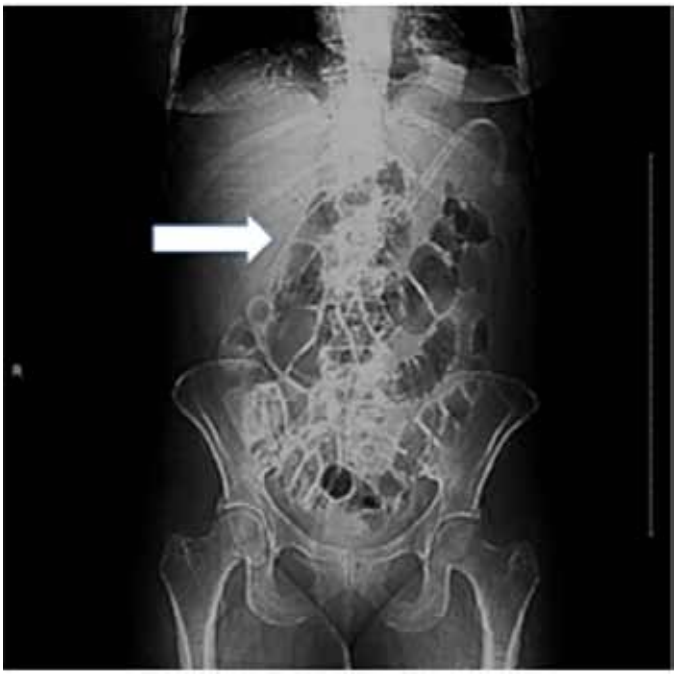


Figure 2: Computed tomography demonstrating migration of stent to the distal stomach and pylorus.

stent. However, as this first case presented here points out, it is a potentially very serious and life-threatening complication.

In this paper, we report five cases in which antegrade migration of endoluminal esophagogastric stents occurred at least once following stent placement for sleeve gastrectomy leaks. One of these cases resulted in a severe complication of jejunal perforation. In each case, repeat endoscopy was performed to reposition the stent and secure the stent in position with an endoluminal suturing technique. In each of those cases of stent fixation utilizing endoluminal sutures to the esophageal wall, no stent migration occurred. No complications of the suture fixation technique occurred.

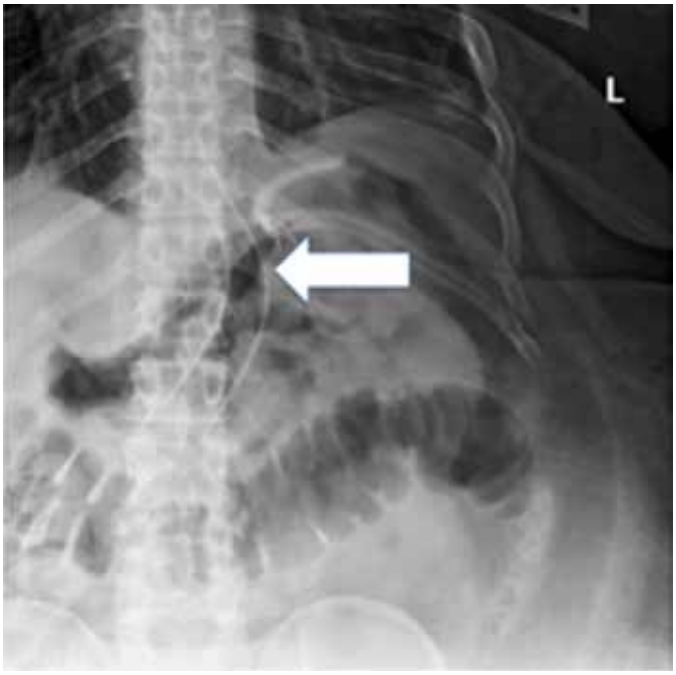


Figure 3: Plain abdominal film demonstrating successfully repositioned stent secured with endoluminal suture. The perigastric drain may be seen in the left upper quadrant.

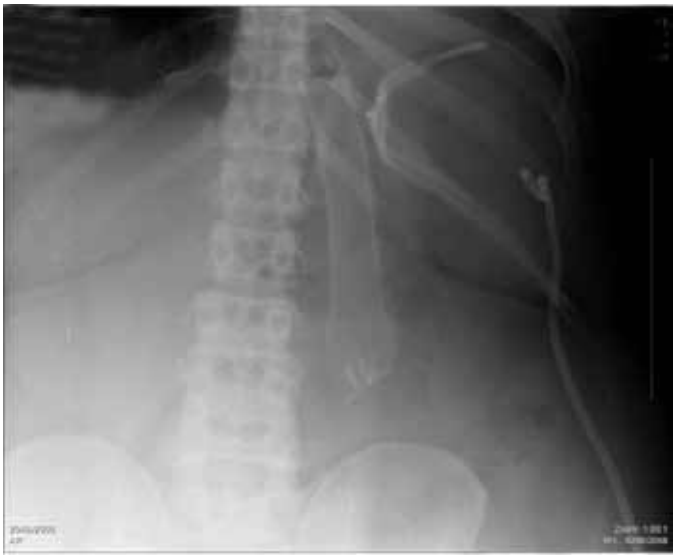


Figure 4: Plain abdominal film showing stent successfully repositioned and secured with endoluminal suture. Sutures are placed at the most proximal/superior portion of the stent.

Migration of endoluminal stents has been a reported and vexing complication occurring in a significant number of cases [2, 5, 6]. Reports that emphasize use of clips, tandem stenting, or other fixation techniques have suggested that the techniques may provide greater security, although that has not been well demonstrated. In case one reported above, the patient experienced a serious complication of jejunal perforation following stent

migration. In an effort to prevent such complications, our practice turned to a procedure of suture fixation utilizing an endoluminal suture device technique (Apollo Overstitch).

Other authors have reported use of endoscopic clip placement, utilization of tandem stents, and the use of partially covered stents in order to reduce stent migration [3, 5]. In our experience, the endoluminal suturing technique has proven successful, potentially more robust than clipping, and easy to perform.

CONCLUSION

Endoluminal suturing is technically straightforward and successful in preventing endoluminal stent migration. After some practice, it adds only a short amount of additional time to the procedure, normally less than 15 minutes. At this point, given our experience resolving a devastating complication of stent migration, and after multiple repeat endoscopies to reposition migrated stents, endoluminal suture fixation has become standard approach to any endoluminal stent placement. Potential risks of routine suture fixation of endoluminal stents include bleeding or perforation from the esophageal wall suture placement, something we have not seen in clinical practice, and we are unaware of its having been reported to date. Concerns of esophageal wall suture placement must be balanced against the frequency and the risks of stent migration.

****

Author Contributions

Kent C. Sasse – 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
David L. Warner – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Jared Brandt – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Ellen Ackerman – 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. Eubanks S, Edwards CA, Fearing NM, et al. Use of endoscopic stents to treat anastomotic complications after bariatric surgery. J Am Coll Surg 2008 May;206(5):935–8; discussion 938–9.
2. Simon F, Siciliano I, Gillet A, Castel B, Coffin B, Msika S. Gastric leak after laparoscopic sleeve gastrectomy: early covered self-expandable stent reduces healing time. Obes Surg 2013 May;23(5):687–92.
3. Gonzalez JM, Garces Duran R, et al. Double-type metallic stents efficacy for the management of post-operative fistulas, leakages, and perforations of the upper gastrointestinal tract. Surg Endosc 2015 Jul;29(7):2013–8.
4. van Boeckel PG, Dua KS, Weusten BL, et al. Fully covered self-expandable metal stents (SEMS), partially covered SEMS and self-expandable plastic stents for the treatment of benign esophageal ruptures and anastomotic leaks. BMC Gastroenterol 2012 Feb 29;12:19.
5. Galloro G, Magno L, Musella M, Manta R, Zullo A, Forestieri P. A novel dedicated endoscopic stent for staple-line leaks after laparoscopic sleeve gastrectomy: a case series. Surg Obes Relat Dis 2014 Jul-Aug;10(4):607–11.
6. Martins BC, Retes FA, Medrado BF, et al. Endoscopic management and prevention of migrated esophageal stents. World J Gastrointest Endosc 2014 Feb 16;6(2):49–54.
7. Fujii LL, Bonin EA, Baron TH, Gostout CJ, Wong Kee Song LM. Utility of an endoscopic suturing system for prevention of covered luminal stent migration in the upper GI tract. Gastrointest Endosc 2013 Nov;78(5):787–93.
8. Irani S, Baron TH, Gluck M, Gan I, Ross AS, Kozarek RA. Preventing migration of fully covered esophageal stents with an over-the-scope clip device (with videos). Gastrointest Endosc 2014 May;79(5):844–51.
9. Kantsevoy SV, Bitner M. Esophageal stent fixation with endoscopic suturing device (with video). Gastrointest Endosc 2012 Dec;76(6):1251–5.
10. Sharaiha RZ, Kumta NA, Doukides TP, et al. Esophageal Stenting With Sutures: Time to Redefine Our Standards? J Clin Gastroenterol 2015 Jul;49(6):e57–60.

CASE SERIES

PEER REVIEWED | OPEN ACCESS

Coronary pulmonary fistula: A case series

Maarten Van Caenegem, Hans Vandekerckhove

ABSTRACT

Introduction: Coronary artery fistulas (CAF) are congenital or acquired coronary artery anomalies, circumventing the myocardial capillary network by directly draining the blood into a great vessel, cardiac chamber, or other vascular structure. Clinical manifestations vary considerably with presentation of dyspnea, congestive heart failure, angina, endocarditis, arrhythmias, or myocardial infarction. Case Report: We report two cases, one between the right coronary artery (RCA) and the pulmonary artery, whereas the other between the circumflex artery (RCX) and the pulmonary artery. We describe our diagnostic methodology and analyze literature on the epidemiology, the diagnostic workout and the treatment possibilities. Conclusion: Despite the role of non-invasive imaging for diagnosis and identification of the location of CAF including the origin and insertion of the recipient vessel, cardiac catheterization and coronary angiography remain the preliminary diagnostic tools for the precise allocation of coronary anatomy, for assessment of its hemodynamic importance, and to show other structural abnormalities.

Maarten Van Caenegem[1], Hans Vandekerckhove[2]

Affiliations: [1]MD, Department of Cardiology, Ghent University Hospital, Ghent, Belgium; [2]MD, Department of Cardiology, Sint Lucas General Hospital, Ghent, Belgium.

Corresponding Author: Maarten Van Caenegem, MD, Provincieweg 351 B-9550 Herzele, Belgium; Email: maarten.vancaenegem@ugent.be

Received: 21 December 2015
Accepted: 19 February 2016
Published: 01 May 2016

Keywords: Coronary artery fistula, Coronarography, Chest pain, Emotional stress, Pulmonary fistula, Sleep apnea syndrome

How to cite this article

Van Caenegem M, Vandekerckhove H. Coronary pulmonary fistula: A case series. Int J Case Rep Imag 2016;7(5):292–295.

Article ID: Z01201605CR10069MC

****

doi:10.5348/ijcri-201608-CS-10069

INTRODUCTION

Coronary artery fistulae (CAF) are anomalous acquired or congenital terminations of the coronary arteries into other vascular structures, such as a cardiac chamber, vena cava, the pulmonary artery, or pulmonary veins. Most of the coronary anomalies are incidental findings during angiographic evaluation for coronary vascular disorders. Coronary artery fistulae are present in 0.002% of the overall population and are documented in almost 0.25% of the patients undergoing coronary angiography [1]. Majority of these fistulas originate from the right coronary artery or the left anterior descending artery. Patients are frequently asymptomatic, but angina due to coronary steal phenomenon or myocardial infarction and dyspnea due to heart failure and endocarditis have been reported in some cases [2, 3]. The management is complex, and recommendations are founded on anecdotal cases of very small retrospective series.

CASE SERIES

Case 1

A 64-year-old male presented to the hospital with dyspnea and angina during emotional stress. He was known with pulmonary sarcoïdosis in remission and obstructive sleep apnea syndrome. He did not take any medication. His general examination was unremarkable. An additional exercise test, however, was clinical suspicious with the reproduction of atypical thoracic complaints in the absence of electrocardiographic changes. Echocardiography showed a preserved left ventricular systolic function with absence of valvular disease. Coronary angiography confirmed slight coronary atheromatosis, however, there was a fistula between the right coronary artery and the pulmonary artery with the presence of minor pulmonary hypertension (Figure 1). A supplementary CT scan of the lungs identified agenesis of the right pulmonary artery with a dysplastic right lung. There was also collateral circulation form the iliac veins and vena cava inferior, an occlusion of the vena cava superior and an arteriovenous malformation of the right middle lobe originating from the right coronary artery (Figure 2). To exclude a vascular steal phenomenon form the right coronary artery, we organized a Thallium stress test, which ruled out stress induced ischemia. A pulmonary function test illustrated an unchanged restrictive pattern due to the dysplastic right lung and his obesity. Concerning the etiology of the absence of the right pulmonary artery and the presence of a dysplastic right lung, we could not differentiate between congenital versus acquired phenomenon (possibly due to external compression of calcified lymph nodes by sarcoidosis at young age). Considering the absence of stress induced ischemia, hemoptysis or pulmonary infections, we suggested a conservative approach with annual evaluation of progressive pulmonary hypertension. A calcium channel blocker was initiated experimentally with full symptom relief.

Case 2

A 46-year-old male presented to the emergency room complaining of dyspnea, slight hemoptysis and atypical chest pain. Physical examination was unremarkable. Electrocardiography showed a normal sinus rhythm, normal QRS morphology and normal repolarization. Transthoracic echocardiography illustrated a normal left and right ventricular morphology with preserved left ventricular systolic function. Pulmonary artery pressure was within normal range. Based on his moderate cardiac risk profile and an inconclusive exercise test, a cardiac catheterization was planned. It revealed non-obstructive coronary artery disease and a large coronary pulmonary fistula communicating from the right circumflex (RCX) coronary artery to a network of collateral circulation to the right lung. An interruption of the right pulmonary artery and vascularization of the right lung by aberrant intercostal arteries and the right mammary artery (Figure 3) are illustrated as well by a supplementary angiography of the pulmonary arteries with the presence of a unique left pulmonary artery. A supplementary cardiac MRI could not detect any other site of an intracardiac of extracardiac shunt. A vascular steal phenomenon was ruled out by a normal Thallium stress test. Considering the absence of pulmonary hypertension, exercise induced ischemia or arrhythmia, a conservative approach was taken.

DISCUSSION

Coronary artery fistulae are uncommon congenital abnormalities of the coronary arteries or more seldom

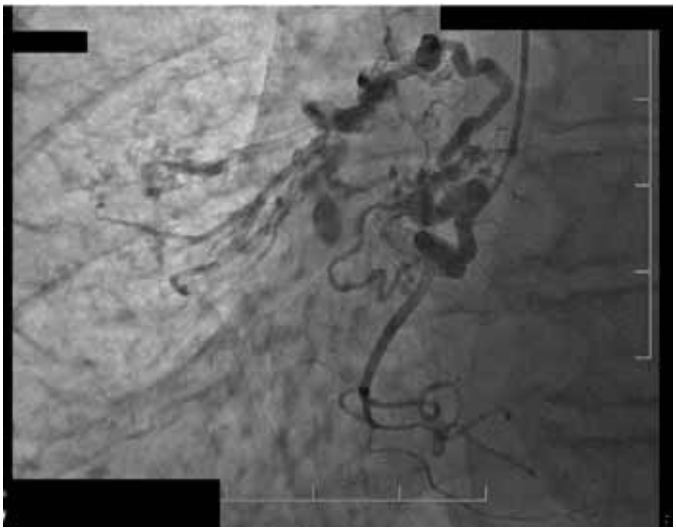


Figure 1: Cardiac catheterization: A coronary pulmonary fistula originating from the proximal right coronary artery (Case 1).

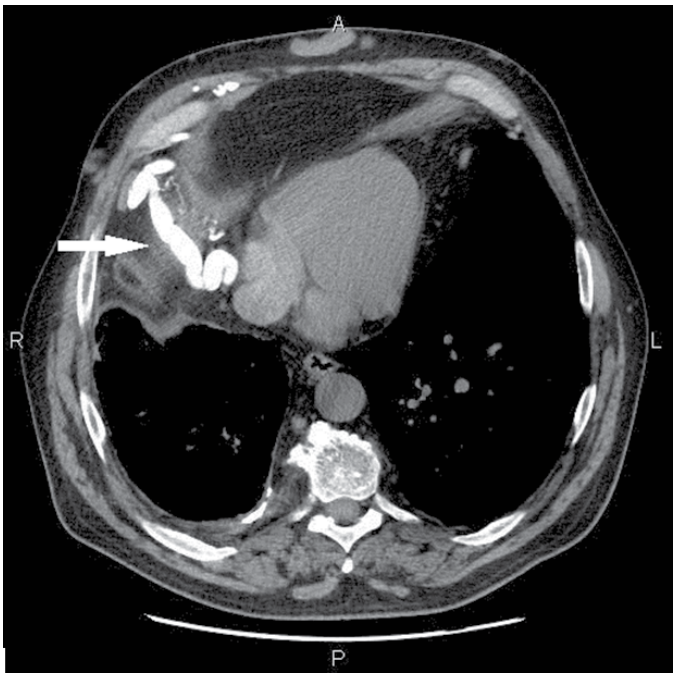


Figure 2: An arteriovenous malformation of the right middle lobe originating from the right coronary artery (Case 1).

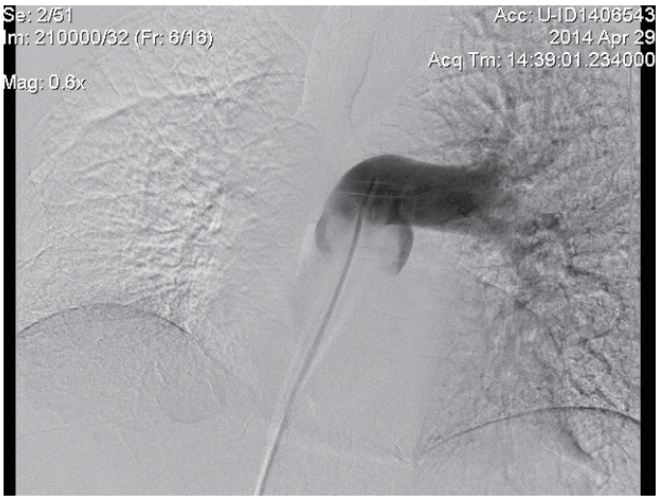


Figure 3: Agenesis of the right pulmonary artery by pulmonary angiography (Case 2).

acquired conditions and can occur iatrogenic after cardiac surgery like coronary artery bypass surgery or cardiac transplantation, or as a complication of coronary angioplasty and myocardial biopsy [4].

These fistulas originate more frequently from the right coronary artery and commonly shunt into one of the right heart chambers. Clinical presentation is dependent on the magnitude of the fistulous connection and could seldom result in a significant left-to-right shunt with congestive heart failure and cardiomegaly in infancy if a large fistula is present [5].

Nevertheless the growing potential of several non-invasive techniques like transthoracic and transesophageal echocardiography or cardiac imaging with magnetic resonance or computed tomography for identification and follow-up, coronary angiography still is the gold standard at present because of its accuracy in defining the artery of origin, as well as the recipient vascular structure.

Coronary artery fistulae cause myocardial ischemia in only in a small number of patients [6]. In planning therapy, evaluation of the hemodynamic importance of the fistulae is crucial. Next to a stress electrocardiography, a stress/rest 99mTc sestamibi single photon emission tomography is trustworthy for assessing the functionality of the anomalies detected by coronary angiography.

The natural history of CAF is unpredictable within one reported case an uncommon spontaneous closure due to spontaneous thrombosis [7]. For that reason, there is still some controversy about the management and follow-up, which is generally based on small retrospective series or anecdotal cases.

Antiplatelet therapy with at least one antiplatelet agent is recommended, especially in patients with distal coronary artery fistulas and abnormally dilated coronary arteries [8]. Based on limited experience and anecdotal cases bacterial endocarditis is a known complication and for that reason prophylactic precautions against subacute bacterial endocarditis are suggested.

The presence of heart failure and myocardial ischemia are predominant clinical symptoms to consider closure of the CAF. In addition to prevent occurrence of symptoms or complications, closure of CAF must also be considered in asymptomatic patients with high-flow shunting, especially in pediatric population [9]. Nevertheless, treatment of non-significant shunting in asymptomatic adult patients is still doubtful. There is no sufficient medical treatment for CAF. The choice between trans-catheter closure of the fistula and surgical intervention is still controversial. However, trans-catheter closure may be indicated if the anatomy is favorable (e.g. non-tortuous vessel) and the distal portion of the fistula is accessible with the closure device and should be narrow to avoid embolization to the drainage site. Catheter closure can be performed with a variety of techniques, including detachable balloons, stainless steel coils, regular and covered stents, and various chemicals. The main goal is to restore the myocardial perfusion by reduction in left to right shunt after occlusion of the treated vessel to the level of first branch. The basic surgical technique is ligation of the fistula and may be performed with or without cardiopulmonary bypass, when there is a simple and easily accessible fistula [10]. Results from the transcatheter and surgical literature show percutaneous closure were associated with lower procedural risk and therefore becomes the preferred method of treatment [11].

There was also reported that recanalization of the treated coronary fistulae can occur, and they suggested follow-up angiography or other imaging modality, like stress electrocardiography or cardiac MRI scan, to be performed annualy in the beginning of follow-up in these patients and decreasing the frequency in case of asymptomatic stable condition [12].

CONCLUSION

Coronary artery fistulae (CAF) are exceptional, isolated abnormalities that are usually asymptomatic. However, certain forms are associated with myocardial ischemia, congestive heart failure, and sudden cardiac death. Identification of signs and symptoms should lead to supplementary testing, especially thorough initial evaluation of coronary artery anatomy using echocardiography to detect shunting, cardiac MRI scan or coronary computed tomography. However, coronary angiography remains the gold standard for diagnosis. Clinically significant CAF has to be considered for elective closure based on the current safety and efficacy of both transcatheter and surgical closure of CAF. The type of intervention will depend on the anatomy and origin of the fistula, the magnitude of the vascular malformation and possible associated defects and certainly the experience of the surgeons and interventional cardiologists.

****

Author Contributions

Maarten Van Caenegem – 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

Hans Vandekerckhove – 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. Raju MG, Goyal SK, Punnam SR, Shah DO, Smith GF, Abela GS. Coronary artery fistula: a case series with review of the literature. J Cardiol 2009 Jun;53(3):467–72.
2. Gowda RM, Vasavada BC, Khan IA. Coronary artery fistulas: clinical and therapeutic considerations. Int J Cardiol 2006 Feb 8;107(1):7–10.
3. Vijayvergiya R, Bhadauria PS, Jeevan H, Mittal BR, Grover A. Myocardial ischemia secondary to dual coronary artery fistulas draining into main pulmonary artery. Int J Cardiol 2010 Apr 15;140(2):e30–3.
4. Ibrahim MF, Sayed S, Elasfar A, Sallam A, Fadl M, Al Baradai A. Coronary fistula between the left anterior descending coronary artery and the pulmonary artery: Two case reports. J Saudi Heart Assoc 2012 Oct;24(4):253–6.
5. Koneru J, Samuel A, Joshi M, Hamden A, Shamoon FE, Bikkina M. Coronary anomaly and coronary artery fistula as cause of angina pectoris with literature review. Case Rep Vasc Med 2011;2011:486187.
6. Rubini G, Ettorre GC, Sebastiani M, Bovenzi F. [Evaluation of hemodynamic significance of arteriovenous coronary fistulas: diagnostic integration of coronary angiography and stress/rest myocardial scintigraphy]. [Article in Italian]. Radiol Med 2000 Dec;100(6):453–8.
7. Sapin P, Frantz E, Jain A, Nichols TC, Dehmer GJ. Coronary artery fistula: an abnormality affecting all age groups. Medicine (Baltimore) 1990 Mar;69(2):101–13.
8. Umaña E, Massey CV, Painter JA. Myocardial ischemia secondary to a large coronary-pulmonary fistula--a case report. Angiology 2002 May-Jun;53(3):353–7.
9. Balanescu S, Sangiorgi G, Castelvecchio S, Medda M, Inglese L. Coronary artery fistulas: clinical consequences and methods of closure. A literature review. Ital Heart J 2001 Sep;2(9):669–76.
10. Cheung DL, Au WK, Cheung HH, Chiu CS, Lee WT. Coronary artery fistulas: long-term results of surgical correction. Ann Thorac Surg 2001 Jan;71(1):190–5.
11. Tirilomis T, Aleksic I, Busch T, Zenker D, Ruschewski W, Dalichau H. Congenital coronary artery fistulas in adults: surgical treatment and outcome. Int J Cardiol 2005 Jan;98(1):57–9.
12. Jama A, Barsoum M, Bjarnason H, Holmes DR Jr, Rihal CS. Percutaneous closure of congenital coronary artery fistulae: results and angiographic follow-up. JACC Cardiovasc Interv 2011 Jul;4(7):814–21.

CASE REPORT PEER REVIEWED | OPEN ACCESS

An unusual presentation of papillary thyroid carcinoma in the lateral aspect of the neck

Ghassan Almaimani, Frank Forst, Thomas Zoedler, Bayan Almaimani

ABSTRACT

Introduction: Papillary thyroid carcinoma (PTC) is the most common thyroid cancer. Papillary thyroid carcinoma presenting as a lateral neck mass in the presence of a normal thyroid is extremely rare. Ectopic thyroid tissue can arise in the lateral aspect of the neck, and, therefore, PTC may arise at this site in ectopic tissue. Alternatively, cervical cystic lymph node metastasis may be a first presentation of occult PTC. Case Report: A case of a 28-year-old male was referred for investigation of a painless left-sided neck swelling. Excisional biopsy revealed PTC, and completion surgery revealed papillary microcarcinoma and a further lymph node metastasis in the definitive resection specimen. Conclusion: This case highlights that PTC always needs to be considered in the differential diagnosis of lateral neck swellings even in presence of a normal thyroid. Establishing the exact etiology of lateral cystic PTC can be challenging.

Keywords: Cervical lymph node, Lateral neck, Metastasis, Papillary thyroid cancer

Ghassan Almaimani[1], Frank Forst[2], Thomas Zoedler[3], Bayan Almaimani[4]

Affiliations: [1]MBBS, Resident, Department of Surgery, Main-Kinzig-Clinics, Gelnhausen, Hessen, Germany; [2]Consultant Surgeon, Main-Kinzig-Clinics, Gelnhausen, Hessen, Germany; [3]Doctorate, Consultant Surgeon, Head of Department, Main-Kinzig-Clinics, Gelnhausen, Hessen, Germany; [4]MBBS, Resident, Maternity and Children Hospital, Makkah, Saudi Arabia.

Corresponding Author: Ghassan Almaimani, Weinbergstr. 6a, Gelnhausen, Hessen, Germany, 63571; Email: dr.gaam@hotmail.com

Received: 04 January 2016
Accepted: 17 February 2016
Published: 01 May 2016

How to cite this article

Almaimani G, Forst F, Zoedler T, Almaimani B. An unusual presentation of papillary thyroid carcinoma in the lateral aspect of the neck. Int J Case Rep Imag 2016;7(5):296–299.

Article ID: Z01201605CR10638GA

****

doi:10.5348/ijcri-201650-CR-10638

INTRODUCTION

Almost 80% of all thyroid cancers are papillary thyroid carcinomas (PTCs), which most commonly present as solitary thyroid nodules [1]. However, PTCs can extremely uncommonly present as lateral neck masses in the absence of thyroid swelling. Here we report a case of PTC presenting as a left-sided neck swelling. Although evaluation of neck swellings usually include ultrasonography, fine-needle aspiration cytology (FNAC), and computed tomography (CT) scan or magnetic resonance imaging (MRI) scan and radionuclide scanning, excisional biopsy of a neck cyst is essential for early diagnosis and management if these results are inconclusive [2]. Our case was diagnosed by histopathological examination after surgical excision of the neck lesion.

CASE REPORT

A 28-year-old male presented with a painless left-sided neck swelling that had gradually increased in size

over the preceding few months. The swelling was sudden in onset and there was no history of preceding infection or trauma. There was no dysphagia, dyspnea, or hoarseness, and the patient had no other medical history of note and a negative family history of thyroid cancer. Neck examination revealed a normal thyroid gland and no cervical lymphadenopathy. However, a 5x3 cm non-tender mass was present in the left posterior triangle. Thyroid function tests were within normal limits. There were no suspicious features such as microcalcifications or solid components on ultrasound examination. Neck MRI scan with contrast revealed a fluid-filled cystic mass measuring 7.6x4.9 cm between the left sternocleidomastoid muscle and anterior scalene muscle, separate from the left lobe of the thyroid (Figure 1). The imaging features were highly suggestive of cystic hygroma. The decision was made to perform excisional biopsy without prior fine-needle aspiration biopsy (FNAB) due to its location close to vascular structures and to establish a definitive diagnosis.

Histopathological examination of the specimen showed features of PTC (Figure 2A). The patient was informed about the need for further surgery and subsequently underwent total thyroidectomy with central compartment neck dissection and a left modified radical neck dissection (MRND). Histopathological analysis of the resection specimen showed a focus of papillary microcarcinoma in the left lobe of thyroid measuring 0.7 cm (Figure 2B–C) and further single lymph node metastasis in a left neck node (Figure 2D).

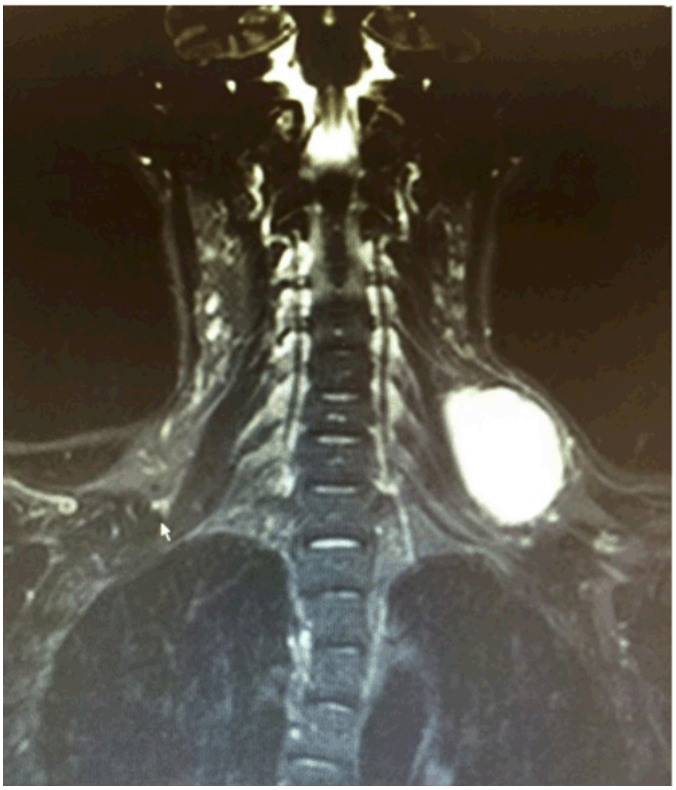


Figure 1: Coronal T2-weighted magnetic resonance imaging scan of the neck showing a large, hyper-intense mass on the left side of the neck.

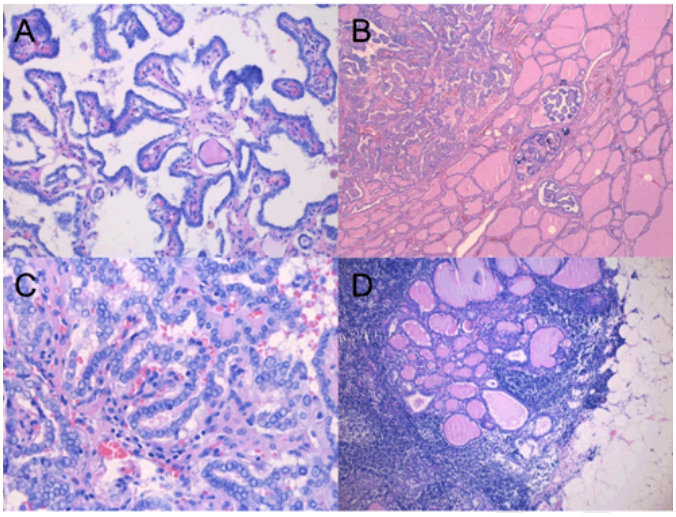


Figure 2: Histopathology of the lesion and resection specimen (A) Photomicrograph showing tumor tissue arranged in papillae and showing the characteristic pathological features of PTC of nuclear clearing and overlapping and psammomatous calcifications (magnification: x20). (B, C) Histological examination of the surgical specimen revealed a 7 mm papillary thyroid microcarcinoma in the left lobe. (B) magnification: x10 and (C) magnification: x40. (D) Photomicrograph of metastatic PTC in a lymph node (magnification: x20). The remaining lymph nodes were normal.

The patient was counseled with respect to the need for radioactive iodine therapy after surgery. He subsequently underwent radioactive iodine therapy postoperatively, and a radioactive iodine scan six weeks later revealed no residual thyroid. The patient remains on thyroxine supplementation and regular follow-up.

DISCUSSION

Papillary thyroid cancers (PTCs) are most common in adult women (mean age of presentation 30 years; F:M 3:1). Cervical metastases are present in 50-75% of patients, depending on the cancer size. Distant metastases are uncommon, but, when metastases occur, lung and bone are the most common sites [1].

Cystic neck masses appearing in the anterior or posterior neck triangles are usually benign but occasionally harbor cancers. Some studies have reported thyroid malignancy in 11% of patients with lateral cervical cysts [2]. However, a solitary cervical cystic mass is an uncommon presentation of PTC with less than 25 reported cases worldwide in the literature.

The thyroid gland is embryologically derived from two anlages: a large median endodermal anlage and two lateral anlages. The median anlage produces most of the thyroid parenchyma, while the lateral anlage is derived from the fourth pharyngeal pouch [3]. Not uncommonly, a failure in descent of the median anlage results in a lingual

thyroid gland. In much rarer cases, failure of fusion of the lateral and median anlages can result in a lateral ectopic thyroid gland [4]. When cancers do arise in the lateral cervical location, some authors consider this to be more likely due to secondary metastatic spread of papillary microcarcinoma to a lymph node that undergoes central liquefaction and cyst formation, while other studies regard these lesions as malignant transformations of ectopic thyroid tissue [5, 6].

With respect to this diagnostic difficulty, Cabibi et al. [7] concluded that the presence of morphologically and immunohistochemically normal-looking follicles in lateral cervical masses without coexisting intrathyroid tumor strongly suggest malignant transformation of ectopic thyroid tissue. Conversely, even if the immunohistochemical pattern in thyroid follicles in lateral cervical masses indicates differentiation resembling normal thyroid parenchyma but there is coexisting intrathyroid tumor, these masses should be considered metastatic in nature.

Verge et al. [8] and Al-Ashaa et al. [9] together reported the largest series of twelve cases of cervical cystic lymph node metastases as the first manifestation of occult PTC. In their reports, in all cases the thyroid tumors were not palpable on physical examination and no abnormalities of the thyroid gland were shown by other diagnostic procedures.

In our case, and based on the histological findings of the presence of another definitive lymph node metastasis and the presence of a focus of papillary microcarcinoma in the left lobe, we consider this lesion to be metastatic in nature. The metastasis is likely to have undergone cystic degeneration to produce the palpable lesion. An in vivo diagnostic method capable of differentiating between ectopic thyroid tissue and metastatic thyroid cancer would be useful to enable accurate preoperative assessment and to guide and refine the decision regarding the extent of the operation required.

We did not consider FNA for investigation of the mass even though the overall sensitivity and specificity of FNA in the diagnosis of neck lesions is approximately 85–90%. This was because: (i) the diagnostic accuracy of cystic mass aspirates may be especially poor due to the likelihood of aspirating acellular material [10]; and (ii) the lesion was in proximity to vascular structures and the risk of post-aspiration hemorrhage was deemed to outweigh the risk and diagnostic benefit of performing excisional biopsy in this case.

CONCLUSION

Literature reports only a few cases of lateral neck swelling as the only manifestation of papillary thyroid carcinoma (PTC). It must always be considered in the differential diagnosis of lateral neck swellings presenting in adult patients. Establishing the exact etiology of these lesions can be difficult and requires systematic histopathological assessment in the absence of appropriate in vivo diagnostic modalities.

****

Acknowledgements
We would like to thank Dr. Sebastian Blasius for preparing the histology illustrations.

Author Contributions
Ghassan Almaimani – 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
Frank Forst – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Final approval of the version to be published
Thomas Zoedler – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published
Bayan Almaimani – Substantial contributions to conception and design, 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. Hundahl SA, Fleming ID, Fremgen AM, Menck HR. A National Cancer Data Base report on 53,856 cases of thyroid carcinoma treated in the U.S., 1985-1995. Cancer 1998 Dec 15;83(12):2638–48.
2. Seven H, Gurkan A, Cinar U, Vural C, Turgut S. Incidence of occult thyroid carcinoma metastases in lateral cervical cysts. Am J Otolaryngol 2004 Jan-Feb;25(1):11–7.
3. Mansberger AR Jr, Wei JP. Surgical embryology and anatomy of the thyroid and parathyroid glands. Surg Clin North Am 1993 Aug;73(4):727–46.
4. Paresi RJ Jr, Shah D. Hashimoto's thyroiditis presenting as an enlarging submandibular mass in a patient with a lingual thyroid. Otolaryngol Head Neck Surg 2005 May;132(5):806–8.

5. LiVolis VA. Thyroid papillary carcinoma in lateral neck cyst: missed primary tumour or ectopic thyroid carcinoma within a branchial cyst? J Laryngol Otol 2001 Jul;115(7):614–5.
6. Loughran CF. Case report: Cystic lymph node metastasis from occult thyroid carcinoma: a sonographic mimic of a branchial cleft cyst. Clin Radiol 1991 Mar;43(3):213–4.
7. Cabibi D, Cacciatore M, Guarnotta C, Aragona F. Immunohistochemistry differentiates papillary thyroid carcinoma arising in ectopic thyroid tissue from secondary lymph node metastases. Thyroid 2007 Jul;17(7):603–7.
8. Verge J, Guixá J, Alejo M, et al. Cervical cystic lymph node metastasis as first manifestation of occult papillary thyroid carcinoma: report of seven cases. Head Neck 1999 Jul;21(4):370–4.
9. Al-Ashaa Y, Hefny AF, Joshi S, Abu-Zidan FM. Papillary thyroid carcinoma presenting as a lateral neck cyst. Afr Health Sci 2011 Jun;11(2):296–300.
10. Gourin CG, Johnson JT. Incidence of unsuspected metastases in lateral cervical cysts. Laryngoscope 2000 Oct;110(10 Pt 1):1637–41.

CASE REPORT PEER REVIEWED | OPEN ACCESS

A case of Stevens–Johnson syndrome in a patient on ipilimumab

Mohini Pathria, Jyoti Mundi, Joshua Trufant

ABSTRACT

Introduction: Stevens–Johnson syndrome is a rare complication of ipilimumab therapy. A brief review of literature on the dermatologic adverse effects of ipilimumab is presented. This paper aims to heighten awareness of the significant risk and potential severity of cutaneous adverse effects associated with the use of ipilimumab. Case Report: A case of 71-year-old woman being treated with ipilimumab for stage IV choroidal melanoma who presented with Stevens–Johnson syndrome 2.5 weeks after her last ipilimumab infusion. Conclusion: Delayed diagnosis of Stevens–Johnsons syndrome in patients receiving ipilimumab therapy can lead to devastating outcomes.

Keywords: Ipilimumab, Melanoma, Stevens–Johnson syndrome

How to cite this article

Pathria M, Mundi J, Trufant J. A case of Stevens–Johnson syndrome in a patient on ipilimumab. Int J Case Rep Imag 2016;7(5):300–302.

Mohini Pathria[1], Jyoti Mundi[2], Joshua Trufant[2]

Affiliations: [1]M.D, University of Florida Department of Internal Medicine, Gainesville, FL -Position: Resident Physician; [2]M.D, Ronald O. Perelman Department of Dermatology, NYU Langone Medical Center, New York NY, Dermatologist.

Corresponding Author: Mohini Pathria, 1600 SW Archer Road, Shands Hospital, Room 4102, Gainesville, Florida USA 32610; Email: mohini.pathria@medicine.ufl.edu

Received: 05 January 2016
Accepted: 16 February 2016
Published: 01 May 2016

Article ID: Z01201605CR10639MP

****

doi:10.5348/ijcri-201651-CR-10639

INTRODUCTION

Ipilimumab is a monoclonal human antibody directed against cytotoxic T lymphocyte antigen-4 (anti-CTLA-4). The medication helps increase T cell activation [1]. The therapy has been recently approved for management of metastatic melanoma. Common adverse effects involve the gastrointestinal tract and skin [2]. Skin reactions can range from mild to severe, including Stevens–Johnson syndrome (SJS) [2]. Stevens–Johnson syndrome is characterized by epidermal death and separation involving less than 10% of the skin surface. If more than 30% of the skin is involved, the syndrome is considered toxic epidermal necrolysis (TEN), with SJS/TEN overlap for involvement of 10–30% of the skin surface [3]. Stevens-Johnson syndrome (SJS) and/or toxic epidermal necrolysis (TEN) with ipilimumab therapy is a rare complication and the exact frequency is unknown. Delayed diagnosis can lead to devastating complications.

CASE REPORT

A 71-year-old female was being treated for stage IV choroidal melanoma with ipilimumab. Two and a half weeks after her third infusion, she presented with a progressively painful rash on her face, trunk, and extremities, ocular discharge, and dysuria. Physical examination revealed conjunctival injection, painful crusted erosions on her vermilion, mucosal lip and labia majora, and dusky targetoid patches on her forehead and extremities. The patient's trunk was diffusely

erythematous and tender. Nikolsky's sign was present and spontaneous sloughing was noted over approximately 25% of the patient's body surface area (Figure 1). The affected epidermis was submitted for frozen sections and a 4-mm punch biopsy was performed (Figure 2A–B). The biopsy demonstrated full-thickness epidermal necrosis with a sparse perivascular lymphocytic infiltrate. The clinical and histologic findings were consistent with Stevens–Johnson syndrome. The patient was transferred to a burn unit. The patient was treated with supportive measures including artificial tears and Lacrilube to the eyes. Daily vitamin A and vitamin D ointment and xeroform was applied to denuded skin. She received intravenous fluid resuscitation and tube feedings to enhance protein intake. She has fully recovered from this episode and has resumed her baseline quality of life.

DISCUSSION

Ipilimumab is a monoclonal immunoglobulin G1 antibody directed against cytotoxic T lymphocyte antigen-4 (anti-CTLA-4). The medication received FDA approval in 2011 as monotherapy for metastatic melanoma for four cycles at a dose of 3 mg/kg administered intravenously every three weeks for 90 minutes [1]. CTLA-4 diminishes T cell activation by competing with CD28 on T cells for co-stimulatory molecules on antigen presenting cells [1]. In metastatic melanoma, T cells are functionally impaired, while inhibitory receptors, such as CTLA-4, are upregulated [4]. In vivo studies have demonstrated that CTLA-4 blockade can promote antitumor immunity [5].

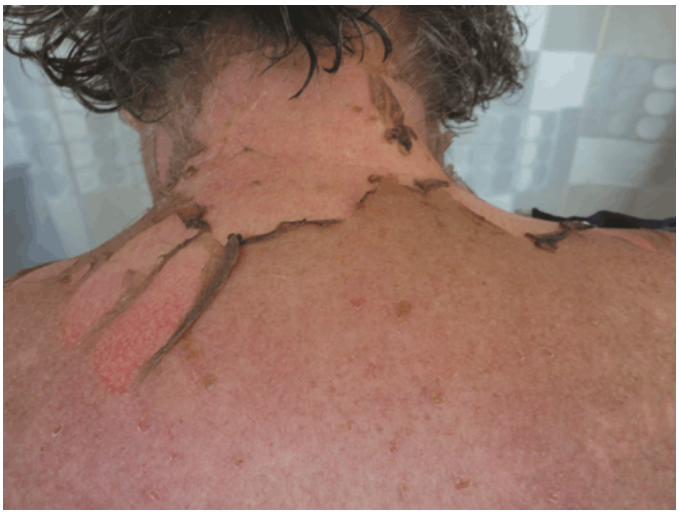


Figure 1: Neck and upper back – Note diffuse erythema and spontaneous sloughing of skin on the upper back.

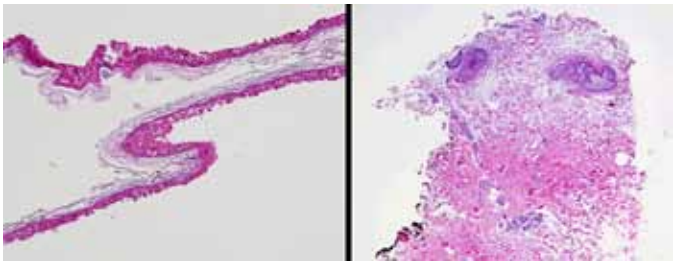


Figure 2: Punch biopsy specimen of skin. (A) Epidermis demonstrating full-thickness epidermal necrosis (H&E stain, x200), (B) Dermis demonstrating sparse perivascular lymphocytic infiltrate (H&E stain, x100).

Antibody inhibition of CTLA-4 is associated with immune-related adverse events (IRAEs), mainly affecting the skin, gastrointestinal, and endocrine systems. Cutaneous IRAEs occur after 2 to 3 weeks, gastrointestinal IRAEs occur after 6 to 7 weeks, and endocrinologic IRAEs have been described after an average of 9 weeks. A dose-dependent increase in the frequency of IRAEs of any grade has been noted [2].

Dermatologic adverse effects include rash, pruritus, and vitiligo. The incidence of all-grade rash in patients receiving ipilimumab was 24.3%. The overall incidence of high-grade rash was 2.4% [1]. Cutaneous lesions associated with ipilimumab have been described as itchy, erythematous, discrete, mildly scaly papules coalescing into thin plaques on the trunk and proximal aspects of extensor extremities. The head and neck can be involved, while palms and soles are usually spared. Koebnerization may occur. Patients can develop a peripheral eosinophilia. On histology, a perivascular CD4+ T cell infiltrate with eosinophils is seen in the superficial dermis. Epidermal spongiosis and, rarely, dyskeratosis are also observed. Other skin findings include alopecia of the scalp, eyebrows, face, pubic region, and trunk as well as a photosensitive eruption [6]. Additionally, anti-CTLA-4 antibodies may stimulate an immune response against melanocytes. The development of vitiligo in a subset of patients and the identification of Melan-A-specific $CD8^+$ T lymphocytes near apoptotic melanocytes in biopsy specimens supports this theory [2, 6].

Stevens–Johnson syndrome and toxic epidermal necrolysis is a rare yet severe cutaneous adverse effect of ipilimumab. Recommended management of SJS and TEN involves early removal of the causative drug. Supportive therapy is initiated with protective measures for the exposed skin and mucosa, early detection and management of infection, fluid and nutritional support, and pain control [3]. Although initiation of prednisone at 1–2 mg/kg is recommended in patients with high grade IRAEs due to ipilimumab [2], caution is urged as the benefit of systemic corticosteroids in the treatment of SJS-TEN is debated and their use may be detrimental [3].

CONCLUSION

Stevens–Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are rare, but potentially fatal occurrences while undergoing ipilimumab therapy. The TEN has a high risk of mortality due to infections and retrospective data supports early referral to a burn unit. Given the increasing use of ipilimumab in patients with

metastatic melanoma, it is important for oncologists and dermatologists to be aware of the significant risk and potential severity of cutaneous adverse effects associated with the medication.

****

Acknowledgements
We would like to acknowledge Dr. Rishi R. Patel, Assistant Professor at the Ronald O. Perelman Department of Dermatology and the Department of Pathology at NYU Langone Medical Center for his contribution to this case. Dr. Rishi R. Patel provided histological confirmation of the diagnosis. Dr. Rishi R. Patel did not receive any financial compensation for his contribution.

Author Contributions
Mohini Pathria – 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
Jyoti Mundi – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Joshua Trufant – 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. Minkis K, Garden BC, Wu S, Pulitzer MP, Lacouture ME. The risk of rash associated with ipilimumab in patients with cancer: a systematic review of the literature and meta-analysis. J Am Acad Dermatol 2013 Sep;69(3):e121–8.
2. Weber JS, Kähler KC, Hauschild A. Management of immune-related adverse events and kinetics of response with ipilimumab. J Clin Oncol 2012 Jul 20;30(21):2691–7.
3. Chave TA, Mortimer NJ, Sladden MJ, Hall AP, Hutchinson PE. Toxic epidermal necrolysis: current evidence, practical management and future directions. Br J Dermatol 2005 Aug;153(2):241–53.
4. Baitsch L, Baumgaertner P, Devêvre E, et al. Exhaustion of tumor-specific CD8? T cells in metastases from melanoma patients. J Clin Invest 2011 Jun;121(6):2350–60.
5. Leach DR, Krummel MF, Allison JP. Enhancement of antitumor immunity by CTLA-4 blockade. Science 1996 Mar 22;271(5256):1734–6.
6. Jaber SH, Cowen EW, Haworth LR, et al. Skin reactions in a subset of patients with stage IV melanoma treated with anti-cytotoxic T-lymphocyte antigen 4 monoclonal antibody as a single agent. Arch Dermatol 2006 Feb;142(2):166–72.

SUGGESTED READING

- Hoos, A., Ibrahim, R., Korman, A., et. al. Development of Ipilimumab: Contribution to a New Paradigm for Cancer Immunotherapy. Semin Oncol. 2010; 37:533-546.
- Voskens CJ, Goldinger SM, Loquai C, Robert C, Kaehler KC, et al. (2013) The Price of Tumor Control: An Analysis of Rare Side Effects of Anti-CTLA-4
- Therapy in Metastatic Melanoma from the Ipilimumab Network. PLoS ONE 8(1): e53745.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Small bowel obstruction in an adult patient with situs ambiguous and mid gut malrotation

Shwe Phyo Han, Jonathan Grassby

ABSTRACT

Introduction: Situs ambiguous or heterotaxy syndrome is defined as the abnormal positioning of internal viscera relative to the normal. Diagnosis in adult is extremely rare as 90–99% of the patients have severe cardiac abnormalities and die by the age of five years. Case Report: A 32-year-old male was presented to hospital with sudden onset abdominal pain, abdominal distension, vomiting and absolute constipation for one day. There was no other medical problems. Examination was also unremarkable. Abdomen was distended and generally tender. Per rectal examination showed empty rectum. Blood tests were unremarkable. Computed tomography scan of abdomen and pelvis showed closed loop mid to distal small bowel obstruction with small bowel wall thickening. Malrotation of the bowel was noted. It also showed that stomach and spleen were on the right side of the body. Emergency laparotomy showed small bowel volvulus and spleen, stomach, duodenojejunal flexure and small bowel were in the right side of the abdomen and colon was in the left side of the abdomen. Ileocecal valve was noted in the left side of the abdomen. Adhesiolysis and derotation of the affected small bowel were performed. Appendicectomy was performed due to its location in left lower quadrant. The patient was discharged from the hospital four days after the operation. Conclusion: There is no case report on adult situs ambiguous presented with acute intestinal obstruction before. We report this case for extremely rare occurrence of situs ambiguous with mid gut malrotation presented with small bowel obstruction in the adult age group.

Keywords: Abdominal pain, Mid gut malrotation, Situs ambiguous, Small bowel obstruction, Vomiting

How to cite this article

Han SP, Grassby J. Small bowel obstruction in an adult patient with situs ambiguous and mid gut malrotation. Int J Case Rep Imag 2016;7(5):303–305.

Article ID: Z01201605CR10640SH

****

doi:10.5348/ijcri-201652-CR-10640

Shwe Phyo Han[1], Jonathan Grassby[2]

Affiliations: [1]MBBS, MS, Surgical Registrar, Department of surgery, Dubbo Base Hospital, Dubbo, NSW, Australia; [2]MBBS, FRACS, Consultant surgeon, Department of Surgery, Dubbo Base Hospital, Dubbo, NSW, Australia.

Corresponding Author: Shwe Phyo Han, Unit 4, 3 Quinn Street, Dubbo, NSW, Australia, 2830; Email: sphmdy@gmail.com

Received: 05 January 2016
Accepted: 17 February 2016
Published: 01 May 2016

INTRODUCTION

Situs ambiguous or heterotaxy syndrome is defined as the abnormal positioning of internal viscera relative to the normal [1]. It is due to a primary defect in lateralization around day-28 of gestation, leading to a deviation from the normal position of viscera [1]. The incidence of situs ambiguous is approximately 1 in 40,000 live births [2]. Diagnosis in adult is extremely rare as 90–99% of the patients have severe cardiac abnormalities and die by the age of five years [2, 3].

CASE REPORT

A 32-year-old male was presented to Dubbo base hospital with sudden onset abdominal pain, abdominal distension, vomiting and absolute constipation for one day. There are no other medical problems. There was no previous abdominal operation. On examination, he was afebrile and observations were within normal limits. Abdomen is distended and soft. There was generalized tenderness in the abdomen. Per rectal examination showed empty rectum and no mass or lesion was noted. Blood tests were unremarkable.

Computed tomography scan of abdomen and pelvis with oral and IV contrast were performed. It showed closed loop mid to distal small bowel obstruction with small bowel wall thickening (Figure 1). Malrotation of the bowel was noted. It also showed that stomach and spleen was on the right side of the body (Figure 2 and Figure 3).

Emergency laparotomy was performed. Small bowel volvulus was noted. Spleen, stomach, duodenojejunal flexure and small bowel were in the right side of the abdomen and colon is in the left side of the abdomen. Ileocecal valve was noted in the left side of the abdomen. Adhesiolysis and derotation of the affected small bowel were performed. Appendicectomy was performed due to its location in left lower quadrant.

There was no complication after the operation and the patient was discharged from the hospital four days after the operation.

DISCUSSION

Situs ambiguous can be divided into two main types. They are left isomerism (situs ambiguous with polysplenia) and right isomerism (situs ambiguous with asplenia). Associated anomalies include congenital heart disease, IVC interruption with azygos or hemiazygos continuation (left sided isomerism), bilateral bilobed lungs (left sided isomerism), bilateral trilobed lungs (right sided isomerism), polysplenia (left sided isomerism), asplenia (right sided isomerism), midline liver and right sided stomach (left sided isomerism) [1, 2].

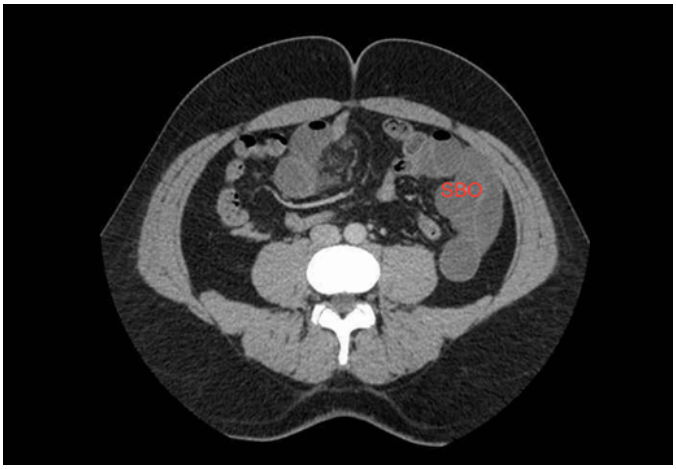


Figure 1: Computed tomography scan of abdomen and pelvis showing closed loop mid to distal small bowel obstruction with small bowel wall thickening.

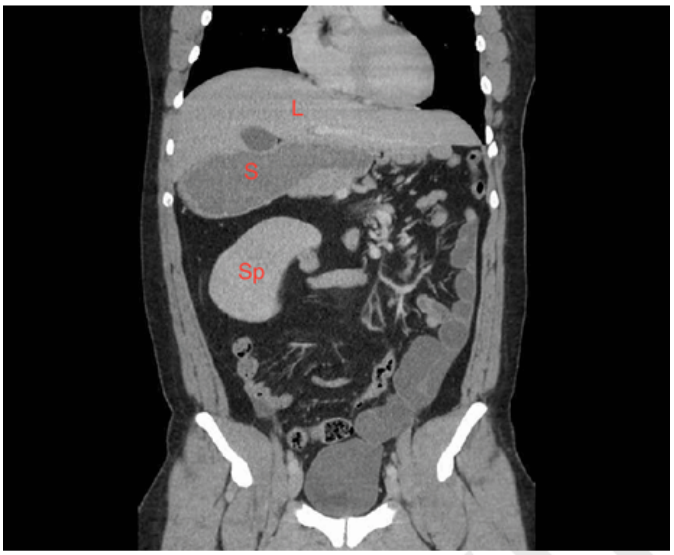


Figure 2: Coronal section of computed tomography scan showing stomach and spleen on the right side of the body.

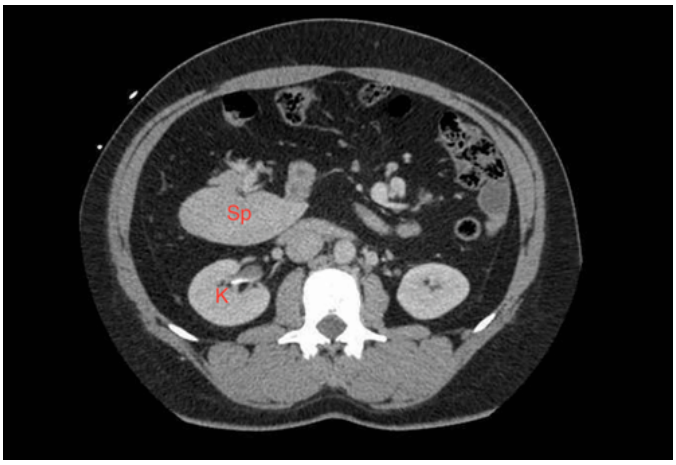


Figure 3: Cross-sectional computed tomography scan showing spleen on the right side of the body.

The number of spleens ranges from one to ten in left sided isomerism and spleens may be located either bilaterally or if unilateral, ipsilateral to the stomach because spleen arises from the dorsal mesogastrium. Patients with situs ambiguous can present in different ways due to different anatomical abnormalities that may be present. Most of the patients will not fit into the two categories described above and many patients will fall somewhere in between. Therefore, individual anatomical abnormalities should be documented rather than using the terms left and right sided isomerism. Thorough imagine studies are required to diagnose the anatomical abnormalities [1].

Congenital heart disease is the major cause of morbidity and mortality in situs patients. Patients with right sided isomerism have very high incidence of

congenital heart disease. Patients with situs ambiguous with asplenia have increased risks of sepsis secondary to encapsulated bacteria. Vaccination against these organisms is recommended [1].

Situs ambiguous may not cause any symptoms but they can present with confusing symptoms when they have common surgical problems such as appendicitis and cholecystitis [1].

Mid gut malrotation including the cases of non-rotation is seen in 1 in 500 births. It is associated with situs ambiguous in 70% of the cases. It arises from failure of counter clockwise rotation around the superior mesenteric artery [1]. Intestinal malrotation in adult is rare with an occurrence between 0.0001–0.19%. Presentation with chronic abdominal pain, vomiting, diarrhea, abdominal distension and early satiety is common. Acute presentations with intestinal obstruction due to small bowel volvulus are less common [4].

CONCLUSION

There are few cases of situs ambiguous reported which were incidentally found during operation and abdominal imaging. There is no case report on adult situs ambiguous presented with acute intestinal obstruction before. We report this case for extremely rare occurrence of situs ambiguous with mid gut malrotation presented with small bowel obstruction in the adult age group.

****

Author Contributions

Shwe Phyo Han – 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

Jonathan Grassby – 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. Mujo T, Finnegan T, Joshi J, Wilcoxen KA, Reed JC. Situs ambiguous, levocardia, right sided stomach, obstructing duodenal web, and intestinal nonrotation: a case report. J Radiol Case Rep 2015 Feb 28;9(2):16–23.
2. Lee JM, Lee HS, Kim CD. Situs ambiguous with left sided isomerism. Dig Liver Dis 2014 Jul;46(7):658.
3. Donatelli G, Costantino F, Dhumane P, et al. Endoscopic intragastric balloon: a bridge toward definitive bariatric surgical management of a morbidly obese patient with situs ambiguous and midgut malrotation (with videos). Gastrointest Endosc 2012 Jan;75(1):217–8.
4. Salústio R, Nabais C, Paredes B, Sousa FV, Porto E, Fradique C. Association of intestinal malrotation and Bochdalek hernia in an adult: a case report. BMC Res Notes 2014 May 13;7:296.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Idiopathic orofacial granulomatosis: A case report

Dhupar Anita, Carvalho Karla M., Spadigam Anita, Syed Shaheen

ABSTRACT

Introduction: Orofacial granulomatosis (OFG) is a lesser known disease entity, which presents a confounding spectrum of oral manifestations. It is a non-specific granulomatous inflammation caused by an elusive etiopathogenesis. It usually manifests as persistent and/or recurrent upper and/or lower labial enlargement. The clinical features, both extra-oral and intra-oral, are highly variable and at times, insidious. This disease can be both cosmetically and functionally debilitating. Case Report: This is a report of an unusual case of orofacial granulomatosis which presented as a persistent severe generalized gingival enlargement in a nine-year-old child. There was no evidence of an underlying allergic or systemic cause. Surgical intervention (i.e., gingivectomy) showed no alleviation of symptoms. Conclusion: Making a prompt and precise diagnosis of orofacial granulomatosis is often challenging, however complete remission is possible with targeted therapy.

Dhupar Anita[1], Carvalho Karla M.[2], Spadigam Anita[3], Syed Shaheen[4]

Affiliations: [1]MDS, Professor; Department of Oral and Maxillofacial Pathology, Goa Dental College & Hospital, Bambolim, Goa, India; [2]MDS, Lecturer; Department of Oral and Maxillofacial Pathology, Goa Dental College & Hospital, Bambolim, Goa, India; [3]MDS, Professor and Head; Department of Oral and Maxillofacial Pathology, Goa Dental College & Hospital, Bambolim, Goa, India; [4]MDS, Assistant Professor; Department of Oral and Maxillofacial Pathology, Goa Dental College & Hospital, Bambolim, Goa, India.

Corresponding Author: Dr. Karla Maria Carvalho, Liberty Apartments, Flat No: 202, Feira Alta, Mapusa, Goa, India, 403507; Email: karlamariac@gmail.com

Received: 05 January 2016
Accepted: 17 February 2016
Published: 01 May 2016

Keywords: Crohn's disease, Diagnosis, Orofacial granulomatosis, Treatment

How to cite this article

Anita D, Carvalho KM, Anita S, Shaheen S. Idiopathic orofacial granulomatosis: A case report. Int J Case Rep Imag 2016;7(5):306–310.

Article ID: Z01201605CR10641DA

****

doi:10.5348/ijcri-201653-CR-10641

INTRODUCTION

A granuloma is a distinct histological entity, formed as an immunological response to a chronic non-degradable product of active hypersensitivity [1]. Orofacial granulomatosis (OFG) is a term used to describe granulomatous lesions affecting the orofacial tissues in the absence of provable systemic granulomatous conditions such as sarcoidosis or Crohn's disease [2]. Till date, some authors consider OFG to be a non-specific disease because of several overlapping clinical and histopathological features with other granulomatous conditions, in particular with Crohn's disease [3–6]. The unknown etiology of both orofacial granulomatosis and Crohn's disease further challenges efforts in categorizing the disease entities [2, 7].

The diagnostic dilemma tackles the question: Is OFG an early oral manifestation of Crohn's disease, or is it a distinct disease entity? This is a rare case of OFG in a child which presented solely as persistent gingival enlargement.

CASE REPORT

A nine-year-old girl, presented with a chief complaint of progressive gingival enlargement in both upper and lower jaws which she had noticed since the past 1 year. She was extremely conscious about her smile as she was being teased in school. She underwent two gingivectomy surgeries, at two different occasions, for both, the upper and lower dental arches, in the span of 1 year. A positive history of bleeding on brushing, difficulty in mastication due to impingement of enlarged gingiva was elicited. There was no relevant systemic history noted. Diet history showed that the patient favored cocoa based foods and showed a daily intake of the same.

Detailed oral examination revealed gross enlargement of upper and lower labial gingiva. The marginal, papillary and interdental gingiva of both jaws (i.e. labial and palatal/lingual) appeared erythematous, with a soft consistency and a granular surface texture (Figure 1). She had dental restorations in her lower right and left first permanent molar and lower right second permanent molar, which were a month old (Figure 2). Periodontal pockets were absent. Permanent canines in both arches were seen in the erupting stage (Figure 3). Examination of other oral sites which included lips, tongue and faucial pillars showed no signs of any abnormalities.

The preliminary investigations included a complete hemogram, bleeding time, clotting time, erythrocyte sedimentation rate (ESR), fasting blood sugar levels, tests for tuberculosis (i.e., sputum for acid fast bacilli, chest radiograph and Mantoux test), renal and liver function tests. Except for a mild eosinophilia (10%) observed on the hemogram, all other results were normal.

An incisional biopsy of the enlarged gingiva, revealed granulomas, multinucleated giant cells, dystrophic calcifications, sheets of plasma cells and lymphocytes .The lymphocytes had an atypical morphology (Hematoxylin and Eosin stain, periodic acid Schiff stain and van Gieson stain). No evidence of a deep fungal infection was seen (Figure 4).

A pediatric consultation followed. Splenomegaly was detected by a positive Castell's sign. A bone marrow aspirate was requested which was negative for malignancy. A normal abdominal ultrasound and no contributory gastrointestinal symptoms ruled out Crohn's disease. Normal C-reactive protein (CRP), angiotensin converting enzyme (ACE) levels and chest radiograph, disproved a diagnosis of sarcoidosis and Wegener's granulomatosis.

Additional tests, such as, LE cell and antinuclear antibody (ANA) proved negative for a frank autoimmune dysfunction. To further support a diagnosis of exclusion, an immunohistochemical panel workup was done. CD 3 and CD 20 (in small lymphocytes), CD 68 (highlighted multiple granulomas) and CD 138 (in plasma cells) markers were found to be positive. A diagnosis of idiopathic orofacial granulomatosis was made (Figure 5).

The patient was advised to strictly eliminate cocoa from her diet. Systemic corticosteroid (i.e., prednisolone) regime with regular follow-up appointments was prescribed which showed immediate positive results

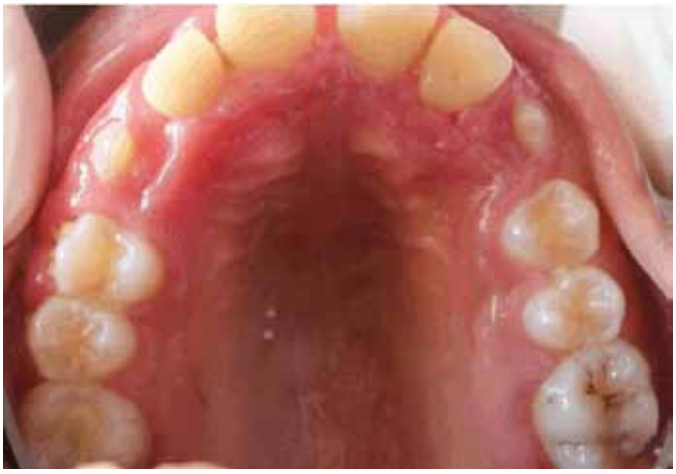


Figure 1: Erythematous enlarged labial gingiva seen in upper and lower dental arches.

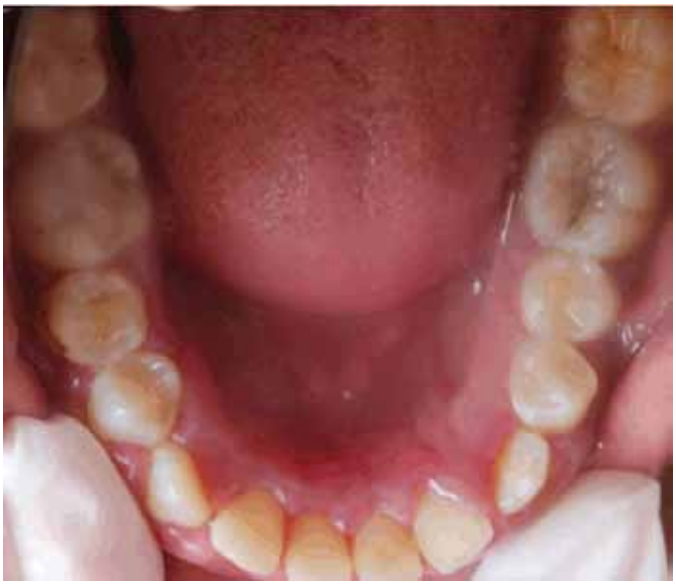


Figure 2: Dental restoration seen in lower right and left permanent molars. Gingiva is enlarged and inflamed in the anterior (incisor) segment of the lingual gingiva.

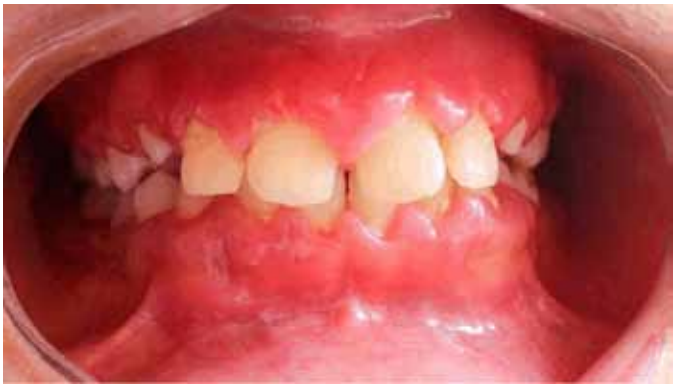


Figure 3: Erupting permanent canines in upper dental arch and mild palatal gingival enlargement seen.

within the first week of treatment. A minimal dose of 1 mg/kg/day for two weeks, followed by gradual taper for four weeks was prescribed. Simultaneous prophylaxis using co-trimoxazole was also initiated. Follow-up after three months showed an almost healthy and well contoured gingiva, in both upper and lower jaw arches (Figure 6A–B).

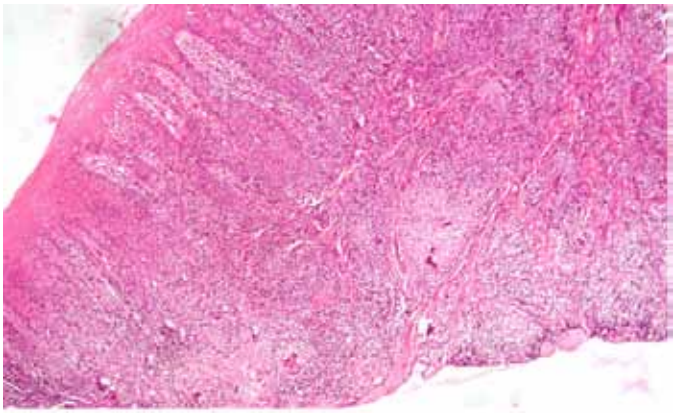


Figure 4: Biopsy section from upper right posterior labial Gingiva showing non-caseating granuloma formation (H&E stain, x100).

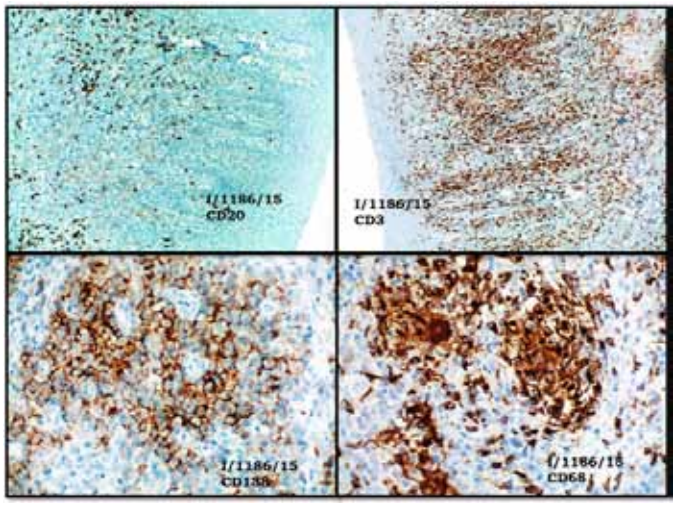


Figure 5: Immunohistochemical panel showing positive CD 3, CD 20, CD 68 and CD 138 markers (Magnifications: x100).

Figure 6: Three month post-treatment follow-up showing complete regression of lesion (A) Labial gingiva, (B) Palatal.

DISCUSSION

An increase incidence of orofacial granulomatosis is seen globally, particularly in the pediatric age group [6]. However, despite advanced diagnostic techniques, it is considered to be a diagnosis of exclusion [3, 5, 8, 9]. Recent evidence has shown OFG to be an immunologically induced granulomatous disorder [1].

The most common clinical symptom is persistent labial enlargement which is non-pitting, and non-tender, involving upper and/ or lower lips [6]. The other oral and facial manifestations include oral ulcers, fissured tongue, mucosal tags, gingival enlargement, facial nerve palsy, facial swelling and cervical lymphadenopathy [2, 8]. Our case mimicked some of the cases studied by Endo et al. and Gale et al., in that; gingival hyperplasia was the sole manifestation [10].

There are a number of theories, both genetic and immunological, which attempt to explain the course and clinical outcome of this disease [6]. Current research in OFG supports an immunologic or allergen triggering factor as the chief aetiological basis [10].

It is on the basis of the above mentioned research, that the association between Crohn's disease (CD) and OFG is still being debated. It is hypothesized that OFG is a subtype of CD since 20–50% of patients with OFG have concurrent intestinal manifestations and 6.4% have a positive family history of CD. In our case, it is also relevant to note that a known 40.4% of children affected with OFG fall victims to CD during their lifetime [9]. Andrew Zbar et al. in their recent review stated that a frank labial enlargement, exfoliative angular stomatitis and oral ulcerations clinically distinguish OFG from CD. These authors have advocated a complete evaluation for underlying systemic conditions in those OFG cases showing gingival enlargement alone [6]. In our case, suspected leukemia induced gingival hyperplasia was refuted by a normal bone marrow aspirate.

The immunohistochemical evaluation showing a predominance of CD3+ T cells support a diagnosis of OFG without systemic Crohn's disease. It has also been suggested that IgE-expressing B cells (CD20+) in the submucosa of OFG patients play an important role in the pathogenesis and provide a link to type IV hypersensitivity, particularly in cases of OFG without CD in young individuals [10]. This may be reflected by the high expression of CD138+ plasma cells. Since cocoa is a proven triggering factor in a number of OFG cases, the daily intake of cocoa based food by the patient could be contributory to these immunological findings. The multinucleated giant cells of macrophage origin were highlighted by the CD68+ marker. Besides a complete absence of gastrointestinal manifestations, the above mentioned immunological findings do not support a diagnosis of Crohn's disease for this case. Therefore a colonoscopy was not advised [6].

A biopsy showing non-caseating granuloma formation is critical for differentiating OFG from all other

granulomatous disease [6]. The differential diagnosis, include Tuberculosis, Sarcoidosis, allergic reaction, deep fungal infection, Wegener's granulomatosis and a leukemic infiltrate, which were proved negative by the investigative protocol followed [3]. "Idiopathic orofacial granulomatosis" is thus a suitable diagnosis for this case of OFG, given that it satisfies the criteria given by Tilakaratne et al. [6].

The treatment for OFG, given its unknown aetiology is non-specific and subjective [2]. In this case, the elimination of a potential allergen (i.e. cocoa), was advocated before prescribing any medication, however, there was no change in the clinical picture [2, 8]. Corticosteroids with or without the use of an immunosuppressant, is considered to be the first line and the mainstay of the recommended treatment regime [2, 3, 8].

Intra-lesional corticosteroid therapy though ideal in this patient, was not possible as she could not afford it. Systemic corticosteroid therapy in a growing child can result in several adverse effects which include growth retardation [6]. However, the dose given to this patient was minimum and advocated for a short duration of time. A strict follow-up schedule at regular intervals ensures a complete systemic review of the patient during the course of the treatment.

Newer drugs which have proved to be effective are thalidomide, tacrolimus, infliximab and adalimomab (i.e. recombinant monoclonal antibiotic against TNF-K). These drugs can be included in therapy based on the clinical course and severity of the disease [8].

Recurrences and subsequent systemic granulomatous manifestations are relatively common in cases of OFG restricted to the oral cavity. A mandatory regular clinical review for these idiopathic OFG cases is therefore strongly advised. A multidisciplinary approach involving a team made up of a dental surgeon, gastroenterologist and pediatrician (when applicable) is essential to provide timely and targeted care [6]. The clinical, histopathological and immunological findings of this case, separate OFG from Crohn's disease. This case thus supports the hypothesis of these two disease entities being independent sub categories caused by underlying idiopathic immunological dysfunction.

CONCLUSION

It is important to recognize and consider orofacial granulomatosis as a differential diagnosis for idiopathic gingival enlargement cases, with/without other allied oral manifestations. The growing incidence of Crohn's disease, particularly in the pediatric and adolescent age group, warrants an exhaustive investigative analysis for suspected OFG, so as to deliver early targeted treatment.

****

Author Contributions

Dhupar Anita – Conception and design, Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
Carvalho Karla M. – Conception and design, drafting the article, Acquisition of data, Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
Spadigam Anita – Analysis and interpretation of data, Critical revision of the article, Final approval of the version to be published
Syed Shaheen – Acquisition of data, 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.

Copyright

REFERENCES

1. James DG. A clinicopathological classification of granulomatous disorders. Postgrad Med J 2000 Aug;76(898):457–65.
2. Leão JC, Hodgson T, Scully C, Porter S. Review article: orofacial granulomatosis. Aliment Pharmacol Ther 2004 Nov 15;20(10):1019–27.
3. Kauzman A, Quesnel-Mercier A, Lalonde B. Orofacial granulomatosis: 2 case reports and literature review. J Can Dent Assoc 2006 May;72(4):325–9.
4. Lazzerini M, Bramuzzo M, Ventura A. Association between orofacial granulomatosis and Crohn's disease in children: systematic review. World J Gastroenterol 2014 Jun 21;20(23):7497–504.
5. Kim SK, Lee ES. Orofacial granulomatosis associated with Crohn's disease. Ann Dermatol 2010 May;22(2):203-5.
6. Ravindran R, Karunakaran A. Idiopathic orofacial granulomatosis with varied clinical presentation. Case Rep Dent 2013;2013:701749.
7. Motemayel AF, Abdolsamadi HR, Irani S, Bardal R. Orofacial granulomatosis of the upper lip: A case report. Dent J Hamadan Univ Med Sci 2009;1(1):37–40.
8. Javadzadeh A, Pakfetrat A, Falaki F, Seyyedi SA. Approach to orofacial granulomatosis and review of literature. Journal of Islamic Dental Association of IRAN 2012;24(1):111–21.

9. Manjunatha BS, Agrawal A, Deepti A, Shah V. Orofacial Granulomatosis: A Review. International Journal of Oral and Maxillofacial Pathology 2013;4(4):16–22.

10. Endo H, Rees TD. Cinnamon products as a possible etiologic factor in orofacial granulomatosis. Med Oral Patol Oral Cir Bucal 2007 Oct 1;12(6):E440–4.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Dorsal penile frenulum: A rare developmental abnormality

Bhavinder Arora

ABSTRACT

Introduction: Dorsal penile frenulum is a very rare abnormality in literature. Case Report: This abnormality was present with normal external urethral meatus in a young adult male. There was no associated penile torsion. Conclusion: This is the second case report in medical literature. A brief description of preputial anatomy and frenar band band is described. Various hypotheses have been discussed for development of dorsal penile frenulum.

Keywords: Dorsal penile frenulum, Penile frenulum, Penile torsion

How to cite this article

Arora B. Dorsal penile frenulum: A rare developmental abnormality. Int J Case Rep Imag 2016;7(5):311–313.

Article ID: Z01201605CR10642BA

****

Bhavinder Arora

Affiliations: MS FIAMS FAIS, Associate Professor, Department of Surgery, Pt BDS University of health sciences, PGIMS Rohtak, India.

Corresponding Author: Bhavinder Arora, MS FIAMS FAIS, Associate Professor, Department of Surgery, Pt BDS University of health sciences, PGIMS Rohtak, India; Email: drbhavinderarora@gmail.com

Received: 15 November 2015
Accepted: 15 March 2016
Published: 01 May 2016

doi:10.5348/ijcri-201654-CR-10642

INTRODUCTION

Preputial anatomy and frenar band

In classical description of prepuce, it is divided into outer and inner (mucosal) layer. The outer layer is a continuation of the shaft skin, up to the rim of junction with the mucosa at the opening of the prepuce at rest. The mucosa or inner preputial skin from the junctional or transitional zone to the sulcus where it continues as the mucosal covering of the glans [1]. Just inside the junctional rim of prepuce at the dorsal aspect is a transverse band of ridges 10–15 mm wide. Except the area of this band rest of the mucosa is smooth. The ridged band is continuous with the frenulum, being a radial band over 60% of the penile diameter–the dorsal and lateral aspects–then turning distally to form a V-shape on the ventral aspect whilst reducing its width to merge with the frenulum [1]. Dorsal penile frenulum is a rare developmental abnormality; only one such case is available in literature [2]. We report another case of dorsal penile frenulum with normal penile anatomy, normal ventral frenulum but a dorsal frenulum present since birth.

CASE REPORT

A 24-year-old male presented with complaint of dyspareunia. There was no history of balanoposthitis in childhood. On examination, the size of penis, scrotum and testis was found to be normal. The external urinary meatus was at its normal position. On retracting the prepuce, there was a band connecting dorsum of glans to prepuce, V-shaped and double layered. A normal ventral frenulum was present. Frenuloplasty of the dorsal penile frenulum was done which led to retraction of normal

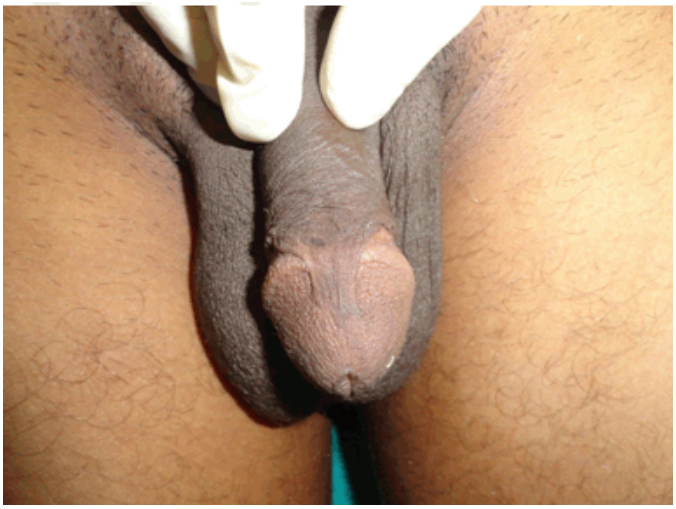


Figure 1: Dorsal penile frenulum with normal external uretheral meatus.

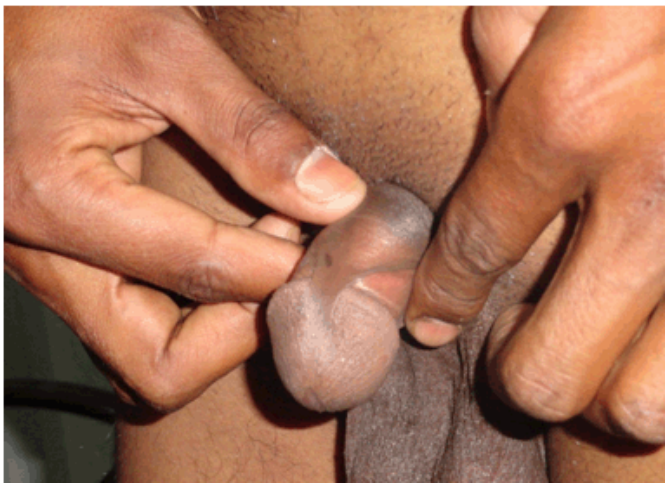


Figure 2: Lateral view of dorsal frenulum.

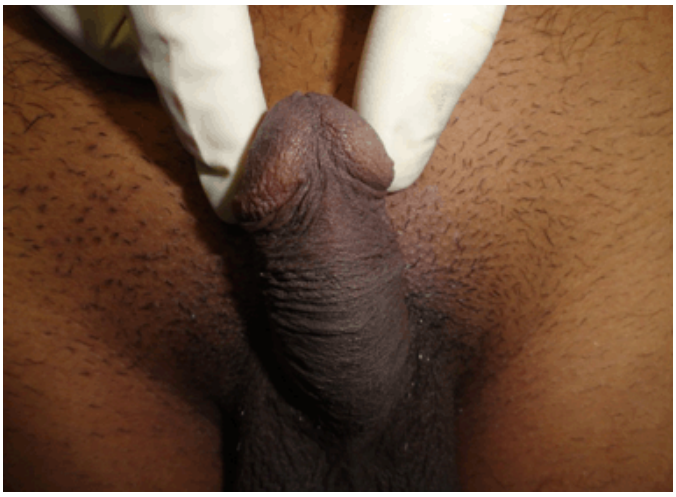


Figure 3: Normal ventral frenulum and median raphe.

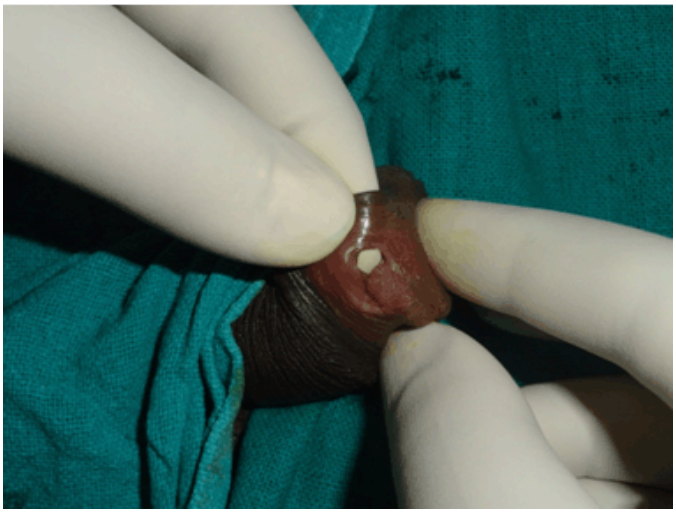


Figure 4: Double layered dorsal frenulum.

prepuce. In postoperative period, patient remained asymptomatic.

DISCUSSION

The embryological development of penile frenulum is related to the development of prepuce. The prepuce is formed by reduplication of the ectoderm covering the distal part of the phallus in third month of fetal life. The prepuce separates from the glans to form preputial sac and residual adhesion of ectoderm to the glans penis on the ventral aspect persists and is called frenulum [3]. The preputial and urethral folds fuse on the ventrum of the glans as the frenulum. Failure of fusion of the urethral folds blocks development of prepuce ventrally especially the frenulum [4]. Penile torsion is a rotation of the shaft of penis, usually to the left (counterclockwise) direction. It results in the urethral meatus being placed in oblique position, such that median raphe makes a spiral curve from the base of the penis to the meatus [5]. Singla et al. Postulated that probably during the embryological development of preputial sac, the latter separated from the glans on all sides except on the dorsal side and residual adhesions on dorsal side formed the dorsal frenulum [2].

CONCLUSION

I agree with this theory of adhesion during embryological life because

- Frenulum thus formed was two layered
- It was adherent to the dorsum of glans at one point only

Simple division of dorsal frenulum led to restoration of normal prepuce.

****

Author Contributions

Bhavinder Arora – 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. Mcgrath K. The Frenular Delta. In: Denniston GC, Hodges FM, Milos MF eds. Understanding Circumcision: A Multi-Disciplinary Approach to a Multi-Dimensional Problem. New York: Kluwer Academic/Plenum Publishers; 2001.
2. Singla SL, Garg P, Bishnoi PK, Singh B. Dorsal penile frenulum. Urol Int 1997;59(1):55–6.
3. Williams PL, Warwick R, Dyson M, Bannister LH. Gray's anatomy. 37ed. Edinburgh: Churchill Livingstone; 1989. p. 733–4.
4. Max M. Normal and Anomalous Development of the Urinary Tract. In: Walsh PC eds. Campbell's Urology. 7ed. London: WB Saunders; 1998. p. 1582–3.
5. Shaeer O. Torsion of the penis in adults: prevalence and surgical correction. J Sex Med 2008 Mar;5(3):735–9.

ABOUT THE AUTHOR

Article citation: Arora B. Dorsal penile frenulum: A rare developmental abnormality. Int J Case Rep Imag 2016;7(5):311–313.

Dr. Bhavinder Arora is Professor Associate at Department of Surgery, Pt BDS PGIMS, University of Health Sciences, Rohtak, India. He earned MBBS degree from PGIMS, Rohtak and postgraduate degree from MS (General Surgery) from PGIMS, Rohtak, India.

He has published 50 research papers in national and international academic journals and authored five chapters in books. His research interests are in peripheral vascular surgery, Day Surgery, Inguinal Hernia, Trauma and critical care. He has delivered ORATIONS in national conferences. He is a national faculty in Association of Surgeons of India, Association of Thoracic and Vascular Surgeons of India, The Indian association of Day Surgery of India. He has read about 50 research papers in national and international conferences and won four research paper awards.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Phlegmasia cerulea dolens complicated by methylenetetrahydrofolate reductase genetic mutation

Jason A. Fried, Lauren M. Wright

ABSTRACT

Introduction: Deep venous thrombosis (DVT) is the formation of a blood clot within a deep vein. Phlegmasia cerulea dolens (PCD) represents a critical acute consequence of DVT. The PCD is a condition caused by massive iliofemoral thrombosis that produces severe venous congestion and obstruction of arterial flow; eventually, causing ischemia in the affected extremity. The treatment goals of PCD are to restore venous outflow by removing thrombus burden, prevent additional thrombus formation, and maintain collateral circulation. However, no therapeutic algorithms exist for PCD. Case Report: We report PCD in a 55-year-old male with a significant past medical history for multiple venous thromboembolisms, requiring placement of an inferior vena cava filter and lifetime anticoagulation. Clinical presentation and accompanying venous duplex results led to the diagnosis of PCD. The 9th American College of Chest Physicians Consensus Conference on Antithrombotic and Thrombolytic Therapy created guidelines for treatment of acute DVT in the absence of gangrene. Guidelines advise to withhold thrombolysis and percutaneous or surgical procedure until treatment with therapeutic heparin anticoagulation proves to be inadequate. Conservative treatment with therapeutic anticoagulation was unsuccessful. therefore, catheter directed thrombolytic therapy, venoplasty, and stent placement were implemented. Conclusion: Due to possible associated morbidity and mortality, it is recommended to implement therapy soon after diagnosis of PCD. It is hoped that this report will provide guidance in management and assist to develop an evidence-based treatment algorithm for PCD.

Keywords: Deep venous thrombosis, Hypercoagulable, Phlegmasia cerulea dolens, Thrombolysis

How to cite this article

Fried JA, Wright LM. Phlegmasia cerulea dolens complicated by methylenetetrahydrofolate reductase genetic mutation. Int J Case Rep Imag 2016;7(5):314–317.

Article ID: Z01201605CR10643JF

****

doi:10.5348/ijcri-201655-CR-10643

Jason A. Fried[1], Lauren M. Wright[2]

Affiliations: [1]DO, USA Division Chief of General Surgery at Western Reserve Hospital, Cuyahoga Falls Surgical Associates, Cuyahoga Falls, OH; [2]DO, General Surgery Resident, Western Reserve Hospital Cuyahoga Falls, OH, USA.

Corresponding Author: Lauren Wright, DO, 1900 23rd St., Cuyahoga Falls, OH, 44223; Email: lwright@westernreservehospital.org

Received: 06 December 2015
Accepted: 28 January 2016
Published: 01 May 2016

INTRODUCTION

Deep venous thrombosis (DVT) is the formation of a blood clot within a deep vein [1]. Principles of Virchow's triad—venous stasis, hypercoagulability, and endothelial injury—are postulated as the cause of venous

thrombosis. Non-specific physical examination findings of the extremity may include pain, swelling, erythema, prominent superficial veins, pain with passive dorsiflexion, and peripheral cyanosis [2]. Phlegmasia cerulea dolens (PCD) represents a critical acute consequence of DVT. PCD—"blue, painful leg" or "blue phlebitis" [3]—is a condition caused by massive iliofemoral thrombosis that produces severe venous congestion and obstruction of arterial flow; eventually, causing ischemia in the affected extremity [2, 3]. The PCD is an uncommon but fulminate manifestation of venous thrombosis, with high morbidity and mortality, and without a developed standard of care [3–5].

CASE REPORT

A 55-year-old male presented to the emergency department complaining of back pain of one day duration. Patient described the pain as severe, dull, with radiation to left buttock. Associated symptoms are paresthesias of bilateral lower extremities, left lower quadrant abdominal pain, and shortness of breath. Patient was a non-smoker with a past medical history significant for coronary artery disease, myocardial infarction status post four vessel coronary artery bypass graft, diabetes mellitus, congestive heart failure, cocaine abuse with cessation six months prior to admission, and multiple venous thromboembolisms requiring placement of an inferior vena cava filter and lifetime anticoagulation—rivaroxaban.

Physical examination revealed tenderness to palpation of left abdomen, bilateral flanks, and lumbar spine. Vascular examination included bilateral warm lower extremities, capillary refill less than two seconds, palpable femoral and posterior tibial (PT) pulses with dorsalis pedis (DP) Doppler signals, and no neurologic deficits. Vital signs and cell counts were within normal limits with a lactic acid of 3.1 mmol/L. Computed tomography (CT) scan of the abdomen and pelvis with intravenous and oral contrast revealed distal para-aortic and left retroperitoneal stranding, interpreted as a probable hematoma.

Patient was admitted to the medicine service with orders to hold anticoagulant therapy. On the second hospital day, patient complained of increasing bilateral leg pain—"tightness"—with subsequent change in vascular examination to bilateral bluish lower extremities which were edematous, tender to palpation, delayed capillary refill, no palpable lower extremity pulses, and bilateral PT Doppler signals. Sensation and motor function remained intact; however, he was unable to bear weight due to extreme pain. Vascular surgery was consulted. Computed tomography angiogram of the aorta with bilateral run-offs revealed diminished blood flow and atherosclerotic changes in the vessels below the knee, more apparent on the left than the right. Lower extremity venous duplex showed DVTs of bilateral lower extremities from the common femoral to distal posterior tibial, peroneal, and gastrocnemius veins.

The patient had not been on anticoagulant therapy due to concerns related to the retroperitoneal hematoma. However, considering the above duplex and clinical findings of PCD, the benefits outweighed the risks and therapeutic heparin [80 units/kg IV bolus, then continuous infusion of 18 units/kg/hr] with leg elevation was initiated. The following day, an interval CT of the abdomen and pelvis with intravenous contrast confirmed stability of the retroperitoneal hematoma. After five days of therapeutic heparin, the patient's clinical response was inadequate due to progressive pain and discoloration of bilateral lower extremities; therefore, decision was made to provide catheter directed thrombolytic therapy. He was taken to the vascular suite for venogram (Figure 1) and a Cragg–McNamera® (Micro Therapeutics Inc., Irvine, CA) catheter was placed for directed thrombolytic therapy (catheter Alteplase rate of 0.5 mg/hr and sheath heparin rate of 500 U/hr). Serial coagulation profiles were monitored and thrombolytic therapy was adjusted accordingly. Postoperative day-one, the patient returned to the vascular suite for interval venography, which revealed decrease in thrombus burden. Thrombolytic therapy was continued and patient returned to the vascular suite postoperative day-two. Venogram showed a patent right iliac vein and vena cava with IVC filter intact. The left external iliac vein had residual thrombus; therefore, venoplasty with 14/100 LifeStar® stent (Bard Peripheral Vascular, Tempe, Arizona) was deployed with subsequent expansion with a 14/40 Atlas® balloon (Bard Peripheral Vascular, Tempe, Arizona). Completion venography revealed a patent venous system with no extravasation or residual stenosis (Figure 2).

Postoperative course consisted of symptomatic treatment and serial vascular examinations. Etiology of condition remained unclear; therefore, hematology/oncology was consulted to exclude underlying malignancy and hypercoagulable state. Patient was found to be hypercoagulable due to a genetic mutation of methylenetetrahydrofolate reductase. The patient was discharged on postoperative day-six, with resolution of presenting symptoms and instructions for resumption of daily activities without functional limitations and continuation of vitamin-K-antagonist. Follow-up at 2nd month and 20th month revealed no recurrence of clinical symptoms.

DISCUSSION

Pathogenesis

Phlegmasia cerulea dolens (PCD) is a rare syndrome with pathogenesis of massive iliofemoral thrombosis causing total or near total venous occlusion. Subsequently, substantial increase in venous pressure produces parallel increase in tissue pressure with consequential fluid sequestration, edema, compromise of arterial circulation,

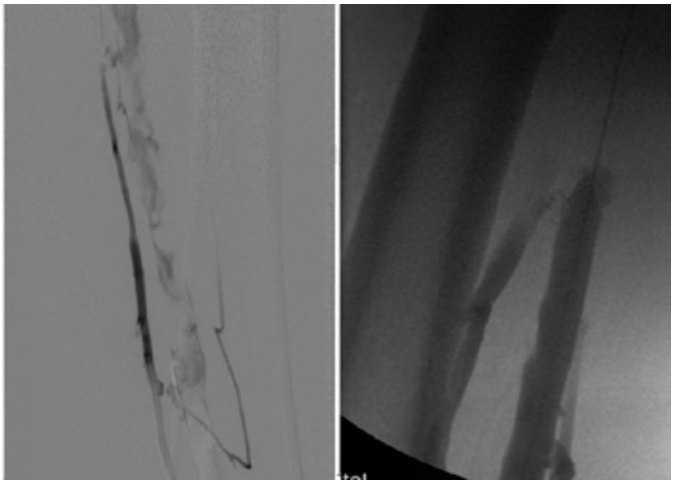


Figure 1: Venogram—extensive iliofemoral occlusive deep venous thrombosis. (A) Right femoral vein, and (B) Left femoral vein.

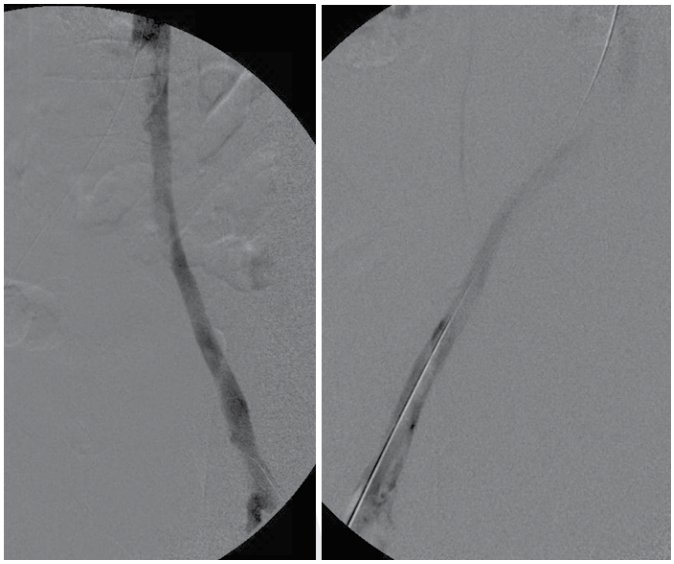


Figure 2: Venogram—completion venogram showing improved patency and flow after 48 hours catheter directed thrombolysis. (A) Right iliac vein and vena cava, and (B) Left iliac vein—venoplasty and stent placement.

and ischemia in the affected extremity [2–4]. Risk factors for development of DVT into PCD include malignancy, hypercoagulable states, previous DVT, trauma, inferior vena cava filter, contraceptive agents, and venous stasis [5]. Of these, malignancy is the most common etiology, with reported rates of 33% [4]. Prompt diagnosis and implementation of treatment is the basis to prevent complications such as gangrene [40–60%], amputation [20–50%], and death [25–40%]—pulmonary embolism being responsible for 30% of deaths [3–6].

Diagnosis

Phlegmasia cerulea dolens is classified based on severity—non-complicated, impending venous gangrene, or venous gangrene [4]. Diagnosis is established by physical examination and duplex ultrasonography (US) [3]. The PCD is clinically defined as a triad of acute extremity edema, cyanosis, and ischemic pain [5]. Ultrasonography is considered first-line imaging due to sensitivity and specificity in diagnosis of symptomatic proximal DVTs, 97% and 94% respectively; detection of occlusion in arteries and veins; and ability to characterize flow [7, 8]. Alternative diagnostic imaging being CT scan and magnetic resonance imaging scan, with catheter venography and arteriography being the gold standard due to ability to confirm diagnosis, help direct treatment from mapping of circulation, and be therapeutic [8].

Surgical strategy

The treatment goals of PCD are to restore venous outflow by removing thrombus burden, prevent additional thrombus formation, and maintain collateral circulation [4, 5]. No therapeutic algorithms exist for PCD. However, the 9th American College of Chest Physicians Consensus Conference on Antithrombotic and Thrombolytic Therapy created guidelines for the treatment of acute DVT in the absence of gangrene. Guidelines advise to withhold thrombolysis and percutaneous or surgical procedure until treatment with therapeutic heparin anticoagulation proves to be inadequate [9]. Conservative therapies for PCD involve elevation of the affected extremity, therapeutic heparin anticoagulation, and fluid resuscitation [3]. If clinical improvement inadequate or massive thrombosis with impending gangrene at presentation, thrombolysis is indicated. Thrombectomy—percutaneous or surgical—is implemented when there is contraindication to thrombolysis or in conjunction with thrombolysis; furthermore, percutaneous transluminal angioplasty with or without stenting and/or fasciotomy are supplementary treatment options depending resolution of thrombus burden and symptoms [3–6].

Defining optimal treatment with current knowledge is impracticable due to the paucity of data and quality studies [3, 10]. An international registry has been suggested to record all cases of PCD, which would assist with creation of an evidence-based approach to treatment [3]. Overall, further structured reporting of PCD is needed in order to augment understanding and education of the efficacy and safety of treatment strategies.

CONCLUSION

Phlegmasia cerulea dolens (PCD) is an uncommon but critical acute consequence of deep venous thrombosis (DVT). Due to the possible associated morbidity and mortality, prompt diagnosis and implementation of therapy are recommended. It is hoped that this report will provide guidance in management and assist to develop an evidence-based treatment algorithm for PCD.

****

Author Contributions

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

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Deep vein thrombosis. 2015. [Available at: http://www.merriam-webster.com]
2. Crnenwett JL, Johnston KW. Acute Deep Venous Thrombosis: Pathophysiology and Natural History. In: Vandy FC, Wakefield TW eds. Rutherford's Vascular Surgery. 7ed. Philadelphia: Saunders/Elselvier; 2010. P. 736–52.
3. Klok FA, Huisman MV. Seeking optimal treatment for phlegmasia cerulea dolens. Thromb Res 2013 Apr;131(4):372–3.
4. Chinsakchai K, Ten Duis K, Moll FL, de Borst GJ. Trends in management of phlegmasia cerulea dolens. Vasc Endovascular Surg 2011 Jan;45(1):5–14.
5. Onuoha CU. Phlegmasia Cerulea Dolens: A Rare Clinical Presentation. Am J Med 2015 Sep;128(9):e27–8.
6. Lorimer JW, Semelhago LC, Barber GG. Venous gangrene of the extremities. Can J Surg 1994 Oct;37(5):379–84.
7. Greenfield LJ, Mulholand MW. Vascular diagnostics. In: Moneta GL ed. Greenfield's Surgery: Scientific Principles and Practice. 5ed. Philadelphia: Wolters Kluwer Health/Lippincott Williams & Wilkins; 2011. P. 1514–36.
8. Kalagher SD, Kane DD. Phlegmasia cerulea dolens: before and after lysis. Intern Emerg Med 2015 Feb;10(1):103–4.
9. Kearon C, Akl EA, Comerota AJ, et al. Antithrombotic therapy for VTE disease: Antithrombotic Therapy and Prevention of Thrombosis, 9th ed: American College of Chest Physicians Evidence-Based Clinical Practice Guidelines. Chest 2012 Feb;141(2 Suppl):e419S–94S.
10. Enden T, Haig Y, Kløw NE, et al. Long-term outcome after additional catheter-directed thrombolysis versus standard treatment for acute iliofemoral deep vein thrombosis (the CaVenT study): a randomised controlled trial. Lancet 2012 Jan 7;379(9810):31–8.

CASE REPORT PEER REVIEWED | OPEN ACCESS

A parathyroid cyst associated with an adenoma causing primary hyperparathyroidism (PHPT): Gamma camera localization for case with MIBI scan-negative PHPT

Takaaki Fujii, Reina Yajima, Hironori Tatsuki, Hiroyuki Kuwano

ABSTRACT

Introduction: A cystic lesion of the parathyroid gland causing hyperparathyroidism is an uncommon finding. Tc-99m MIBI scintigraphy is a useful preoperative diagnostic tool in primary hyperparathyroidism (PHPT). We report a rare case of a patient with PHPT with a functional parathyroid cyst in whom MIBI scintigraphy failed to detect a parathyroid tumor. Case Report: A 70-year-old Japanese man was referred to our hospital for the examination and treatment of hypercalcemia and a high level of intact PTH. Computed tomography scan, Magnetic resonance imaging scan, and ultrasonography of his neck showed a cystic mass in the lower right side of the neck measuring approximately 3.0×2.0 cm, whereas MIBI scintigraphy failed to detect a parathyroid tumor. Resection of the right cystic mass was performed. Scintigraphy images of the neck were acquired by an eZ-SCOPE hand-held gamma camera before the skin incision, and ex vivo imaging of the specimen was performed, which was useful for the navigation surgery and minimally invasive parathyroidectomy. Histopathology showed a parathyroid adenoma with prominent cystic degeneration. Conclusion: False-negative diagnostic results have been reported in cystic parathyroid adenomas. In cases of cystic hyperfunctioning scan-negative parathyroid tumors, the eZ-SCOPE may be useful for the localization and navigation surgery of primary hyperparathyroidism due to a cystic parathyroid tumor.

Keywords: Gamma camera, Navigation surgery, Primary hyperparathyroidism, Parathyroid cyst, Sestamibi scintigraphy

Takaaki Fujii[1], Reina Yajima[1], Hironori Tatsuki[1], Hiroyuki Kuwano[1]

Affiliations: [1]Department of General Surgical Science, Gunma University Graduate School of Medicine. Maebashi, Gunma 371-8511, Japan.

Corresponding Author: Takaaki Fujii, M.D., Ph.D., FACS, Department of General Surgical Science, Graduate School of Medicine, Gunma University, 3-39-22 Showa-machi, Maebashi, Gunma 371-8511, Japan; E-mail: ftakaaki@med.gunma-u.ac.jp

Received: 19 October 2015
Accepted: 23 November 2015
Published: 01 May 2016

How to cite this article

Fujii T, Yajima R, Tatsuki H, Kuwano H. A parathyroid cyst associated with an adenoma causing primary hyperparathyroidism (PHPT): Gamma camera localization for case with MIBI scan-negative PHPT. Int J Case Rep Imag 2016;7(5):318–322.

Article ID: Z01201605CR10644TF

****

doi:10.5348/ijcri-201656-CR-10644

INTRODUCTION

Primary hyperparathyroidism (PHPT) is a condition characterized by an excess secretion of parathyroid hormone by adenomatous or hyperplastic glands [1, 2]. The accurate preoperative localization of parathyroid adenomas is important to reduce the operative failure rate [2, 3]. Tc-99m sestamibi (MIBI) scintigraphy is a useful preoperative diagnostic tool for PHPT [4, 5].

The association of a functional parathyroid cyst with a parathyroid adenoma is an uncommon finding [6–9]. It was reported that in some patients with PHPT and a functioning parathyroid cyst, Tc-99m MIBI scintigraphy failed to detect a parathyroid tumor [6, 7]. There have also been some reports of radio-guided surgery for PHPT [10–19]. We recently demonstrated that in scan-positive patients identified by preoperative Tc-MIBI, Tc-MIBI scintigraphy with the use of a hand-held gamma camera, the eZ-SCOPE AN (Anzai Medical Co. Ltd., JAPAN), is useful for navigation surgery for PHPT and minimally invasive parathyroidectomies [18, 19].

The eZ-SCOPE AN is designed to be used as a hand-held, regional diagnostic imaging device, which is a new compact-type semiconductor gamma camera based on the use of a cadmium zinc telluride (CdZnTe) [18–21].We report here a rare case of a patients with a functioning parathyroid cyst in whom MIBI scintigraphy failed, but in whom a gamma camera could detect a parathyroid tumor

DEVICE DESCRIPTION

The eZ-SCOPE AN is designed to be used as a hand-held, regional diagnostic imaging device [17–20]. This compact-type semiconductor gamma camera made of CdZnTe has 256 semiconductors representing the same number of pixels. Each semiconductor is a 2-mm square and is located in 16 lines and rows on the surface of the detector. The outer dimensions of the camera are 74×72×210 mm, and it weighs 820 g. The most significant differences between it and previous cameras include the spatial resolution, sensitivity, high count rate characteristics, and energy resolution [18–21].

CASE REPORT

The patient was a 70-year-old Japanese man with a history of hepatocellular carcinoma with chronic hepatitis C. Hypercalcemia and a high level of intact parathyroid hormone (PTH) had been detected 27 months prior to his admission to our hospital. Neck ultrasonography and computed tomography (CT) scan revealed a right parathyroid tumor, but a Tc-99m MIBI scintigraphy examination failed to detect a parathyroid tumor. He had been followed up because he was also suffering from the hepatocellular carcinoma with liver cirrhosis. His serum calcium level had been significantly elevated, and thus he was referred to our hospital for further examination and treatment of hypercalcemia and the high level of intact PTH. On admission, his serum calcium was 14.4 mg/dL, albumin was 3.7 g/dL, and the intact PTH level was 423 pg/mL. Computed tomography scan of his neck showed a hypodense mass in the right side of the neck with peripherally enhancing walls measuring approximately 3.0×2.0 cm (Figure 1).

Magnetic response imaging (MRI) scan showed a mass in the right neck, which showed low to intermediate signal intensity on T1-weighted images and high signal intensity on T2 images compared to the surrounding tissues (Figure 2). Neck ultrasonography (US) revealed a cystic mass measuring 3.0×1.5 cm in the right neck (Figure 3). Tc-99m MIBI scintigraphy was performed to diagnose primary hyperparathyroidism and examine other parathyroid glands, but it failed to detect a parathyroid tumor (Figure 4). As there was suspicion of hyperparathyroidism due to the functioning parathyroid

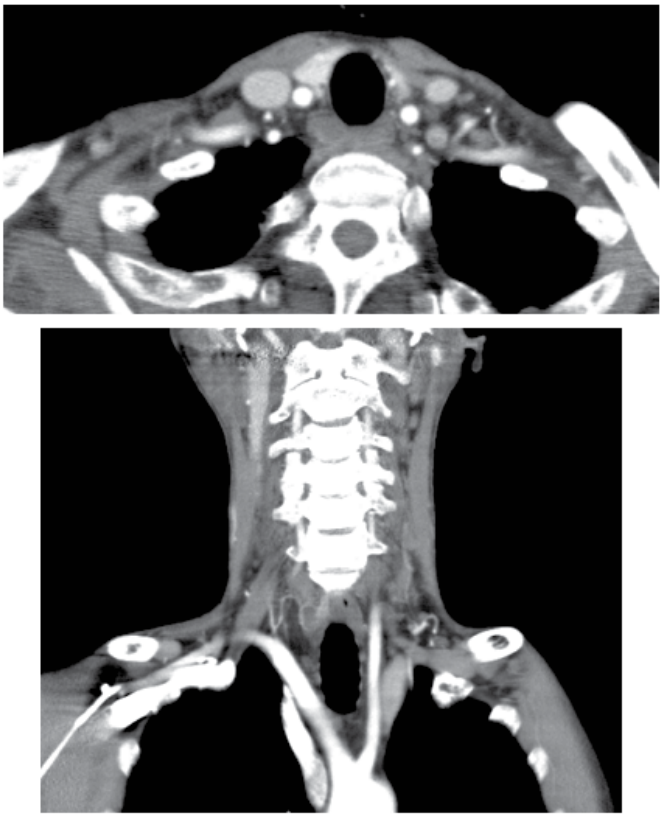


Figure 1: Computed tomography of neck showing a hypodense mass in the right side of the neck with peripherally enhancing walls measuring approximately 3.0 × 2.0 cm.

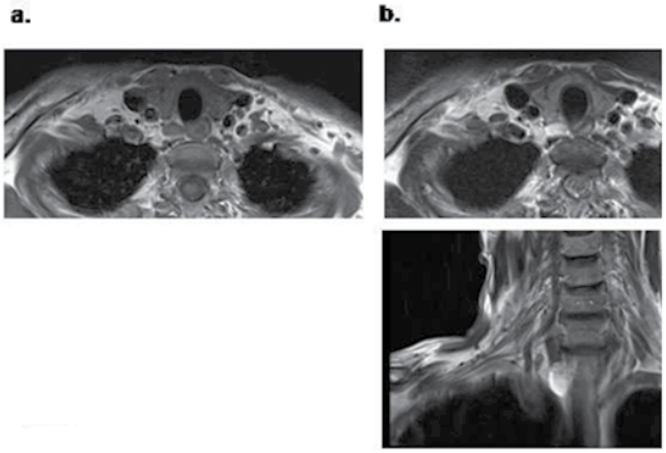


Figure 2: Magnetic resonance imaging showing a mass in the right neck which showed low to intermediate signal intensity on T1-weighted images (a) and high signal intensity on T2 images (b) compared to the surrounding tissues.

cyst, we performed resection of the right cystic mass. Scintigraphy images of the neck were acquired with the eZ-SCOPE AN before the skin incision, and ex vivo imaging of the specimen was performed (Figure 5). This gamma camera was useful for the navigation surgery and minimally invasive parathyroidectomy.

The intact PTH level was measured 10 min after the removal of the cystic mass, and we confirmed a decrease in the intact PTH level. Histopathology showed a parathyroid adenoma with prominent cystic degeneration (Figure 6). Our follow-up of the patient has remained uneventful, with no sign of recurrent disease.

DISCUSSION

Tc-99m MIBI scintigraphy has been widely used with high accuracy for the detection and localization of parathyroid tumors [4, 5]. Here we encountered a rare case of a patient with PHPT with a functioning parathyroid cyst in whom Tc-99m MIBI scintigraphy failed to detect a parathyroid tumor. Cystic lesions of the parathyroid gland are very uncommon, and the association of a functional parathyroid cyst with a parathyroid adenoma is an uncommon finding [6–9]; the most common cause is cystic degeneration of the adenomas, which is consistent with our case. Computed tomography scan, magnetic resonance imaging scan and ultrasonography could demonstrate our patient's cystic parathyroid adenoma, whereas the Tc-99m MIBI scintigraphy failed to detect the cystic adenoma.

Although the exact mechanisms of the visualization of abnormal and hyperfunctioning parathyroid tissue by MIBI scintigraphy are not clear, false-negative diagnostic results have been reported in cystic parathyroid adenoma [6, 7]. In our previous study, we found that for single adenomas and cases with positive Tc-MIBI scans, radio-guided surgery is an effective tool in the surgical management of primary hyperparathyroidism [18, 19]; however, in cases that are scan-negative due to cystic disease, radio-guided surgery is also thought to be useful.

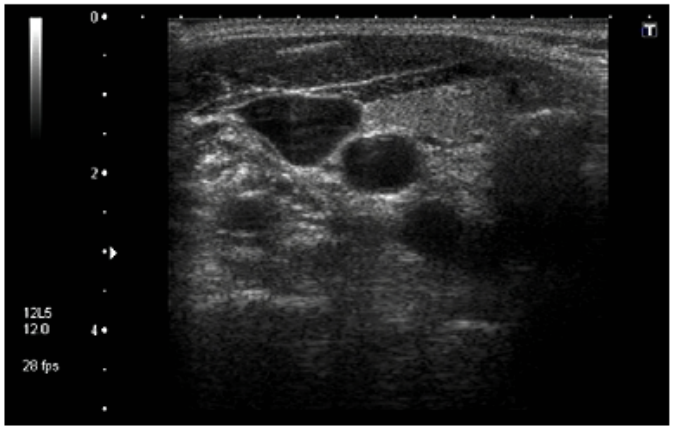


Figure 3 : Neck ultrasound revealed a cystic mass measuring 3.0×1.5 cm on the right of neck.

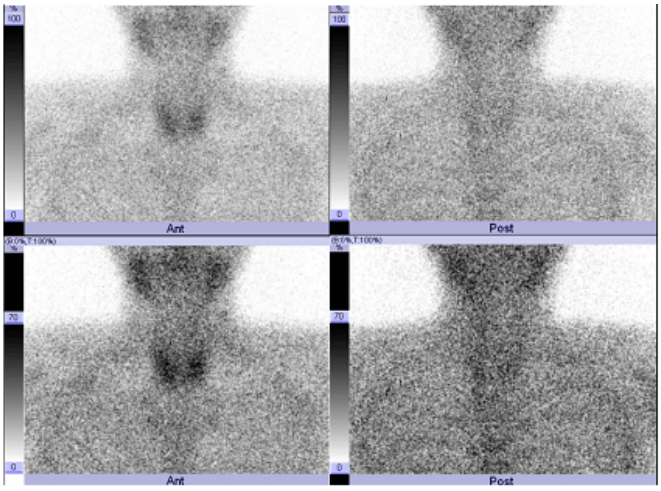


Figure 4: Tc-99m MIBI scintigraphy was performed to diagnose primary hyperparathyroidism and examine other parathyroid glands, but it failed to detect a parathyroid tumor.

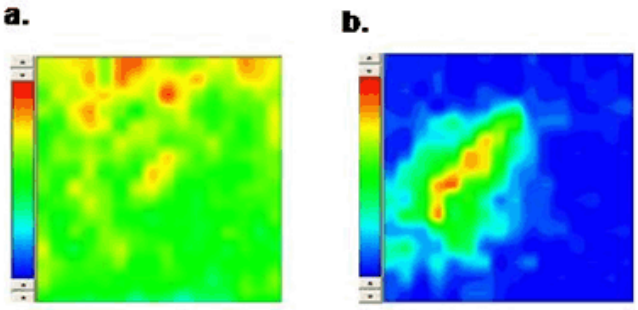


Figure 5: Scintigraphy images of the neck were acquired with the eZ-SCOPE AN before the skin incision (a), and ex vivo imaging of the specimen was performed (b). This gamma camera was useful for the navigation surgery and minimally invasive parathyroidectomy.

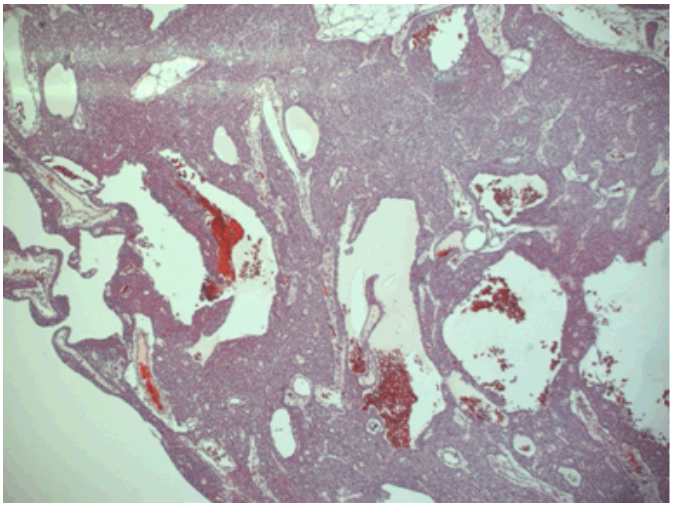


Figure 6: Histopathology revealed a parathyroid adenoma with prominent cystic degeneration (stain, x100).

CONCLUSION

We have reported a rare case of a patient with a cystic parathyroid adenoma presenting hyperparathyroidism in whom a gamma camera could detect the parathyroid tumor. Preoperative diagnoses can be challenging for cystic parathyroid adenomas. In some cystic hyperfunctioning scan-negative parathyroid tumor cases, the eZ-SCOPE AN may be useful for the localization and the navigation surgery of primary hyperparathyroidism due to a cystic parathyroid tumor.

****

Acknowledgements
We would like to thank Saitoh Y, Yano T, Matsui Y, Ishida A, Ishikubo A for their secretarial assistance.

Author Contributions
Takaaki Fujii – 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
Reina Yajima – 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
Hirnori Tatsuki – Substantial contributions to conception and design, Drafting the article, Final approval of the version to be published
Hiroyuki Kuwano – 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.

REFERENCES

1. Heath H 3rd, Hodgson SF, Kennedy MA. Primary hyperparathyroidism. Incidence, morbidity, and potential economic impact in a community. N Engl J Med 1980 Jan 24;302(4):189–93.
2. Seki K, Hashimoto K, Hisada T, et al. A patient with classic severe primary hyperparathyroidism in whom both Tc-99m MIBI scintigraphy and FDG-PET failed to detect the parathyroid tumor. Intern Med 2004 Sep;43(9):816–23.
3. Casas AT, Burke GJ, Mansberger AR Jr, Wei JP. Impact of technetium-99m-sestamibi localization on operative time and success of operations for primary hyperparathyroidism. Am Surg 1994 Jan;60(1):12–6; discussion 16–7.
4. Goldstein RE, Blevins L, Delbeke D, Martin WH. Effect of minimally invasive radioguided parathyroidectomy on efficacy, length of stay, and costs in the management of primary hyperparathyroidism. Ann Surg 2000 May;231(5):732–42.
5. Denham DW, Norman J. Cost-effectiveness of preoperative sestamibi scan for primary hyperparathyroidism is dependent solely upon the surgeon's choice of operative procedure. J Am Coll Surg 1998 Mar;186(3):293–305.
6. Ak I, Acikalin MF. Hyperparathyroidism with a functioning parathyroid cyst. Clin Nucl Med 2007 Sep;32(9):713–5.
7. Wirowski D, Wicke C, Böhner H, et al. Presentation of 6 cases with parathyroid cysts and discussion of the literature. Exp Clin Endocrinol Diabetes 2008 Aug;116(8):501–6.
8. Ardito G, Fadda G, Danese D, et al. Coexistence of a parathyroid adenoma and parathyroid cyst causing primary hyperparathyroidism. J Endocrinol Invest 2003 Jul;26(7):679–82.
9. McCoy KL, Yim JH, Zuckerbraun BS, Ogilvie JB, Peel RL, Carty SE. Cystic parathyroid lesions: functional and nonfunctional parathyroid cysts. Arch Surg 2009 Jan;144(1):52–6; discussion 56.
10. Goldstein RE, Billheimer D, Martin WH, Richards K. Sestamibi scanning and minimally invasive radioguided parathyroidectomy without intraoperative parathyroid hormone measurement. Ann Surg 2003 May;237(5):722–30; discussion 730–1.
11. Rubello D, Casara D, Saladini G, Piotto A, Pagetta C, Pelizzo MR. 99mTc-MIBI radio-guided surgery in primary hyperparathyroidism: a prospective study of 128 patients. Tumori 2002 May-Jun;88(3):S63–5.
12. Casara D, Rubello D, Pelizzo MR, Shapiro B. Clinical role of 99mTcO4/MIBI scan, ultrasound and intra-operative gamma probe in the performance of unilateral and minimally invasive surgery in primary hyperparathyroidism. Eur J Nucl Med 2001 Sep;28(9):1351–9.
13. Usmani S, Khan HA, al Mohannadi S, et al. Minimally invasive radionuclide-guided parathyroidectomy using 99mTc-sestamibi in patients with primary hyperparathyroidism: a single-institution experience. Med Princ Pract 2009;18(5):373–7.
14. Cassinello N, Ortega J, Lledo S. Intraoperative real-time (99m)Tc-sestamibi scintigraphy with miniature gamma camera allows minimally invasive parathyroidectomy without ioPTH determination in primary hyperparathyroidism. Langenbecks Arch Surg 2009 Sep;394(5):869–74.
15. Ortega J, Ferrer-Rebolleda J, Cassinello N, Lledo S. Potential role of a new hand-held miniature gamma camera in performing minimally invasive

parathyroidectomy. Eur J Nucl Med Mol Imaging 2007 Feb;34(2):165–9.
16. Kitagawa W, Shimizu K, Akasu H. Radioguided parathyroidectomy for primary hyperparathyroidism using the solid-state, multi-crystal gamma camera. Med Sci Monit 2003 Jun;9(6):CS53–6.
17. Casella C, Rossini P, Cappelli C, Nessi C, Nascimbeni R, Portolani N. Radioguided Parathyroidectomy with Portable Mini Gamma-Camera for the Treatment of Primary Int J Endocrinol 2015;2015:134731.
18. Fujii T, Yamaguchi S, Yajima R, et al. Use of a handheld, semiconductor (cadmium zinc telluride)-based gamma camera in navigation surgery for primary hyperparathyroidism. Am Surg 2011 Jun;77(6):690–3.
19. Fujii T, Yajima R, Yamaguchi S, Tsutsumi S, Asao T, Kuwano H. Could the eZ-SCOPE AN gamma camera replace intraoperative measurement of iPTH for PHPT? Int Surg 2012 Apr-Jun;97(2):99–103.
20. Abe A, Takahashi N, Lee J, et al. Performance evaluation of a hand-held, semiconductor (CdZnTe)-based gamma camera. Eur J Nucl Med Mol Imaging 2003 Jun;30(6):805–11.
21. Otake H, Higuchi T, Takeuchi Y, et al. Evaluation of efficiency of a semiconductor gamma camera. [Article in Japanese]. Kaku Igaku 2002 Nov;39(4):549–53.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Micro invasive papillary thyroid lesion in a post living donor liver transplant patient: A case report

Khaled Abdel Wahab, Essam Attia, Mohammad Arafa, Mohamed El Sorogy, Mohamed Abdel Wahab

ABSTRACT

Introduction: Liver transplant recipients are at increased risk of developing de novo malignancies. Skin cancers and lymphoma are common in adult transplant recipients, while post-transplant lymphoproliferative disorder (PTLD) is the most common concern in pediatric transplant recipients. De novo malignancy is a well-recognized complication of solid organ transplantation and associated immunosuppression. The development of such malignancies can be caused by a multifactorial combination of individual and regional predispositions to malignancy, pretransplantation disease states, recipient viral status, and the use and intensity of various immunosuppressive regimens to maintain allografts. Case Report: This report describes a papillary microinvasion of the thyroid in an adult Egyptian male following liver transplantation and declares the need for a high level of suspicion and careful investigation into any abnormal lesion in the long-term follow-up of solid organ transplant patients. Conclusion: Care should be taken towards any significant symptom or sign suspicious for malignancy for post-transplant patients. The idea of having a routine screening program to detect de novo tumors for those patients should be discussed more.

Keywords: De novo malignancy, Immunosuppressive therapy, Liver transplant, Thyroid lesion

Khaled Abdel wahab[1], Essam Attia[1], Mohammad Arafa[2], Mohamed El Sorogy[3], Mohamed Abdel Wahab[3]

Affiliations: [1]Surgical Oncology Unit, Mansoura Oncology center, Mansoura University; [2]Pathology Department, Mansoura University, Egypt; [3]Gastroenterology center, Mansoura university, Egypt.

Corresponding Author: Khaled Abdel Wahab, Lecturer of Surgical Oncology, Surgical Oncology Unit, Mansoura Oncology Center, Mansoura University, Egypt; Email: khaled14eg@hotmail.com

Received: 11 December 2015
Accepted: 23 January 2016
Published: 01 May 2016

How to cite this article

Wahab KA, Attia E, Arafa M, El Sorogy M, Wahab MA. Micro invasive papillary thyroid lesion in a post living donor liver transplant patient: A case report. Int J Case Rep Imag 2016;7(5):323–326.

Article ID: Z01201605CR10645KW

****

doi:10.5348/ijcri-201657-CR-10645

INTRODUCTION

Liver transplant recipients are at increased risk of developing de novo malignancies compared to the general population [1–3]. The reported incidence rates range between 3% and 15%, twice that of the general population [4–5].

Skin cancers and lymphoma are common in adult transplant recipients, while post-transplant lymphoproliferative disorder (PTLD) is the most common concern in pediatric transplant recipients [6].

This report describes a papillary micro invasion of the thyroid in an adult male patient following liver transplantation and declares the need for a high level of suspicion and careful investigation into any abnormal lesion in the long-term follow-up of solid organ transplant patients.

CASE REPORT

A 48-year-old Egyptian male had successfully undergone liver transplantation 18 months earlier for hepatocellular carcinoma on top of liver cirrhosis. He had a smooth postoperative period. Then, he used to take cyclosporine and mycophenolate sodium as immunosuppressive therapy.

Eighteen months post-transplant, the patient came with a thyroid nodule that was discovered accidentally after doing a neck ultrasound. There was no prior history of irradiation exposure or any family history of thyroid disease. There was no associated lymphadenopathy, and systemic examination did not reveal any significant findings. He was euthyroid with normal thyroid function.

Neck ultrasound revealed asymmetrically enlarged both thyroid lobes showing foci of calcification and increased perinodular vascularity with the largest nodule was on the left side measuring 1.8x1.2 cm.

Although the initial fine needle aspiration cytology (FNAC) results were inconclusive (colloid nodule with secondary hemorrhage), we decided to perform total thyroidectomy due to the sonographic suspicious criteria. Total thyroidectomy confirmed the diagnosis of micro invasive papillary carcinoma with no capsular and vascular invasion.

Grossly, the lesion appeared as solid firm whitish nodule of about 1.5 cm in diameter. Microscopically, it showed the typical features of papillary carcinoma of the thyroid. There were papillae showing complexity and branching. The papillae were lined by cuboidal cells with stratification. The nuclear features were mostly apparent in the form of ground glass (optically clear), overlapping and grooving (Figures 1 and 2)

The tumor was T1N0M0 on TNM staging with no lymph node invasion or distant metastases. Post-operative, suppressive dose of eltroxin was prescribed for the patient with close follow-up.

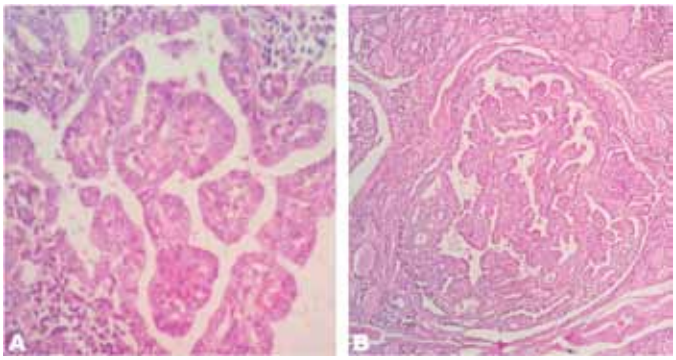


Figure 1 (A, B): Microscopic picture of the micro invasive papillary thyroid lesion.

DISCUSSION

Acquired immunodeficiency conditions are always associated with an increased risk for de novo malignancy. Organ transplantation is considered as an induced state of immunosuppression and, is commonly associated with higher probability for developing neoplasms [7].

De novo malignancy is a well-recognized complication of solid organ transplantation and associated immunosuppression. The risk of developing de novo malignancy after liver transplantation is about 1% per year, but the incidence varies between 3% and 15%, rates that are far greater than those in the general population [1–3].

The development of such malignancies can be caused by a multifactorial combination of individual and regional predispositions to malignancy, pretransplantation disease states, recipient viral status, the use and intensity of various immunosuppressive regimens to maintain allografts [8].

This complication is a major cause of late death in liver transplant recipients, responsible for 25% of the deaths occurring in patients who have survived more than three years post-transplantation [9].

The improvement in immunosuppression and better management of transplanted patients has led to a marked increase in post-LT survival. However, two main pitfalls exist: recurrence of HCV and the long-term side effects of immunosuppression [10].

Protocol for immunosuppressive drugs has changed in the past decades. Overall, the triple therapy for immunosuppression was the standard of care from 1991 to 1995 (cyclosporine, azathioprine and steroids). After 1995, significant changes were developed, consisting primarily in the use of more potent agents [mofetil mycophenolate (MMF), tacrolimus, sirolimus, anti-IL2 receptor antibodies] and the earlier stoppage of second line drugs, such as steroids [7].

Based on the recent introduction of newer and more potent immunosuppressive drugs in the LT arena, Benlloch et al. hypothesized, first, that the number of de novo tumors would increase in recent years; and second, that these tumors may follow a more aggressive behavior [7].

Schmilovitz et al. in their study failed to observe a correlation between tumor development and the type of immunosuppression, the occurrence of rejection episodes, or OKT3 use [11].

Accurate data such as the median interval between solid organ transplantation and the occurrence of the thyroid carcinoma, age range and sex ratio were not discussed in literature.

The patient was commenced on suppressive thyroxine supplements and thyroglobulin levels were monitored to detect recurrent or persistent disease following total thyroidectomy.

CONCLUSION

Care should be taken towards any significant symptom or sign suspicious for malignancy for post-transplant

patients. The idea of having a routine screening program to detect de novo tumors for those patients should be discussed more.

****

Author Contributions

Khaled Abdel Wahab – 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
Essam Attia – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Mohammad Arafa – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Mohamed El Sorogy – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Mohamed Abdel Wahab – 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. Benlloch S, Berenguer M, Prieto M, et al. De novo internal neoplasms after liver transplantation: increased risk and aggressive behavior in recent years? Am J Transplant 2004 Apr;4(4):596–604.
2. Finkenstedt A, Graziadei IW, Oberaigner W, et al. Extensive surveillance promotes early diagnosis and improved survival of de novo malignancies in liver transplant recipients. Am J Transplant 2009 Oct;9(10):2355–61.
3. Haagsma EB, Hagens VE, Schaapveld M, et al. Increased cancer risk after liver transplantation: a population-based study. J Hepatol 2001 Jan;34(1):84–91.
4. Debray D, Baudouin V, Lacaille F, et al. De novo malignancy after solid organ transplantation in children. Transplant Proc 2009 Mar;41(2):674–5.
5. Jonas S, Rayes N, Neumann U, De novo malignancies after liver transplantation using tacrolimus-based protocols or cyclosporine-based quadruple immunosuppression with an interleukin-2 receptor antibody or antithymocyte globulin. Cancer 1997 Sep 15;80(6):1141–50.
6. Penn I. De novo malignances in pediatric organ transplant recipients. Pediatr Transplant 1998 Feb;2(1):56–63.
7. Benlloch S, Berenguer M, Prieto M, et al. De novo internal neoplasms after liver transplantation: increased risk and aggressive behavior in recent years? Am J Transplant 2004 Apr;4(4):596–604.
8. Sanchez EQ, Marubashi S, Jung G, De novo tumors after liver transplantation: a single-institution experience. Liver Transpl 2002 Mar;8(3):285–91.
9. Pruthi J, Medkiff KA, Esrason KT, et al. Analysis of causes of death in liver transplant recipients who survived more than 3 years. Liver Transpl 2001 Sep;7(9):811–5.
10. Sapisochin G, Bilbao I, Dopazo C, et al. Evolution and management of de novo neoplasm post-liver transplantation: a 20-year experience from a single European centre. Hepatol Int 2011 Jun;5(2):707–15.
11. Schmilovitz-Weiss H, Mor E, Sulkes J, et al. De novo tumors after liver transplantation: a single-center experience. Transplant Proc 2003 Mar;35(2):665–6.

ABOUT THE AUTHORS

Article citation: Wahab KA, Attia E, Arafa M, El Sorogy M, Wahab MA. Micro invasive papillary thyroid lesion in a post living donor liver transplant patient: A case report. Int J Case Rep Imag 2016;7(5):323–326.

Khaled Mohamed Abdel Wahab is a lecturer of surgical oncology at Mansoura oncology center, Mansoura university in Egypt. He earned the undergraduate degree (M.B.B.CH) from Faculty of medicine, Mansoura university in 2005, Master degree of general surgery in 2010, Master of breast reconstruction from La Sapienza university in Rome, Italy in 2013 and MD degree from Mansoura faculty of medicine in 2015. He has published 13 research papers in national and international academic journals and is an editorial member in four international journals. His research interests include breast surgery and head and neck surgery.
E-mail: khaled14eg@hotmail.com

Essam Attia is working in Surgical Oncology Unit, Mansoura Oncology center, Mansoura University.

Mohammad Arafa is working in Pathology Department, Mansoura University, Egypt.

Mohamed El Sorogy is working in Gastroenterology center, Mansoura university, Egypt.

Mohamed Abdel Wahab is working in Gastroenterology center, Mansoura university, Egypt.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Gallbladder torsion: A case study

Yasser Arafat, Marianna Zukiwskyj, Tarana Lucky, Mojgan Rahimi, Polbert Diaz, Suntharalingam Shivananthan

ABSTRACT

Introduction: Torsion of the gallbladder is a rare condition and an important differential of an acute surgical abdomen. While early surgical intervention reduces the risk of disease progression to life-threatening complications, and thus avoids high morbidity and mortality, it is difficult to make a preoperative diagnosis of gallbladder torsion. Case Report: In this report, we describe a case of acute gallbladder torsion in an elderly male whose clinical presentation mimicked acute acalculous cholecystitis with local inflammatory reaction resulting from gangrenous gallbladder. The 81-year-old male was treated with emergency exploratory laparotomy. Except for developing an ileus requiring longer term recovery his post-operative period was otherwise uneventful and no further complications were evident in his follow-up clinic assessment. Conclusion: There is a wide range of differential diagnoses for patients presenting with acute surgical abdomen. Rare but life-threatening condition such as gallbladder torsion still remains a diagnostic challenge. Early diagnosis is the paramount in managing patients with gallbladder torsion as immediate surgical intervention is required. Keeping high index of clinical suspicion for patients presenting to emergency department with acute abdomen, especially in elderly population followed by diagnostic imaging can aid prompt diagnosis and thus facilitate early treatment.

Keywords: Abdominal pain, Acute abdomen, Gallbladder torsion, Gallbladder volvulus, Gastrojejunostomy

Yasser Arafat[1], Marianna Zukiwskyj[2], Tarana Lucky[3], Mojgan Rahimi[4], Polbert Diaz[5], Suntharalingam Shivananthan[6]

Affiliations: [1]Principal House Officer, Department of Surgery, Bundaberg Base Hospital, Bundaberg, Queensland 4670, Australia; [2]Registrar, Department of Surgery, Rockhampton Base Hospital, Rockhampton, Queensland 4700, Australia; 3Principal House Officer, Logan Hospital, Meadowbrook, Queensland 4131, Australia; [4]Locum Medical Officer, Ipswitch Hospital, Chelmsford Avenue, Ipswich, Queensland 4305, Australia; [5]Visiting Medical Officer, Department of Surgery, Hervey Bay Hospital, Hervey Bay, Queensland 4655, Australia 6Staff Specialist, Department of Surgery, Hervey Bay Hospital, Hervey Bay, Queensland 4655, Australia.

Corresponding Author: Yasser Arafat, Associate Lecturer, University of Queensland, & Principal House Officer, Department of Surgery, Bundaberg Base Hospital, Bundaberg, Queensland 4670, Australia; Email: dryarafat@gmail.com

Received: 27 December 2015
Accepted: 27 February 2016
Published: 01 May 2016

How to cite this article

Arafat Y, Zukiwskyj M, Lucky T, Rahimi M, Diaz P, Shivananthan S. Gallbladder torsion: A case study. Int J Case Rep Imag 2016;7(5):327–331.

Article ID: Z01201605CR10646YA

****

doi:10.5348/ijcri-201658-CR-10646

INTRODUCTION

Acute abdomen is a commonly encountered presentation in general surgical patient, for which there is a wide spectrum of surgical diagnoses. Torsion of

the gallbladder (also known as gallbladder volvulus) is an extremely rare but an important differential in the context of acute surgical abdomen. Since its initial description in 1898 by Wendel [1], only approximately 500 cases of gallbladder torsion have been reported in literature. In absence of early recognition and prompt surgical intervention gallbladder torsion may incur significant morbidity and mortality. The condition poses a diagnostic challenge preoperatively to both surgeons and radiologists, and often presents as a surprise to surgeons intraoperatively [2].

Gallbladder torsion occurs due to rotation of the gallbladder along the axis of the cystic duct and vascular pedicle. It can be either incomplete (rotation less than 180 degrees, usually gradual onset) or complete (rotation more than 180 degrees, usually acute onset) [3]. Both clockwise (occurs as a result of gastric and duodenal peristalsis) and anticlockwise rotations (secondary to colonic peristalsis) have been described [4].

While definite aetiology remains uncertain, multiple reasons for torsion of the gallbladder have been postulated by authors. In a review by Pottorf et al. [5], absence of gallbladder fixation to the liver resulting in excessive mobilization ability of gallbladder, relaxation and atrophy of a previously normal mesentery in the elderly (also known as visceroptosis) causing mesenteric elongation and thinning, atherosclerosis of the cystic artery and a tortuous cystic duct, and congenital anomalies that predispose individuals to elongated mesenteries have been reported as possible explanations.

The underlying pathophysiology related to congenital anomalies is thought to be secondary to a redundant wide mesentery or a mesentery that covers the cystic duct and artery. This causes the suspended gallbladder to twist along the axis of the cystic duct and cystic artery and result in a volvulus [6]. While peristaltic movement of the stomach and colon have also been reported as a possible attribute to the torsion of the gallbladder, the mechanism of movement that may actually cause the torsion still remain unclear. Loss of visceral fat with liver atrophy from conditions such as ageing may also result in acquired long mesentery which may explain the increased incidence of gallbladder torsion in elderly thinner population [5]. Gallbladder torsion is more frequently encountered in the elderly population with 85% of the cases reported between the ages of 60 and 80 years. While there is a female preponderance with a female to male ratio of 3:1, it is reported to be more common amongst boys than girls in the pediatric population, with a pediatric male : female ratio of 4:1 [7].

We report a case of gallbladder torsion in an elderly male which presented as a case of acute acalculous cholecystitis. This report also aims to review information currently available in literature relevant to the clinical aspect of this rare condition.

CASE REPORT

An 81-year-old male was presented to the emergency department with gradually worsening abdominal pain with symptoms of obstipation for approximately two weeks. He had presented to emergency department two days prior, and had been discharged with aperients, with little effect. His past medical history included gastrojejunostomy in 1968 for duodenal ulcers, appendicectomy many years before, and multiple spinal fusions.

He had a low-grade temperature measured by the ambulance staff. However, he was afebrile with a blood pressure of 95/58 mmHg and heart rate 86 bpm on presentation. On physical examination, signs of dehydration and diffuse tenderness and guarding in the right upper quadrant (RUQ) and epigastrium were noted. There was no palpable RUQ mass or jaundice, and the Murphy's sign was negative. Laboratory evaluations showed mild leukocytosis (10,700 /mm^3), and an elevated CRP of 327. Otherwise, the investigations including the liver function test were grossly normal.

Computed tomography (CT) scan with IV contrast on day of presentation revealed dilatation of intra- and extra-hepatic bile ducts, and a dilated and non-well-defined gallbladder with some free fluid adjacent to liver. However, there was no evidence of calculi and radiologist could not appreciate any specific cause to explain dilated bile ducts (Figure 1). An ultrasound scan was ordered in the context of diagnostic uncertainty, however was not carried out as his clinical condition deteriorated. Hence decision to operate was made based on physical examination, radiological findings, and presence of raised inflammatory markers.

An emergency exploratory laparotomy was performed, revealing copious pus and fibrin upon entry into the peritoneal cavity. The pericholecystic area was inflamed, and a torted and gangrenous, but un-perforated gallbladder was found (Figure 2). The rest of abdomen was unremarkable. The cystic duct was dissected and clipped and an intraoperative cholangiography (IOC) was performed which demonstrated a significantly dilated common bile duct and some kinking of the bile duct in the mid-third (Figure 3a). However, no filling defects was appreciated and free flow to the duodenum was noted (Figure 3b). Due to technical difficulty an antegrade cholecystectomy followed by copious peritoneal lavage was carried out. Drains were inserted to Morrison's pouch and pelvis, and abdomen was closed following satisfactory hemostasis. Histopathology of the surgical specimen showed an acute gangrenous cholecystitis.

Postoperatively, the patient developed an ileus which resolved with conservative management. His recovery was otherwise uneventful. The patient was discharged on the eighth postoperative day with a planned follow-up in the outpatient clinic in 4-6 weeks time. Follow-up clinic assessment was satisfactory with no evidence of any further short-term or long-term complications.

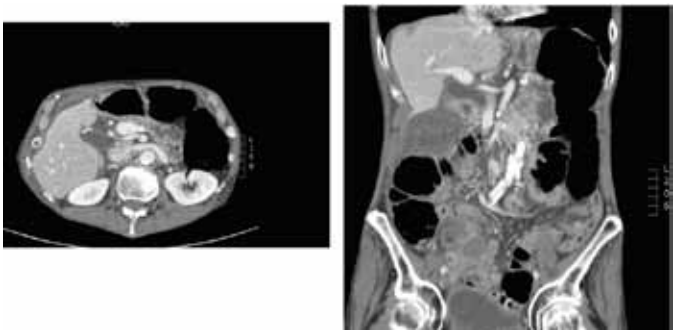


Figure 1: Abdomen computed tomography with intravenous contrast: a fluid filled, not well-defined structure noted inferior to the liver with small amount of fluid adjacent to the liver, likely dilated gallbladder.

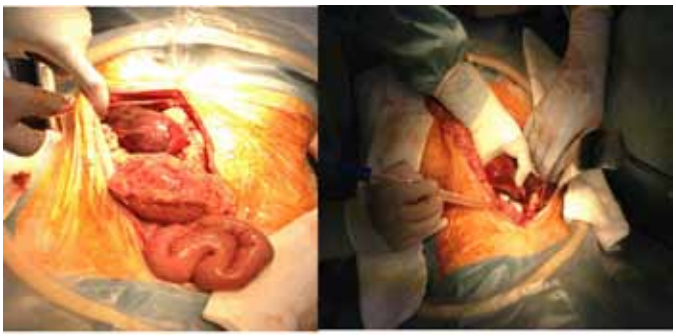


Figure 2: Operative findings: A torted and gangrenous gallbladder.

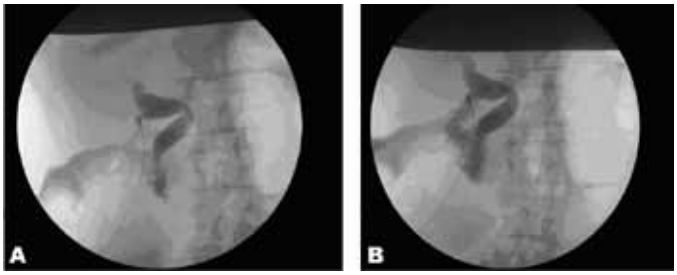


Figure 3: (A) Intraoperative cholangiogram showing dilated common bile duct and some kinking of the bile duct in the mid third but, (B) no filling defects and free flow to the duodenum.

DISCUSSION

Torsion of the gallbladder is a rare clinical condition of the hepatobiliary system, with a reported clinical incidence of 1 in 365,520 hospital admissions [2]. Review of the current literature shows that gallbladder volvulus commonly presents as an acute cholecystitis, although isolated cases of gallbladder torsion mimicking acute appendicitis have also been reported [6, 8]. Since no single clinical, serologic, or radiographic finding is pathognomonic, this condition is often misdiagnosed as acute acalculous cholecystitis, making a correct preoperative diagnosis challenging [4, 5, 9]. According to a review by Reilly et al. [4], a preoperative diagnosis of gallbladder torsion was made in 32 of 125 (26%) patients reported within the last 20 years and death as an outcome was reported in seven of the 113 patients presenting after 1991 representing a mortality rate of 6%. The main complications associated with delayed diagnosis and treatment includes necrosis, gangrene and subsequent perforation of the gallbladder resulting in contamination of the abdominal cavity with bilious material. Given the diagnostic dilemma and significant morbidity and mortality, gallbladder torsion should be considered as a differential diagnosis in any patients presenting with acute surgical abdomen, especially in those who show clinical deterioration despite antibiotic treatment.

The clinical features of gallbladder torsion usually include low grade fever and jaundice, poor response to antibiotic therapy, and acute onset of abdominal pain. These may be helpful in the differential diagnosis from acute cholecystitis. Moreover, a highly suggestive sign of gallbladder torsion observed by ultrasonography or CT is a markedly enlarged “floating” gallbladder with a continuous hypoechoic line indicating edematous change in the wall. Magnetic resonance studies (i.e. MRCP) can also be useful in establishing preoperative diagnosis. Diagnostic features in MRCP may include V-shaped distortion of extrahepatic bile duct, twisting interruption and tapering of the cystic duct, midline shift of the gallbladder with distension and enlargement, and also observation of different intensities of gallbladder, extrahepatic bile ducts, and cystic duct [3]. A triad of triads has also been reported in literature that is used to recognize potential gallbladder volvulus. Originally described by Lau et al., the clinical features for gallbladder torsion can be grouped into three triads. These include a triad of physical characteristics (usually thin built, elderly patient with either chronic chest disease or a spinal deformity); a triad of symptoms (relatively short history of presenting symptoms including abdominal pain and vomiting of early onset); and a triad of physical signs (presence of an abdominal mass along with absence of toxaemia or jaundice and a discrepancy between pulse and temperature). Lau et al. [10] suggested that if most of these features are present, torsion of the gallbladder should be a diagnostic consideration. As expected, our patient demonstrated most of the aforementioned clinical features.

Current literature suggests that an early emergency cholecystectomy should be performed when gallbladder torsion is suspected, preferably done using a laparoscopic approach as it is minimally invasive with a reported postoperative recovery period of 2–3 days [2, 6]. Moreover, compared to a classic cholecystectomy a laparoscopic approach for gallbladder volvulus is considered to be easier since a long mesentery is often present and the gallbladder is minimally adherent to the liver bed [2]. In our case, however, an exploratory laparotomy was performed. The decision to proceed with laparotomy was made in view of patient’s clinical presentation and subsequent rapid deterioration, gross distension of gallbladder evident through imaging, and

also diagnostic uncertainty. There was in fact a torted necrotic gallbladder which was removed, although there was no difficulty identifying the cystic duct and artery. While the postoperative period was otherwise uneventful he developed a postoperative ileus requiring a longer term hospital stay and recovery. It may be postulated that had the diagnosis of gallbladder torsion been considered preoperatively, laparoscopic approach would have been more favorable which would possibly have resulted a shorter recovery time.

CONCLUSION

The diagnosis of gallbladder torsion should be considered in all elderly patients presenting with symptoms suggestive of acute or non-resolving symptoms and signs of cholecystitis, particularly in the absence of gallstones. It is a rare condition and requires a high index of clinical suspicion. To achieve the best patient outcome, early diagnostic imaging and prompt surgical intervention are crucial. Recognition of the typical presentation described herein along with utilization of early diagnostic imaging studies can lead to accurate preoperative diagnosis. Thus, with early emergent cholecystectomy mortality and morbidity related to delayed treatment of this rare clinical condition can be reduced.

****

Author Contributions

Yasser Arafat – Substantial contributions to conception and design, Acquisition of data, Analyses and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of version to be published
Marianna Zukiwskyj – Substantial contributions to conception and design, Acquisition of data, Analyses and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of version to be published
Tarana Lucky – Substantial contributions to conception and design, Analyses and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of version to be published
Mojgan Rahimi – Substantial contributions to conception and design, Acquisition of data, Analyses and interpretation of data, Drafting the article, Final approval of version to be published
Polbert Diaz – Substantial contributions to conception and design, Analyses and interpretation of data, Revising it critically for important intellectual content, Final approval of version to be published
Suntharalingam Shivananthan – Substantial contributions to conception and design, Analyses and interpretation 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

REFERENCES

1. Wendel AV. VI. VI. A Case of Floating Gall-Bladder and Kidney complicated by Cholelithiasis, with Perforation of the Gall-Bladder. Ann Surg 1898 Feb;27(2):199–202.
2. Janakan G, Ayantunde AA, Hoque H. Acute gallbladder torsion: an unexpected intraoperative finding. World J Emerg Surg 2008 Feb 22;3:9.
3. Pu TW, Fu CY, Lu HE, Cheng WT. Complete body-neck torsion of the gallbladder: a case report. World J Gastroenterol 2014 Oct 14;20(38):14068–72.
4. Reilly DJ, Kalogeropoulos G, Thiruchelvam D. Torsion of the gallbladder: a systematic review. HPB (Oxford) 2012 Oct;14(10):669–72.
5. Pottorf BJ, Alfaro L, Hollis HW. A Clinician's Guide to the Diagnosis and Management of Gallbladder Volvulus. Perm J 2013 Spring;17(2):80–3.
6. Vedanayagam MS, Nikolopoulos I, Janakan G, El-Gaddal A. Gallbladder volvulus: a case of mimicry. BMJ Case Rep 2013 Jan 17;2013. pii: bcr2012007857.
7. Kitagawa H, Nakada K, Enami T, et al. Two cases of torsion of the gallbladder diagnosed preoperatively. J Pediatr Surg 1997 Nov;32(11):1567–9.
8. Bauman Z, Ruggero J, Lim J. Gallbladder Volvulus Presenting as Acute Appendicitis. Case Rep Surg 2015;2015:629129.
9. Nakao A, Matsuda T, Funabiki S, et al. Gallbladder torsion: case report and review of 245 cases reported in the Japanese literature. J Hepatobiliary Pancreat Surg 1999;6(4):418–21.
10. Lau WY, Fan ST, Wong SH. Acute torsion of the gall bladder in the aged: a re-emphasis on clinical diagnosis. Aust N Z J Surg 1982 Oct;52(5):492–4.

CASE REPORT PEER REVIEWED | OPEN ACCESS

A case report on cervico-medullary epidermoid tumor presenting with hydrocephalus

Sunil Munakomi, Binod Bhattarai, Iype Cherian

ABSTRACT

Introduction: Epidermoid tumors in the cervico-medullary junction are rare entities. Their presentation with features of acute hydrocephalus is a rare epiphenomenon. Case Report: Herein we discuss a rare tumor in the posterior fossa presenting with features of hydrocephalus. We discuss the diagnostic modalities, differential diagnosis and management undertaken in the same. Conclusion: Epidermoids can present with features of acute hydrocephalus. In them, surgical removal of the lesion is the therapeutic target. Specific problems pertaining to them is the insinuating nature of the lesion to surrounding neurovascular structures that may preclude its complete removal. Chemical meningitis, pseudo-meningocele and recurrence may complicate the postoperative period.

Keywords: Epidermoid tumor, Loss of consciousness, Posterior fossa, Hydrocephalus, Weakness

How to cite this article

Munakomi S, Bhattarai B, Cherian I. A case report on cervico-medullary epidermoid tumor presenting with hydrocephalus. Int J Case Rep Imag 2016;7(5):332–335.

Sunil Munakomi[1], Binod Bhattarai[2], Iype Cherian[3]

Affiliations: [1]MCh resident, College of Medical Sciences, Bharatpur, Nepal; [2]Associate Professor College of Medical Sciences, Bharatpur, Nepal; [3]Head of the Department, College of Medical Sciences, Bharatpur, Nepal.

Corresponding Author: Munakomi S., MCh resident, College of Medical Sciences, Bharatpur, Nepal, Email: sunilmunakomi@gmail. com

Received: 05 January 2016
Accepted: 22 February 2016
Published: 01 May 2016

Article ID: Z01201605CR10647SM

****

doi:10.5348/ijcri-201659-CR-10647

INTRODUCTION

Intracranial epidermoid tumors are rare congenital inclusion cysts constituting 0.2–1.8% of primary intracranial neoplasms [1, 2]. First described by a French artist [3], the first full description of this entity was given by the French pathologist Cruveilhier in 1829 [4]. They form between the third and fifth week of embryonic development as a result of displaced epithelial remnants that persists despite neural tube closure [1, 3, 5]. Herein we describe a similar tumor locating in the cervico-medullary region presenting with features of progressive hydrocephalus. Though benign, these tumors insinuate into the surrounding vital structures thereby making their complete excision difficult.

CASE REPORT

A 47-year-old female from Lamjung, Nepal presented to the emergency department of our hospital with a history of on and off headache since last six months and persistent projectile vomiting for last two days. There was no history of trauma, loss of consciousness, aura, weakness of body parts, bladder bowel incontinence or fever with chills and rigor. There was no significant past medical or surgical illnesses. Her Glasgow Coma Scale (GCS) was 15/15. Higher mental function was normal. All cranial nerves were intact. There were no sensory or motor deficits. Cerebellar signs were absent. Fundoscopy revealed bilateral papilledema. Computed tomography (CT) scan of head revealed hypodense and

isodense lesion with anterior nodule pressing upon the fourth ventricle and extending inferiorly up to cervico-medullary junction (Figure 1). There was no contrast enhancement within the lesion. There was also evolving hydrocephalus. Fluid attenuated inverse recovery (FLAIR) and diffusion weighted image (DWI) sequence was performed to differentiate it from the arachnoid cysts. The diagnosis of posterior fossa epidermoid tumor with evolving hydrocephalus was made. Detailed counseling was done and the consent for the surgery was taken. She underwent midline sub-occipital craniectomy with gross excision of tumor. Intraoperatively tumor consisting of large bed of pearls with pseudocapsule adhering to midbrain and surrounding cisterns was seen (Figure 2). The tumor was insinuating basal nerves and extending caudally to C1 arch. Gross removal of the tumor was done (Figure 3). The tumor bed was thoroughly irrigated with normal saline so as to prevent chemical meningitis and recurrence. The dura was repaired and the wound was closed in layers. Postoperatively, she had uneventful recovery. Repeat CT scan showed no contrast enhancement with minimal extra-axial collection and resolving hydrocephalus. The patient was monitored for features of progressive hydrocephalus till her stay at the hospital. The patient was discharged home on the 10th postoperative day on prophylactic antiepileptic (sodium valproate 300 mg per oral three times daily). Patient followed up in the outpatient clinic one month later with a complain of swelling over the surgical site. Examination revealed soft fluctuant swelling suggestive of pseudo-meningocele. Therapeutic tap was done and the patient was started on tab acetazolamide 250 mg per oral three times daily and tapered over three weeks. There was no further recurrence. Patient was advised for six monthly follow-up.

DISCUSSION

These tumors typically occur in the cerebellopontine angle cisterns [1]. Although, few cases are reported in the fourth ventricle, this location is the second most common for an epidermoid tumor in the posterior fossa [6].

Epidermoids histologically consists of an outer capsule surrounding a layer of keratinized stratified squamous epithelium and inner cystic fluid that usually includes debris, keratin, water and cholesterol. As the epithelial layer desquamates, the cells accumulate and form a cholesterol-rich layer that gives the tumor its characteristic bed of pearls appearance. On contrary to the dermoid tumors, these tumors do not contain any dermal appendages. Though benign, these tumors tend

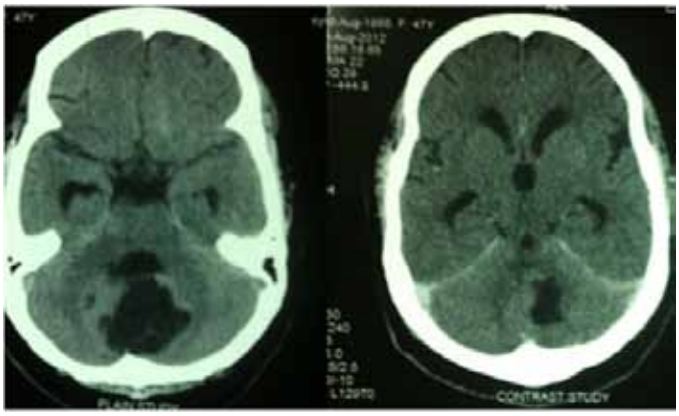


Figure 1: Plain and contrast computed tomography images showing lesion with mixed density in the posterior fossa with evolving hydrocephalus.

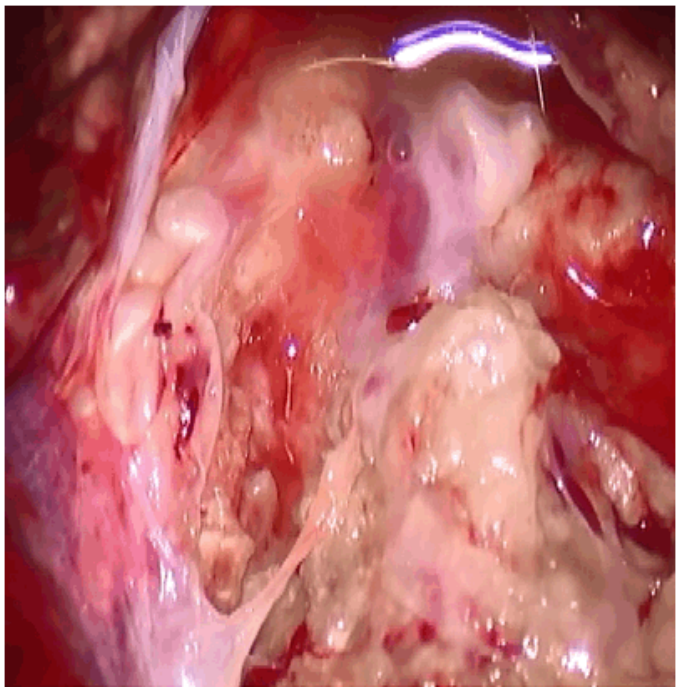


Figure 2: Intraoperative picture showing the bed of pearls appearance of the lesion

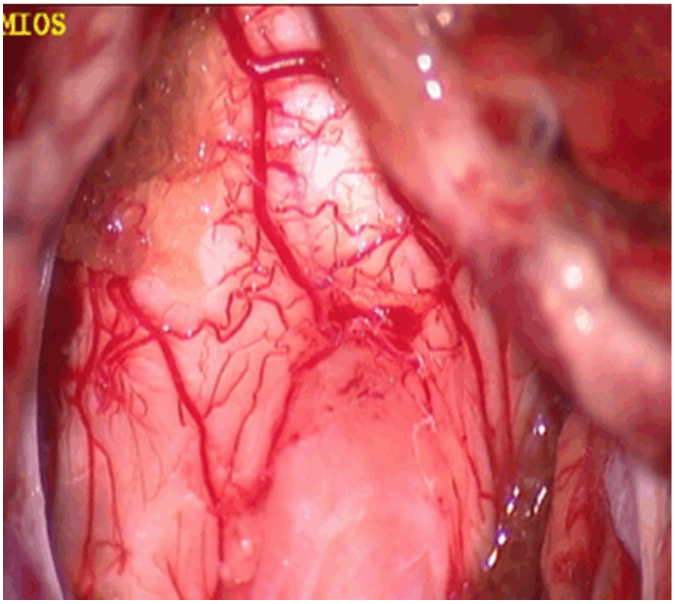


Figure 3: Tumor bed after gross complete removal of the lesion.

to insinuate and encasing adjacent vessels and nerves thereby making its complete removal difficult.

Current armantarium of radio imaging shows cerebrospinal fluid like characteristics but having insinuation to adjacent neurovascular structures [1]. Early distinction from the arachnoid cyst can be easily done by applying the Fluid attenuated inverse recovery (FLAIR) and diffusion weighted image (DWI) sequences. On contrary to arachnoid cysts, it is hyperintense on FLAIR and has restricted diffusion on DW images.

Majority of the patients present with gait problems due to involvement of cerebellar vermis [7]. On contrary to other solid tumors, the incidence of hydrocephalus is rare in such tumors due to egress of the cerebral spinal fluid within the fissures of the tumors to the surrounding foramina [7]. Another presentation may be chemical meningitis due to tumor leakage into the subarachnoid space [6]. The reported rate of recurrence of epidermoid tumors in literature is highly variable. In case of suspected recurrence, main focus should be on the clinical background [6]. While undergoing re-surgery, main attempt should be made removing the areas of abnormal enhancing portion seen on the image studies and sending them for histopathological diagnosis.

CONCLUSION

These tumors though benign in nature, the insinuating nature of the tumor marks the complete removal of the tumor most often cumbersome. Characteristic findings of hyperintensity on FLAIR and restricted diffusion on DWI MR sequences afford easy distinction from arachnoid cyst. Intraoperatively, they have characteristic bed of pearls appearance marking its apparent diagnosis. Postoperative period may be complicated by chemical meningitis, pseudomeningocele and recurrences.

****

Author Contributions

Sunil Munakomi – 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
Binod Bhattarai – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Iype Cherian – 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. Osborn AG, Preece MT. Intracranial cysts: radiologic-pathologic correlation and imaging approach. Radiology 2006 Jun;239(3):650–64.
2. Osborn AG. eds. Diagnostic imaging: Brain. Salt Lake City, Utah: Amirsys; 2004.
3. Kaido T, Okazaki A, Kurokawa S, Tsukamoto M. Pathogenesis of intraparenchymal epidermoid cyst in the brain: a case report and review of the literature. Surg Neurol 2003 Mar;59(3):211–6.
4. Kachhara R, Bhattacharya RN, Radhakrishnan VV. Epidermoid cyst involving the brain stem. Acta Neurochir (Wien) 2000;142(1):97–100.
5. Recinos PF, Roonprapunt C, Jallo GI. Intrinsic brainstem epidermoid cyst. Case report and review of the literature. J Neurosurg 2006 Apr;104(4 Suppl):285–9.
6. Tancredi A, Fiume D, Gazzeri G. Epidermoid cysts of the fourth ventricle: very long follow up in 9 cases and review of the literature. Acta Neurochir (Wien) 2003 Oct;145(10):905–10; discussion 910–1.
7. Nassar SI, Haddad FS, Abdo A. Epidermoid tumors of the fourth ventricle. Surg Neurol 1995 Mar;43(3):246–51.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Pseudoxanthoma elasticum: A case report

Ayman Elgendy, Abdalaziz Altaweel, Eslam Alshawadfy, Eman Ali, Magdy Afify, Morad Abouelela, Karem Khalil, Ahmed M. Elsaidi

ABSTRACT

Introduction: Pseudoxanthoma elasticum (PXE) is a rare genetic disorder that mainly involves the skin, eyes, and cardiovascular system. Case Report: We reported a case of pseudoxanthoma elasticum which involves axillae, neck, abdomen and thighs, together with angioid streaks of fundi, but without cardiovascular events. Skin biopsy specimen was taken and was stained with hematoxylin and eosin (H&E) that revealed clumping and fragmentation of elastic fibers. Conclusion: These features confirmed histopathological diagnosis of PXE.

Keywords: Angioid streaks, Elastic fibers, Pseudoxanthoma elasticum

How to cite this article

Elgendy A, Altaweel A, Alshawadfy E, Ali E, Afify M, Abouelela M, Khalil K, Elsaidi AM. Pseudoxanthoma elasticum: A case report. Int J Case Rep Imag 2016;7(5):336–340.

Ayman Elgendy[1], Abdalaziz Altaweel[1], Eslam Alshawadfy[2], Eman Ali[2], Magdy Afify[3], Morad Abouelela[3], Karem Khalil[1], Ahmed M. Elsaidi[4]

Affiliations: [1]Dermatology Department, Benha University; [2]Dermatology Department, National Research Center; [3]Ophthalmology Department, Saudi Airlines Medical Service; [4]Ain shams University.

Corresponding Author: Ayman Mohamed Elgendi, Dermatology Department, Benha University; Email: aymanelgendy91@yahoo.com

Received: 09 January 2016
Accepted: 13 February 2016
Published: 01 May 2016

Article ID: Z01201605CR10648AE

****

doi:10.5348/ijcri-201660-CR-10648

INTRODUCTION

Pseudoxanthoma elasticum (PXE) is an inherited systemic disease involves elastic tissue of the skin, eyes and cardiovascular system. Skin and ocular manifestations of pseudoxanthoma elasticum are referred to as Grönblad-Strandberg syndrome. The condition was named by Darier in 1896, who sought to differentiate PXE from common xanthomas. Cutaneous changes are usually the first manifestation of pseudoxanthoma elasticum, but do not become recognizable until the second or third decade of life [1].

Pseudoxanthoma elasticum can be transmitted as an autosomal dominant trait (Type I and Type II) or an autosomal recessive trait (Type I and Type II). PXE has been estimated to have a prevalence ranging from 1 in 70,000 to 1 in 1 million [2, 3].

In some families the cutaneous changes may be predominant with relatively mild eye or cardiovascular involvement, while in other families the involvement of eye and cardiovascular system may be severe with limited skin findings [4]. Pathognomonic alterations of some organs may be attributed to certain types of mutations. Polymorphisms of p.R1268Q is associated with early onset of angioid streaks [5, 6], while the stop codon mutation p.R1141X is correlated with to cardiovascular involvement independent of hyperlipidemia [7]. The first lesions to be noted are on the skin in the lateral part of the neck. Skin lesions begin in childhood but they are not usually noted until adolescence. Small, yellow papules are seen in a linear or reticular pattern and may coalesce to form plaques. Clinically,

patients show characteristic ocular manifestations, including peau d'orange, angioid streaks, and choroidal neovascularizations [8]. Intermittent claudication is the most common cardiovascular symptom (30% of patients) and often represents the first sign of atherosclerosis. Slowly progressive calcification of the elastic media and intima of the blood vessels leads to various cardiovascular manifestations [9].

CASE REPORT

A 24–year–old male presented with 10-year history of yellow-orange lesions on sides of the neck, axillae and the thighs.

On clinical examination, it was found that the patient had yellowish papular lesions with cobblestone–like appearance, symmetrically distributed on the sides of the neck (Figure 1), the axillae (Figure 2) and abdomen.

These lesions were asymptomatic and rendered no difficulty to the patient. Fundoscopy revealed bilateral angioid streaking of the fundi (Figure 3).

Hair, nails, mucous membranes, and other systemic examinations were normal. The patient was referred to cardiologists and subjected to blood pressure measurement, an electrocardiogram (ECG) and also an examination of arterial stiffness. Furthermore, the patient has been subjected to an abdominal echography. Patient had no cardiovascular or hemorrhagic events. Complete blood cell count and urine analysis were done, which were within the normal limits. There was no family history. Skin biopsy was taken and sent for histopathological examination. Hematoxylin and eosin (H&E) staining demonstrated clumping, degeneration and fragmentation of elastic fibers in the mid–dermis (Figure 4).

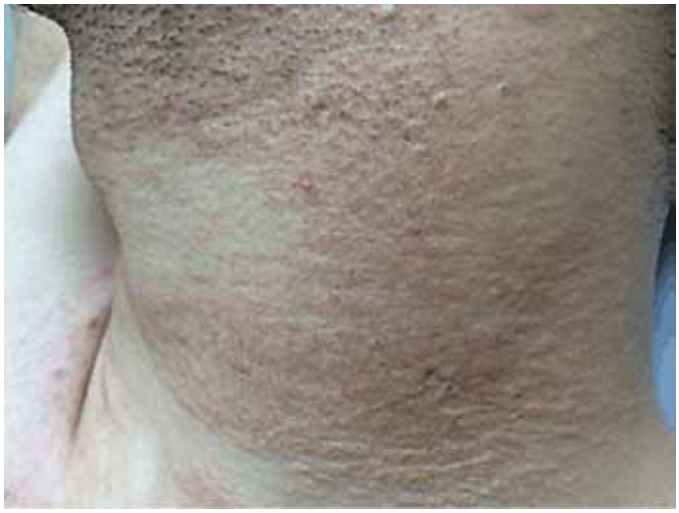


Figure 1: Cobblestone-like yellowish papules, symmetrically distributed on the sides of the neck.

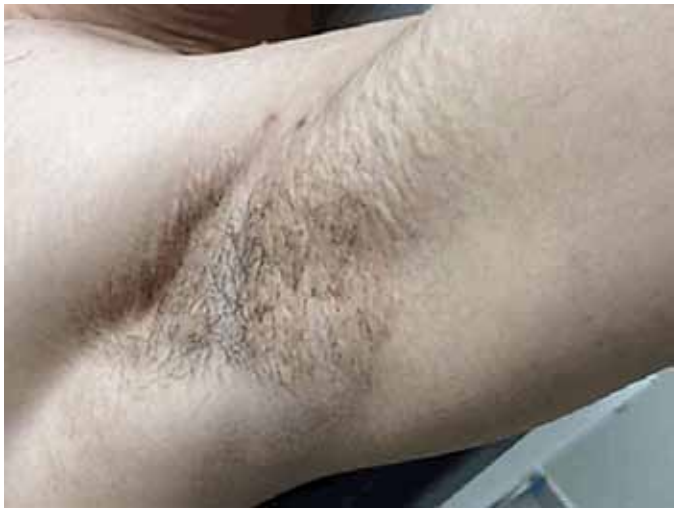


Figure 2: Cobblestone-like yellowish papules and comedones, distributed on the axilla.

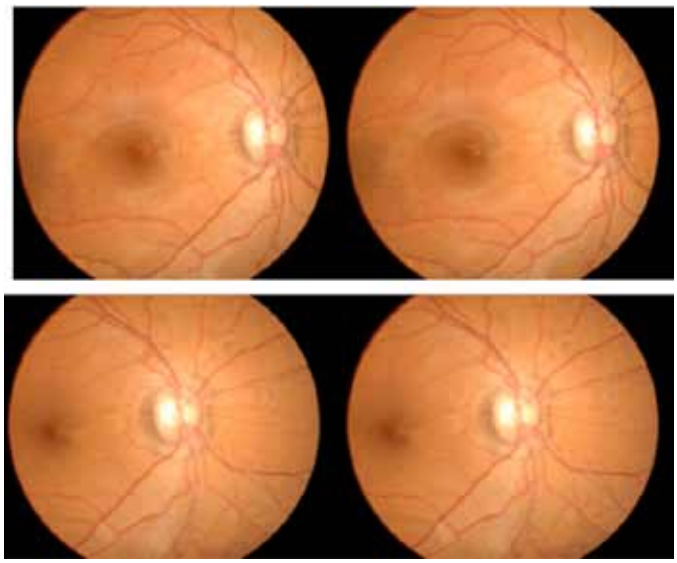


Figure 3 (A, B): Bilateral angioid streaking of the fundi.

DISCUSSION

Pseudoxanthoma elasticum (PXE) is a rare progressive disorder that affects the elastic tissue of the skin, the eyes and blood vessels. It is also known as Gronblad-Strandberg syndrome. The basic fault in PXE appears to relate to The mutations in the transporter genes MRP6 or ABCC6, which has been mapped to chromosome 16p13. Mutations in the ABCC6 gene cause absence or non-functional MRP6 protein which may cause impairment of release of ATP from cells. As a result calcium and other minerals accumulate in elastic fibers of the skin, blood vessels, eyes, and other tissues affected by PXE [9]. There are two types of autosomal dominant pseudoxanthoma elasticum; type I is characterized by a classic skin lesions, intermittent claudication, severe recurrent angina, and severe chorioretinitis, even blindness, and type II is a much milder form, with a macular rash, mild retinal degeneration and no vascular complications [2]. Autosomal recessive pseudoxanthoma elasticum also has two types: recessive type I has the characteristic

flexural distributed skin rash, moderately severe ocular disease, and increased risk to gastrointestinal bleeding, and recessive type II is much rarer and affects the entire skin which is soft, lax and wrinkled. It shows extensive infiltration with degenerated elastic fibers [3].

Cutaneous changes are usually the first manifestation of pseudoxanthoma elasticum. These lesions starts during childhood and progresses sluggishly during adulthood. Small, yellowish papular lesions in a linear or reticular pattern are seen on the neck, axillae, groin, and flexural creases. On advancement of the disease, the skin may become loose, lax and redundant and it hangs down in folds producing the typical plucked chicken appearance [10].

The histology of PXE is characteristic: skin lesions show clumped and fragmented elastic fibers with calcium deposits in the mid and deep reticular dermis. Similar changes occur in elastic fibers of Bruch's membrane of the eye, the blood vessels, endocardium and other organs [11]. Abnormal calcium deposits can be demonstrated with the von Kossa stain in the connective tissue. Initially, mineralization of the elastic fiber has been seen as a central core of electron density on electron microscopy, then core density increases as mineralization continues. Dermal mineralized areas show deposits of thread-like material and collagen fibrils of irregular diameter. Ultrastructurally, extracellular matrix components such as proteoglycans, fibronectin, vitronectin, and have been found to be accumulated in lesional skin. Raised levels of glycosaminoglycans were seen in affected skin and urine of some patients with PXE [12]. It has been found that 87% of patients with pseudoxanthoma elasticum have angioid streaks (AS), seen as irregular radiations from a ring-like area around the disc and extending out into the fundus. Angioid streaks are visible as dark red-to-brown bands and are variable in their pigmentation. Angioid streaks result from crack-like breaks in Bruch's membrane due to its abnormal structural composition, which predisposes to these localized areas of rupture. During fluorescein angiography, angioid streaks reveal increased fluorescence in the early phase resulting from increased visibility of the choroid due to the local defect in Bruch's membrane and in the later phase due to the leakage from the adjacent choriocapillaris. The frequent clinical association of angioid streaks and disciform degeneration at the macular area and the similarity of the histopathological changes indicate some pathogenetic link between them. Macular involvement with loss of vision usually appears after age 40 years. Angioid streaks may progress slowly or remain stationary for years. Two possibilities are suggested to be the cause of angioid streaks: [1] Degenerative changes in Bruch's membrane which are incidental to several other pathological conditions; [2] Primary degeneration of the Bruch's membrane which is bilateral and inherited with elastic tissue degeneration in other tissues of the body. Bruch membrane defects predispose to choroidal neovascularization, which may cause subretinal hemorrhage and ultimately disciform degeneration. Loss visual field due to optic disk drusen has been reported in some patients with pseudoxanthoma elasticum who have angioid streaks. The prognosis is often very poor because of choroidal ruptures and retinal hemorrhages which may occur in patients with pseudoxanthoma elasticum due to minor ocular trauma [13, 14]. Choroidal neovascularization (CNV) can be treated with surgery, photocoagulation, and photodynamic therapy with varying success [15]. Intravitreal antivascular endothelial growth factor (anti-VEGF) agents should be considered for patients with choroidal neovascularization. Intravitreal injection of aflibercept or ranibizumab or the off-label use of bevacizumab seems to maintain visual acuity [15]. Cardiovascular manifestations include calcifications within the elastic tissue of the intima and media of blood vessels leading to intermittent claudication, coronary and cerebrovascular disease [10]. Valvular changes, mainly mitral valve prolapse, may be present. Early PXE-related coronary artery disease is often severe, most cases presenting as early angina pectoris or myocardial infarction. In some cases, coronary artery disease has led to sudden death [16]. Stroke may also occur as the consequence of ischemic or hemorrhagic cerebrovascular disease. Gastrointestinal hemorrhages are often dramatic and recurrent [17]. Acquired form of PXE has been mentioned in many case reports as an PXE with skin and ocular manifestations, similar to the hereditary PXE but without ABCC6 mutations that do not carry any genetic basis. This form of PXE may be associated with other conditions like autoimmune thyroiditis, and congenital anemia like the sickle−cell disease, and cases of spherocytosis. PXE-like skin lesions in combination with ocular and/or vascular symptoms and calcified elastic fibers were detected in some patient with beta-thalassemia. No disease-causing variant was found in the ABCC6 gene, indicating that this was a phenocopy of PXE. All these conditions related to acquired PXE show degeneration and fragmentation of the elastic fibers infiltrated with calcium and produce clinical and

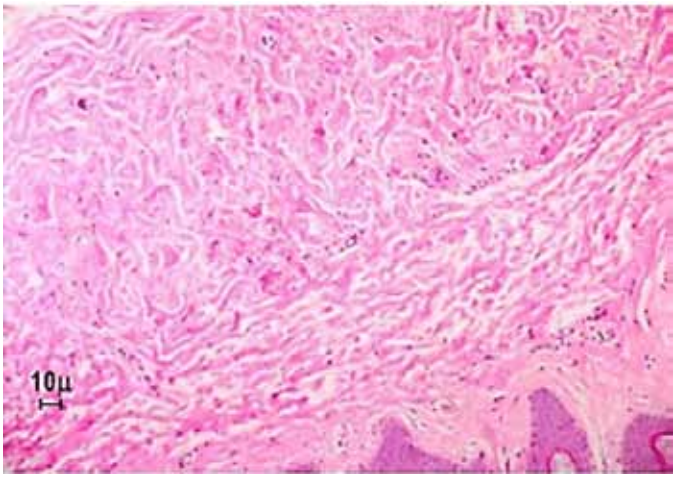


Figure 4: Optical microscope (H&E stain, x100). Elastin fibers are distorted and altered with calcium deposits.

histological changes that become evident. Injury to the elastic fibers in the above conditions may result from the focal, mechanical, and biochemical irritation to the connective tissue inducing a foreign−body reaction and leading to their degeneration [6, 18]. Differential diagnosis includes papillary dermal elastolysis, papular elastorrhexis, perforating periumbilical pseudoxanthoma elasticum, severe actinic damage to the lateral part of the neck long-term, penicillamine therapy and cutis laxa. Severe ocular lesions like those of PXE may occur in some cases. The occurrence angioid streaks in hemochromatosis may be a reflection of iron deposits. Also, angioid streaks were demonstrated in patients with Paget disease of bone and with tumoral calcinosis with hyperphosphatemia due to the deposition of calcium in a relatively normal Bruch membrane [19]. There is no treatment that directly interferes with this multifaceted disorder, although dietary restriction of calcium has been tried with limited results. For excessive areas of skin, plastic surgery may be needed. Avoidance of head trauma and heavy straining is needed to prevent retinal hemorrhage [20].

CONCLUSION

The early diagnosis of pseudoxanthoma elasticum (PXE) may be important to minimize the serious complications and long-term impact on quality of life. This case report may help the clinicians for the early recognition and minimization of the complications of these rare disorders.

****

Author Contributions

Ayman Elgendy – 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
Eslam Alshawadfy – 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
Eman Ali – 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
Magdy Afify – 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
Morad Abouelela – 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
Karem Khalil – 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
Ahmed Elsaidi – Substantial contributions to conception and design, Acquisition of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

REFERENCES

1. Neldner KH. Pseudoxanthoma elasticum. Clin Dermatol 1988 Jan-Mar;6(1):1–159.
2. Pope FM. Autosomal dominant pseudoxanthoma elasticum. J Med Genet 1974 Jun;11(2):152–7.
3. Pope FM. Two types of autosomal recessive pseudoxanthoma elasticum. Arch Dermatol 1974 Aug;110(2):209–12.
4. Pfendner EG, Vanakker OM, Terry SF, et al. Mutation detection in the ABCC6 gene and genotype-phenotype analysis in a large international case series affected by pseudoxanthoma elasticum. J Med Genet 2007 Oct;44(10):621–8.
5. Sato N, Nakayama T, Mizutani Y, Yuzawa M. Novel mutations of ABCC6 gene in Japanese patients with Angioid streaks. Biochem Biophys Res Commun 2009 Mar 13;380(3):548–53.
6. Li Q, Sadowski S, Uitto J. Angioid streaks in Pseudoxanthoma Elasticum: role of the p.R1268Q mutation in the ABCC6 gene. J Invest Dermatol 2011 Mar;131(3):782–5.
7. Köblös G, Andrikovics H, Prohászka Z, Tordai A, Váradi A, Arányi T. The R1141X loss-of-function mutation of the ABCC6 gene is a strong genetic risk factor for coronary artery disease. Genet Test Mol Biomarkers 2010 Feb;14(1):75–8.
8. Sabán-Ruiz J, Fabregate Fuente R, Sánchez-Largo Uceda E, Fabregate Fuente M. Pseudoxanthoma elasticum: case report with arterial stiffness evaluated by a research cardiovascular profiling system. Eur J Dermatol 2010 Nov-Dec;20(6):785–7.
9. Babu RS, Nair IK, Suresh MK, Dalus D. Gronblad Strandberg syndrome with vertibrobasilar dolichoectasia. J Assoc Physicians India 2011 Jan;59:54–7.
10. Uitto J, Shamban A. Heritable skin diseases with molecular defects in collagen or elastin. Dermatol Clin 1987 Jan;5(1):63–84.

11. Miki K, Yuri T, Takeda N, Takehana K, Iwasaka T, Tsubura A. An autopsy case of pseudoxanthoma elasticum: histochemical characteristics. Med Mol Morphol 2007 Sep;40(3):172–7.
12. Maccari F, Volpi N. Structural characterization of the skin glycosaminoglycans in patients with pseudoxanthoma elasticum. Int J Dermatol 2008 Oct;47(10):1024–7.
13. Finger RP, Charbel Issa P, Ladewig MS, et al. Pseudoxanthoma elasticum: genetics, clinical manifestations and therapeutic approaches. Surv Ophthalmol 2009 Mar-Apr;54(2):272–85.
14. Bercovitch L, Terry P. Pseudoxanthoma elasticum 2004. J Am Acad Dermatol 2004 Jul;51(1 Suppl):S13–4.
15. Finger RP, Charbel Issa P, Ladewig M, Holz FG, Scholl HP. Intravitreal bevacizumab for choroidal neovascularisation associated with pseudoxanthoma elasticum. Br J Ophthalmol 2008 Apr;92(4):483–7.
16. Combrinck M, Gilbert JD, Byard RW. Pseudoxanthoma elasticum and sudden death. J Forensic Sci 2011 Mar;56(2):418–22.
17. Schulz V, Hendig D, Schillinger M, et al. Analysis of sequence variations in the ABCC6 gene among patients with abdominal aortic aneurysm and pseudoxanthoma elasticum. J Vasc Res 2005 Sep-Oct;42(5):424–32.
18. Baccarani-Contri M, Vincenzi D, Cicchetti F, Mori G, Pasquali-Ronchetti I. Immunochemical identification of abnormal constituents in the dermis of pseudoxanthoma elasticum patients. Eur J Histochem 1994;38(2):111–23.
19. Lin Y, Chang C, Wang C, Tsai R, Hsu Y. Pseudoxanthoma elasticum: A systemic disorder of elastic tissue diagnosed by skin - A case report. Tzu Chi Med J 2006;18:445–7.
20. Shukla P, Moidin S, Jose M, Shenoy M. Pseudoxanthoma elasticum - "The multifaceted disorder". J Cranio Max Dis 2015;4(1):107–9.

CASE REPORT PEER REVIEWED | OPEN ACCESS

Large cutaneous horn in a young African-American female

Jeremy Bosworth, Ashley Modica, Ikenna Nweze, George Angus

ABSTRACT

Introduction: Cutaneous horns are hyperkeratotic lesions that can be present anywhere on the body. These horns are not themselves malignant. The base of these horns can harbor a malignant histopathological diagnosis and thus special care must be taken to not only excise the horns, but to obtain clear margins while doing so. Case Report: A case of a young African-American female with a large cutaneous horn. Conclusion: All horns must be presumed malignant until a pathological diagnosis proves otherwise. While there are general risk factors for the malignant potential of these horns, as reported in other cases, there are no consistent predictors to determine which horns will be malignant.

Keywords: Cutaneous, Horn, Malignancy, Verruca vulgaris

How to cite this article

Bosworth J, Modica A, Nweze I, Angus G. Large cutaneous horn in a young African-American female. Int J Case Rep Imag 2016;7(5):341–345.

Jeremy Bosworth[1], Ashley Modica[1], Ikenna Nweze[1], George Angus[1]

Affiliations: [1]MD, Nassau University Medical Center, Department of Surgery, 2201 Hempstead Turnpike, East Meadow, NY 11554, USA.

Corresponding Author: George Angus L.D MD, Nassau University Medical Center, Department of Surgery, 2201 Hempstead Turnpike, East Meadow, NY 11554, USA; Email: langus@numc.edu

Received: 10 December 2015
Accepted: 16 January 2016
Published: 01 May 2016

Article ID: Z01201605CR10649JB

****

doi:10.5348/ijcri-201661-CR-10649

INTRODUCTION

Cutaneous horns are hyperkeratotic lesions that can be present anywhere on the body; these horns are not themselves malignant. However, they may be harboring malignant cells at their base [1–3]. There is a large variation in the appearance of these cutaneous horns and though some features are more likely to define a hidden malignancy these are not hard and fast rules [1–5]. Studies have shown that some of the most important risk factors for horns concealing malignancy include horns present in sun exposed areas and those with tenderness at the base [1, 2, 4–6]. Cutaneous horns are rare and that owes to the difficulty of having an exact epidemiologic description and treatment plan [2, 3]. It is widely accepted across literature that surgical excision of cutaneous horns is warranted with at least a shave margin of the base to achieve a histologic diagnosis in the area where malignancy is most likely to be located [4, 5]. The recurrence rate of cutaneous horns after excision is not well documented in literature; Mical et al. were able to present multiple cases of giant cutaneous horns (17–25 cm) that were excised without malignancy and were followed from 2–15 years without any evidence of recurrence [7]. Clinicians must be aware that there is not a definite connection between features of a cutaneous horn and their individual tendencies to be malignant therefore every horn encountered must be treated as potentially malignant [1].

CASE REPORT

A 28-year-old African-American female with past medical history significant only for bipolar disease and recent childbirth, presented to the general surgery clinic with a two-year history of a growth on her frontal scalp. She reported the mass grown over a two-year period, became painful, and occasionally bled from the base. She denied any other associated symptoms. Prior to two years ago, she reported no similar issues, no associated family history, and reported family was all from America.

Physical Examination large, spiral, horn-shaped mass protruding from frontal scalp. Length of the mass was approximately 22 cm total length and 3 cm in diameter at the base. Hard and smooth in texture with the feeling similar to a thick finger nail. No active bleeding noted. There was some scaling around the base. However, it was mobile with the scalp and did not appear fixed to the skull (Figure 1).

On follow-up examination, imaging studies consisting of a skull X-ray and CT scan were reviewed (Figure 2 and Figure 3) and the patient was scheduled for excision of the mass. Plastic surgery was consulted in the event a skin graft was necessary.

Patient was taken to the operating room and the mass was excised down to galea with 5 mm margin at the periphery of the base of the horn (Figure 4). The scalp was further mobilized to create skin flaps allowing approximation of wound edges. The wound was closed primarily. The patient was extubated and subsequently discharged with post-surgical follow-up instructions.

On initial follow-up appointment the patient reported doing well with minimal pain. The wound was noted to be healing well with no signs of dehiscence or infection (Figure 5). The pathologic report returned the description of a 5.0x5.0 cm hair bearing skin base in continuity with a 21.0x4.0x4.0 cm smooth, firm, yellow-white serpiginous structure with a diagnosis of *Verruca vulgaris* completely

Figure 1: Cutaneous horn on initial physical examination.

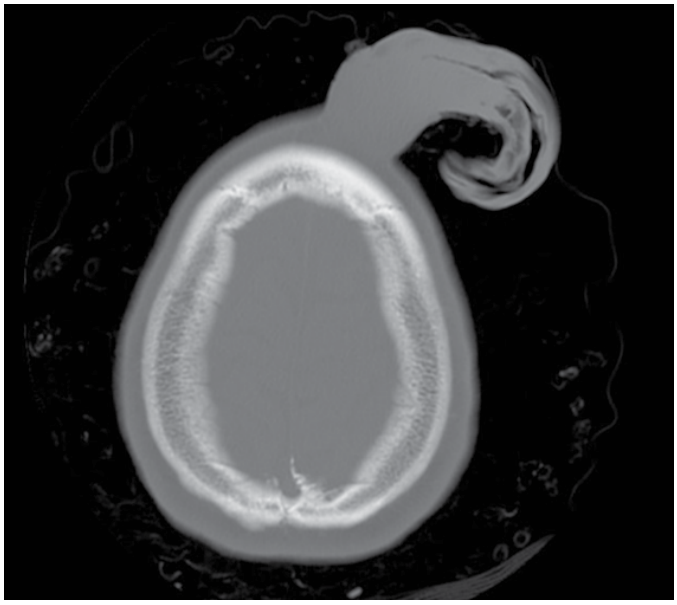


Figure 2: Lesion on computed tomography scan. No bony attachment was seen.

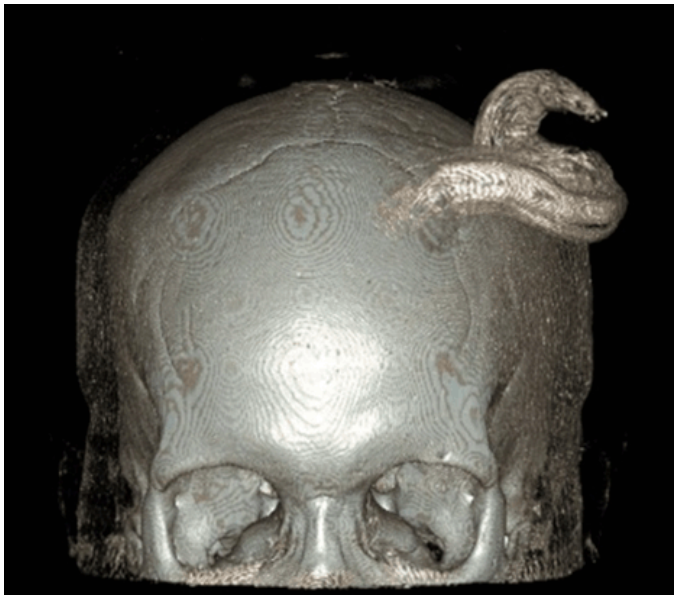


Figure 3: Computed tomography scan, 3-D reconstruction.

Figure 4: Excised specimen showing 5 mm margin.

Figure 5: Postoperative surgical scar.

excised. No evidence of malignancy. The remainder of the patient's postoperative course was uneventful.

DISCUSSION

Cutaneous horn is a clinical diagnosis referring to a conical protrusion from the skin that is comprised mostly of compacted keratin [1, 2]. The mechanism of hyperkeratosis and compaction of the keratin is unknown [2, 3]. Cutaneous horns have a similar appearance to animal horns; however they lack the presence of a central bone [3, 4, 8]. These horns can come in different sizes, shapes and multiplicities. The histopathological area of importance is the base of the horn where the diagnosis of an underlying benign, pre-malignant, or malignant lesion can be made [3, 8].

There are no consistently defined distinguishing features between benign or malignant lesions in cutaneous horns. However, there are some observations that have been noted. Malignancy is more likely in the setting of horns that are tender at the base, those found on older male patients, in horns with wide bases or small height to base ratios, and in those patients whose horn is located on a sun-exposed area of their body [1, 2, 4, 5, 8]. Additionally noted, less reliable risk factors favoring malignancy include hardness at the base, the length of time a horn has been growing and a history of trauma to the area of growth [2]. Size by itself is not an indication of malignancy [2, 4]. Given these observations, our patient, a young female was at lower risk for malignancy, and although she did have some pain and bleeding at the base, these symptoms are non-specific. The pain at the base of the horn is noted as a malignancy risk factor [4, 5], however the size of our patient's horn was so large that it is possible that the weight of the horn was actually causing her pain and discomfort.

Sun exposure is believed by many to be the most important risk factor for cutaneous horn formation. Indeed, the incidence of horn formation is greater in lighter skinned patients and rare in African–Americans [5, 6]. The most common histopathological diagnoses noted at the hyper-proliferative base include actinic keratosis and squamous cell carcinoma, and about 30% of the overall known cases of cutaneous horns have been found on the upper face and scalp [4, 5]. Given the rarity of cutaneous horns, there is no clear consensus on different epidemiological factors [2,3]; however, retrospective case reviews have been done to evaluate trends with respect to these horns. In a retrospective study of 222 cases of cutaneous horns in Brazil, it was noted that these horns were more common in females [65%], more common in patients age greater than 50, and the head and the upper limbs were the two most common locations [35% and 31%, respectively]. In terms of histopathological diagnosis, 41% were benign and 58% were either pre-malignant [83% actinic keratosis] or malignant [93% squamous cell carcinoma] [1]. The most common benign diagnoses at the base of the horns studied in Brazil were warts and keratoacanthoma [5]. Other benign findings identified at the base of these horns included seborrheic keratosis, thrichilemmoma, molluscum contagiosum and benign epithelial hyperplasia [1, 5]. Quite often, the process of hyperkeratosis needed to form a cutaneous horn develops over the area of a hyper-proliferative lesion [1]. In the Brazilian study, 80% of the 222 cases reviewed were white subjects and only 15% were "non-white." In a literature review run by Gomes et al. on the major electronic databases, there are very few reports of cutaneous horns, and of those cases there are only a small number of cases reported on darker skinned people [5]. Other reviews have also demonstrated that cutaneous horns are very rare in the African population [6]. It has been reported that these horns can form in an area of repeated trauma such as reported by Gomes et al. Gomes presented a case where a woman had a history of repeated trauma by scratching with her fingernails to an area on her scalp where her giant cutaneous horn grew from. This repetitive trauma was noted as a possible reason for progression of the horn to squamous cell carcinoma [5]. In terms of treatment and removal of these cutaneous horns, it is important to obtain a tissue diagnosis from the base of the horn. There is little literature regarding specific margins and work-up for lesions, however some consistent guidelines have been identified. A shave excision is advised in removal of these horns to obtain adequate tissue sampling and free margins [4, 5]. A 3-mm area of margin is suggested especially in the face where the incidence of malignant lesions is more prominent [2]. Lymph nodes should also be examined and any suspicious nodes should be biopsied [5].

CONCLUSION

Cutaneous horns are rare but can be visually striking as demonstrated by our case. The presence of these horns has significant clinical implications and as such, physicians are to be reminded of their malignant potential. The pathology from the base of the lesion must be obtained to adequately treat the patient and to plan further surveillance after removal. Our patient had a benign growth at the base of her cutaneous horn and has been without recurrence.

****

Author Contributions

Jeremy Bosworth – 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
Ashley Modica – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Ikenna Nweze – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
George Angus – 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. Mantese SA, Diogo PM, Rocha A, Berbert AL, Ferreira AK, Ferreira TC. Cutaneous horn: a retrospective histopathological study of 222 cases. An Bras Dermatol 2010 Mar-Apr;85(2):157–63.
2. Popadic M. Squamous cell carcinoma presenting as a giant cutaneous horn of the lower lip. Indian J Dermatol Venereol Leprol 2014 Jan-Feb;80(1):74–6.
3. Bhangale DI, Patel M, Shah KB. Cutaneous Horn of Scalp: A Rare Entity. International Journal of Neruo and Spinal Sciences 2013;1:1.
4. Fernandes NF, Sinha S, Lambert WC, Schwartz RA. Cutaneous horn: a potentially malignant entity. Acta Dermatovenerol Alp Pannonica Adriat 2009 Dec;18(4):189–93.
5. Gomes CA, Nogueira Castanõn MCM, Gomes CC, et al. Giant Cutaneous Horn in Afro-Brazilian Descendent Patient: Case Report and Literature Review. J Clin Exp Dermatol Res 2011;2:8.
6. Oludiran OO, Ekanem VJ. Cutaneous horns in an african population. J Cutan Aesthet Surg 2011 Sep;4(3):197–200.
7. Michal M, Bisceglia M, Di Mattia A, Gigantic cutaneous horns of the scalp: lesions with a gross similarity to the horns of animals: a report of four cases. Am J Surg Pathol 2002 Jun;26(6):789–94.
8. Copcu E, Sivrioglu N, Culhaci N. Cutaneous horns: are these lesions as innocent as they seem to be? World J Surg Oncol 2004 Jun 3;2:18.

ABOUT THE AUTHORS

Article citation: Bosworth J, Modica A, Nweze I, Angus G. Large cutaneous horn in a young African-American female. Int J Case Rep Imag 2016;7(5):341–345.

Jeremy Bosworth is currently a PGY-2 surgical resident at the Nassau University Medical Center, East Meadow, New York. He earned his undergraduate degree (BS) in Biology from the University of Arizona and his MD degree from St George's University, Grenada

Ashley Modica is a PGY-3 General surgery resident at Nassau University medical Center, East Meadow , New Yok. She did her undergraduate studies at the State University of New York at Geneseo where she obtained a Bachelor's of Science degree in Biology. She subsequently attended the American University of the Carribean where she obtained her MD degree.

Ikenna Nweze is currently a PGY-3 General Surgery Resident at Nassau University Medical Center, East Meadow, New York, USA. He is a native of Canada and attended Medical School at the University of Jos, Nigeria where he obtained a Bachelor of Medicine and Bachelor of Surgery (MBBS). He has published over 10 articles during his surgical training, and has presented at various regional and national surgical conferences in the United States. His research interest lies in understanding molecular pathways involved in sepsis and shock in severely injured patients. Recently, he has been studying trauma care and substance abuse in the underserved population and he plans to continue being involved in various research studies in the future. He intends to pursue an academic carreer and continue to study pathways in sepsis and shock in future.

George Angus is Director of Trauma and Vice-Chair of surgery at Nassau University Medical Center, East Meadow, New York. He is also an Associate Professor of Clinical Surgery in SUNY @ StonyBrook. He earned a BS degree in biology from Fordham University, Bronx, New York and an MD degree from the Albert Einstein College of Medicine, Bronx, New York. He also received a Master in Public Health form Columbia University, New York, New York. He has published over 65 research papers in national and international academic journals. His research interests include trauma care and management, critical care and acute care surgery and more recently the historical aspects of surgery worldwide. He is also interested in Bariatric surgery and created the first multidisciplinary surgical weight reduction center on Long Island, New York. E-mail: langus@numc.edu

CASE REPORT PEER REVIEWED | OPEN ACCESS

Respiratory epithelial adenomatoid hamartoma of the maxillary sinus: A case report

Gerry Raymond Joviolo, Kartono Sudarman, Agus Surono

ABSTRACT

Introduction: Hamartoma is non-neoplastic tumor characterized by excessive proliferation of tissue in parts of the human body. Hamartoma is a very rare in head and neck regions. Case Report: A case of 33-year-old woman who was initially diagnosed as a chronic sinus inflammation. After Caldwell-Luc approach for surgery, histopathological examination showed respiratory epithelial adenomatoid hamartoma (REAH) of the left maxillary sinus. It was an atypical localization because the most common site for REAH was in the nasal cavity. Conclusion: Even though REAH is very rare, otolaryngologist should be aware of the pathologic entity of this disease to differentiate REAH with inverted papilloma, adenocarcinoma or other paranasal sinuses inflammation. Misinterpretation of REAH as a chronic sinus inflammation may lead to inadequate treatment.

Keywords: Neoplasm, Paranasal sinus, Respiratory epithelial adenomatoid hamartoma, Surgery

How to cite this article

Joviolo GR, Sudarman K, Surono A. Respiratory epithelial adenomatoid hamartoma of the maxillary sinus: A case report. Int J Case Rep Imag 2016;7(5):346–349.

Gerry Raymond Joviolo[1], Kartono Sudarman[1], Agus Surono[1]

Affiliations: [1]Department of Otorhinolaryngology-Head and Neck Surgery, Faculty of Medicine, Universitas Gadjah Mada, Sekip Utara, Yogyakarta, Indonesia.

Corresponding Author: Agus Surono MD ORL-HNS PhD, Department of Otorhinolaryngology-Head and Neck Surgery, Faculty of Medicine, Universitas Gadjah Mada, Sekip Utara, Yogyakarta, Indonesia; Email: agus_srn@yahoo.com

Received: 20 December 2015
Accepted: 27 February 2016
Published: 01 May 2016

Article ID: Z01201605CR10650GJ

****

doi:10.5348/ijcri-201662-CR-10650

INTRODUCTION

Hamartoma term was firstly introduced by Albrecht [1] as the primary non-neoplastic malformations of tissue composed of excessive proliferation of cells and tissues indigenous to particular site of the body. Hamartoma may occur in any part of the body such as the surface of epithelium, seromucous glands, fibrous stroma, and vessels [2]. They are common in the spleen, lung, liver, and kidney and intestinal, but rarely found in the head and neck regions [3].

Hamartoma of the head and neck was initially described by Wenig and Heffner [4], a subgroup of hamartoma more often involving upper aerodigestive tract, which was the respiratory epithelial adenomatoid hamartoma (REAH). They found 31 cases with lesion occurred in the nasal cavity, paranasal sinuses, and nasopharynx associated with tumor originating from surface epithelium with proliferation and accumulation of glands and ducts, lined by ciliated respiratory epithelium, and surrounded by edematous or inflammatory background and sometimes invagination of the respiratory epithelium is observed. The proliferation is not derived from seromucous glands that usually found in the region [3–5]. The pathogenesis of REAH is still speculative. Congenital and prolonged inflammatory are often considered to be predisposing factors. The REAH mostly affect men with male to female ratio of 7:1, ranging from third to ninth decades of life with a median age in the sixth decade of life [3, 6].

CASE REPORT

A 33-year-old female presented with more than five months history of nasal blockage with posterior

rhinorrhea. The patient also complained toothache since five years ago. The patient had taken antibiotic treatment but the symptoms were still present. There was no other significant medical history. The physical examination demonstrated swollen of left inferior turbinate, posterior rhinorrhea and upper left 1st molar caries without any mass found. Computed tomography (CT) scan revealed hypodense mass at the left maxillary sinus suggesting a mucocele of the left maxillary sinus (Figure 1). The patient underwent Caldwell-Luc approach of surgery under general anesthesia to evacuate the presence of mass in the left maxillary sinus. Mass of the left maxillary sinus was shown like a sac filled with serous fluid.

Histopathological examination revealed that the lesion was composed of tumor with glandular proliferation covered with pseudostratified ciliated respiratory epithelial cells. The lesion surface was lined in direct continuity with the ciliated respiratory epithelium creating a papillary appearance with elongated invagination to the submucosa. Other abnormal features including hyalinization with eosinophilic basement membrane covering the glands, stromal edema with chronic inflammatory proliferation was also noted (Figure 2). The patient was well recoved and without recurrence of symptoms at first year follow-up.

DISCUSSION

Hamartoma has been described as a mass that developed with abnormal tissue growth. Unlike neoplasms, hamartoma has no capacity to grow continuously, therefore their proliferation is self-limiting. Respiratory epithelial adenomatoid hamartoma (REAH) is a rare type of hamartoma and found in the nasal cavity, the paranasal sinuses and nasopharynx. Wenig and Heffner [4] performed the largest study of REAH in 1995 by identifying 31 cases from Tumor Registry at the Armed Forces Institute of Pathology which consisted of 27 men and 4 women, aged from 27–81 years old, with mean age of 58 years, and the youngest reported was nine years old. In this case report, the patient was woman with age of 33 years old showing an incidence of REAH at the 4th decade of life.

The REAH has been commonly identified at the nasal cavity (70%), most often at the posterior nasal septum. The other sites are the ethmoid and frontal sinuses, nasopharynx and rarely occurred in the maxillary sinus. Di Carlo et al. [6] reported 15 cases of REAH found in the anterior half of the olfactory clefts bilaterally. Only limited literatures of REAH were published, and mostly were case reports. The previous reports of REAH have described at least ± 60 cases confirming rare entity of this disease. This makes a challenge for this lesion not to be misdiagnosis as malignant because it can be treated with simple excision rather than radical surgery. In this case, REAH developed in the left maxillary sinus without any mass in the nasal cavity. Only three cases of REAH in the maxillary sinus had been published in literature.

The presenting symptoms of REAH have been shown as nasal obstruction, nasal stuffiness, rhinorrhea, epistaxis, anosmia/hyposmia and chronic sinusitis [3]. Kessler [7] reported a case of maxillary sinus of REAH presenting as a periapical radiolucency of the first molar without any sinonasal symptom. Symptoms usually occur for a few months up to eight years. Our case showed the symptoms of nasal blockage and posterior rhinorrhea. Opacification of the affected sinus and some connection to the nasal septum are the most common finding of REAH radiologically. The lesion tends to grow slowly and be able to cause bone expansion rather than bone erosion. Lima et al. [8] found that REAH significantly enlarged the width of the olfactory cleft.

Pathological examination showed that REAH tends to have a polypoid appearance, fleshy to firm, dark brown to white masses with varying size and area of induration

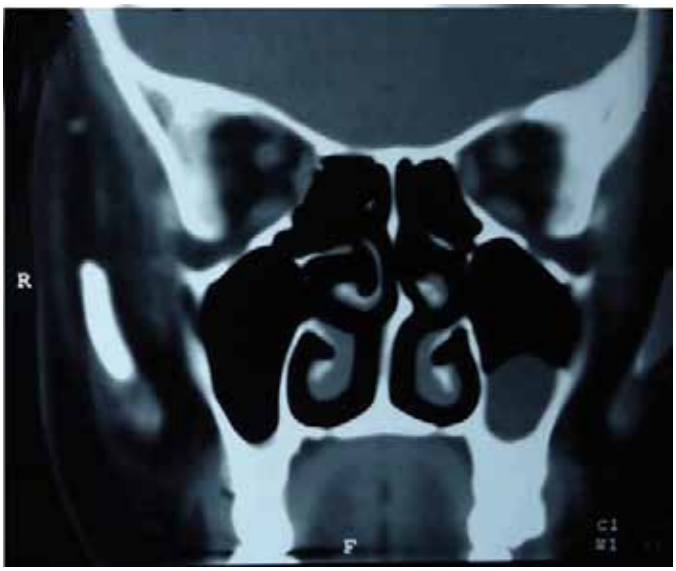


Figure 1: Coronal view of computed tomography scan showed the mass in left maxillary sinus.

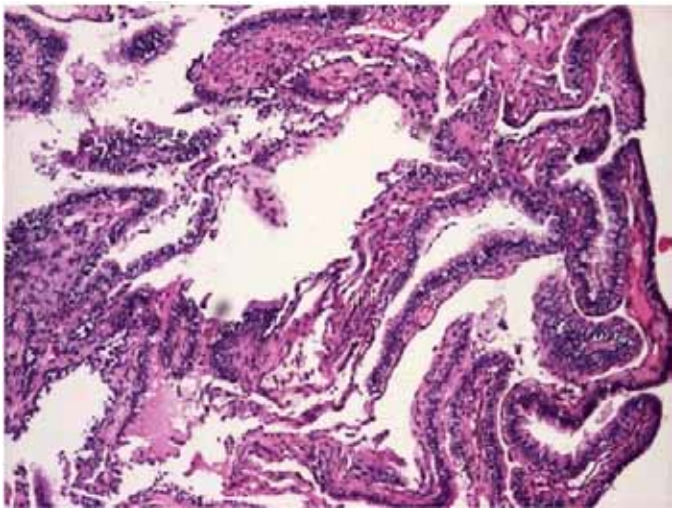


Figure 2: Microscopic appearance of respiratory epithelial adenomatoid hamartoma (REAH). The tissue showed glandular proliferation with invagination of surface epithelium to the submucosa.

noted when the tumor is cut. The histologic features are dominated by the presence of glandular proliferation with the gland covered by the ciliated respiratory epithelium originating from the surface respiratory epithelium with proliferation tends to be submucosal. Other features are like stromal hyalinization and thickening of basement membrane, stromal edema, seromucous gland proliferation, chronic inflammatory cell, and no dysplastic or neoplastic changes seen [9, 10].

The pathogenesis of REAH is unknown. As chronic inflammatory cell often found in REAH histologic view, inflammation is hypothesized may induced REAH [3]. In this case, we found the patient who had teeth caries of upper left 1st molar, could be considered as a risk factor for sinusitis. Prolonged inflammation of the maxillary sinus may develop into REAH [3, 6]. It is a challenge for otolaryngologist to make a distinction of REAH with inflammatory disorders in maxillary sinus because small number of reported cases of REAH in maxillary sinus. The treatment of REAH is complete local excision and there has not been any report of recurrence or progressivity of the disease in literature. Respiratory epithelial adenomatoid hamartoma (REAH) in paranasal sinuses have often been differentially diagnosed with inverted papilloma, adenocarcinoma and nasal polyp [4]. In this case, CT examination suggested mucocele of maxillary sinus prior to pathologic result.

The REAH can be differentiated with nasal polyp from the gross examination of indurations and histological findings of extensive glandular proliferation and stromal hyalinization in REAH. These findings cannot be found in nasal polyp. When differentiating of inverted papilloma with REAH, examination should carefully be performed. The treatments between both diseases are slightly different. The REAH needs local excision while inverted papilloma needs more extensive surgical excision. Inverted papilloma clinically has the ability to destroy bone and invade adjacent vital structures. Histologically, epithelial proliferation with marked thickening, intra epithelial mucous cysts and the presence of inflammatory cells in the epithelium are specific finding of inverted papilloma. The REAH tends to be a single cell layer ciliated respiratory epithelium and inverted papilloma lined by multiple layers of squamous, ciliated, columnar or transitional epithelium. The REAH is also difficult to be differentiated against low grade adenocarcinoma. Stroma identified between the ciliated glands in the case of REAH is the best way to differentiate it from low grade adenocarcinoma. Immunohistochemistry examination using antibody MIB-1 (KI-67) shows higher immunoreactivity of adenocarcinoma compared to hamartoma [3, 9, 11].

CONCLUSION

A case of respiratory epithelial adenomatoid hamartoma (REAH) has been reported. Despite REAH is very rare, it is important for otolaryngologist to be aware of this pathological entity and to differentiate REAH with inverted papilloma or adenocarcinoma in order to avoid unnecessary aggressive surgery. Misinterpretation of REAH as any sinus inflammatory may result in inadequate treatment.

****

Acknowledgements

We would like to thank to staffs of Department of Pathological Anatomi Faculty of Medicine for preparing the result of histological examination.

Author Contributions

Gerry Raymond Joviolo – 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
Kartono Sudarman – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Agus Surono – 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. Albrecht E. Uber hamartoma. Ver Dtsch Ges Pathol 1904;7:153–7.
2. Endo R, Matsuda H, Takahashi M, Hara M, Inaba H, Tsukuda M. Respiratory epithelial adenomatoid hamartoma in the nasal cavity. Acta Otolaryngol 2002 Jun;122(4):398–400.
3. Avilés Jurado FX, Guilemany Toste JM, Alobid I, Alós L, Mullol I Miret J. The importance of the differential diagnosis in rhinology: respiratory epithelial adenomatoid hamartoma of the sinonasal tract. Acta Otorrinolaringol Esp 2012 Jan-Feb;63(1):55–61.
4. Wenig BM, Heffner DK. Respiratory epithelial adenomatoid hamartomas of the sinonasal tract and nasopharynx: a clinicopathologic study of 31 cases. Ann Otol Rhinol Laryngol 1995 Aug;104(8):639–45.

5. Seol JG, Livolsi VA, O'Malley BW Jr, Chen JY, Loevner LA. Respiratory epithelial adenomatoid hamartoma of the bilateral olfactory recesses: a neoplastic mimic? AJNR Am J Neuroradiol 2010 Feb;31(2):277–9.
6. Di Carlo R, Rinaldi R, Ottaviano G, Pastore A. Respiratory epithelial adenomatoid hamartoma of the maxillary sinus: case report. Acta Otorhinolaryngol Ital 2006 Aug;26(4):225–7.
7. Kessler HP, Unterman B. Respiratory epithelial adenomatoid hamartoma of the maxillary sinus presenting as a periapical radiolucency: a case report and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2004 May;97(5):607–12.
8. Lima NB, Jankowski R, Georgel T, Grignon B, Guillemin F, Vignaud JM. Respiratory adenomatoid hamartoma must be suspected on CT-scan enlargement of the olfactory clefts. Rhinology 2006 Dec;44(4):264–9.
9. Fitzhugh VA, Mirani N. Respiratory epithelial adenomatoid hamartoma: a review. Head Neck Pathol 2008 Sep;2(3):203–8.
10. Punia RPS, Mundi I, Dass A, Mohan H. Respiratory epithelial adenomatoid hamartoma of the nasal cavity: A case report. Surg science 2012;3:165–7.
11. Sarioglu S. Update on inverted epithelial lesions of the sinonasal and nasopharyngeal regions. Head Neck Pathol 2007 Sep;1(1):44–9.

ABOUT THE AUTHORS

Article citation: Joviolo GR, Sudarman K, Surono A. Respiratory epithelial adenomatoid hamartoma of the maxillary sinus: A case report. Int J Case Rep Imag 2016;7(5):346–349.

Gerry Raymond Joviolo is Faculty of Medicine in Department of Otorhinolaryngology -Head and Neck Surgery, Universitas Gadjah Mada, Sekip Utara, Yogyakarta, Indonesia.

Kartono Sudarman is Faculty of Medicine in Department of Otorhinolaryngology -Head and Neck Surgery, Universitas Gadjah Mada, Sekip Utara, Yogyakarta, Indonesia.

Agus Surono is Faculty of Medicine in Department of Faculty of Medicine -Head and Neck Surgery, Universitas Gadjah Mada, Sekip Utara, Yogyakarta, Indonesia. His area of interest include head and neck cancer and bronchoesophagology.
E-mail: agus_srn@yahoo.com

CASE REPORT PEER REVIEWED | OPEN ACCESS

Sebaceous cell carcinoma of axilla: A rare case report

Aftab Shaikh, Rajesh Chincholkar, Samarth Agarwal, Aman Singh, Arshad Khan, Dhiraj Patil

ABSTRACT

Introduction: Sebaceous carcinoma is an uncommon and aggressive malignant cutaneous tumor. This neoplasm is thought to arise from sebaceous glandsin the skin and, therefore, may originate anywhere in the body where these glands are found. This region is a common site of origin because the periocular region is rich in this type of gland. However, axilla has rich sebaceous gland as well which may give rise to carcinoma. Case Report: We hereby present a case of a 55-year-old female with sebaceous cell carcinoma of the axilla. Conclusion: Though extra ocular sebaceous cell carcinoma is rare aggressive tumor.

Keywords: Axilla, Malignant cutaneous, Sebaceous cell carcinoma, Tumor

How to cite this article

Shaikh A, Chincholkar R, Agarwal S, Singh A, Khan A, Patil D. Sebaceous cell carcinoma of axilla: A rare case report. Int J Case Rep Images 2016;7(5):350–353.

Article ID: Z01201606CR10651AS

****

Aftab Shaikh[1], Rajesh Chincholkar[1], Samarth Agarwal[1], Aman Singh[1], Arshad Khan[1], Dhiraj Patil[1]

Affiliations: [1]Department of General Surgery, Grant Government Medical College and Sir JJ , Group of Hospitals, Byculla, Mumbai.

Corresponding Author: Dr. Samarth Agarwal, Room – 637, 300 Resident Quarters, JJ Hospital Campus, Byculla, Mumbai, Maharashtra, India; Email: rebellite@gmail.com

Received: 07 October 2015
Accepted: 28 January 2016
Published: 01 May 2016

doi:10.5348/ijcri-201663-CR-10651

INTRODUCTION

Sebaceous carcinoma is an uncommon and an aggressive malignant cutaneous tumor [1]. This neoplasm is thought to arise from sebaceous glands in the skin and, therefore, may originate anywhere in the body where these glands are found. This region is a common site of origin [2, 3] because the periocular region is rich in this type of gland. However, axilla has rich sebaceous gland as well which may give rise to carcinoma. We hereby present a case of sebaceous cell carcinoma of the axilla.

Sebaceous cell carcinoma is a disease of 6th and 7th decade of life and occurs more in women than men [4]. Sebaceous cell carcinoma form less than 1% of all cutaneous malignancies. Extra-ocular sebaceous carcinoma is rare comprising only 25% of all reported cases of sebaceous cell carcinoma. Overall only few cases of sebaceous carcinoma are reported in literature [5].

CASE REPORT

A 55-year-old female presented with ulceroproliferative growth over right axilla since two months. The swelling was small to begin with but then rapidly increased in size.

Since last 15 days swelling developed ulceration and a foul smelling discharge. On physical examination there was an ulceroproliferative lesion in right axilla of size 15x12 cm with presence of maggots. There was no regional lymphadenopathy (Figure 1).

Investigations were within normal limits with no evidence of metastasis which was ruled out clinically and radiologically. Edge biopsy confirmed malignant tumor suggestive of adenocarcinoma. Patient underwent wide excision with local axillary clearance followed by daily cleaning and dressing.

Histopathology on gross examination was suggestive of 12x12 cm size mass. Cut surface shows yellowish-

white tumor with areas of hemorrhage and necrosis. On microscopic examination the stratified squamous epithelium with underlying tissue showed pleomorphic, multi-vacuolated highly atypical cells arranged in rounded nests around the glands (Figures 2 and 3).

Five lymph nodes were examined. They showed inflammatory changes with no evidence of metastasis. Skin grafting was done (Figure 4) after six weeks of cleaning and dressing. The patient was followed up for a period of 12 months which was uneventful.

DISCUSSION

Sebaceous cell carcinoma is a rare but aggressive cutaneous tumor. It was first described by Allaire in 1891 [6]. This tumor arises from sebaceous glands in the skin and approximately 75% of these tumors arise from periocular region [7, 8]. Incidence among females is more as compared to males, 57–77% of patients being females [9, 10]. However, incidence of extraocular sebaceous cell carcinoma is higher among males.

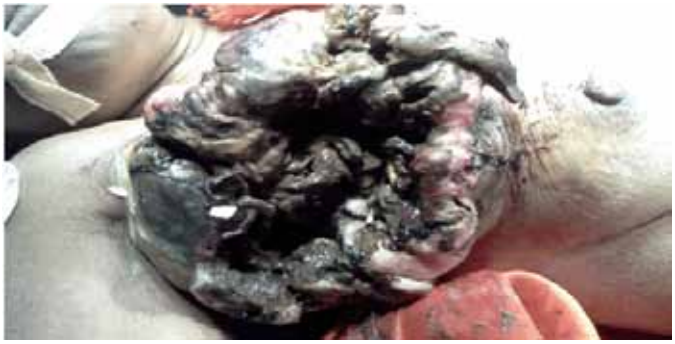


Figure 1: Ulceroproliferative growth over right axillary region with maggots.

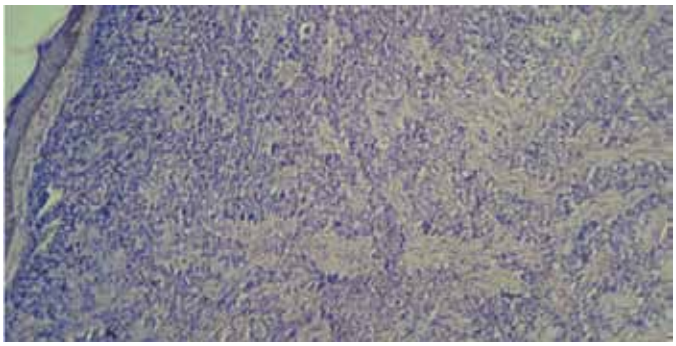


Figure 2: Stratified squamous epithelium with underlying tissue showing pleomorphic multivacuolated highly atypical cells.

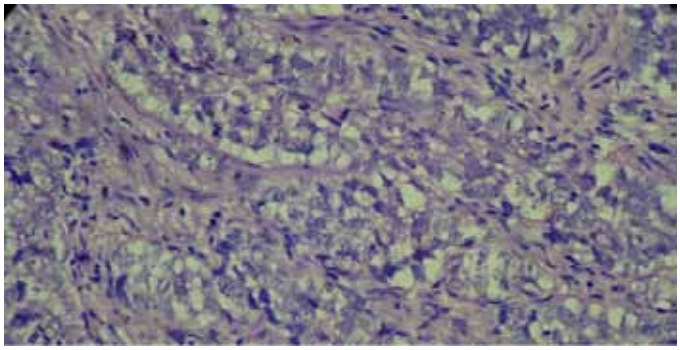


Figure 3: High grade pleomorphism, hyperchromatism, moderate to abundant cytoplasm with multi vacuolation.

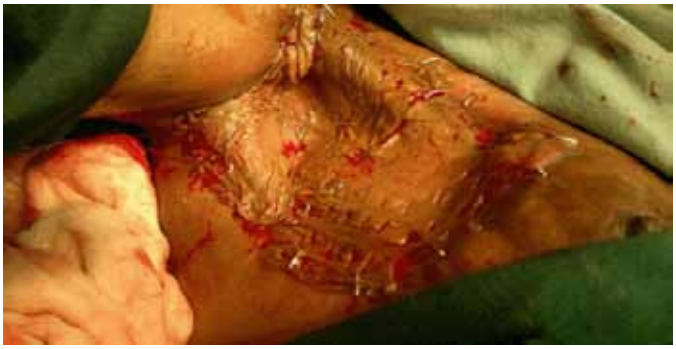


Figure 4: Post skin grafting.

Although it is found from early childhood through the nineties, it is mostly seen in sixth and seventh decade of life [11]. It has been associated with Muir–Torre syndrome which is an autosomal dominant genodermatosis consisting of sebaceous neoplasm viz. sebaceous adenoma, sebaceous carcinoma, or sebaceous epithelioma with or without keratoacanthomas and associated with one or more visceral malignancies [12].

Clinical presentation of sebaceous gland carcinoma is often non-specific and is usually described as a nodule that is pink to yellow red. Most often patient presents with a firm, painless, enlarging nodule on the upper eyelid which is mistaken as chalazion. Present case is a female patient in fifth decade of life with extraocular sebaceous cell carcinoma of axilla with neither ocular involvement nor visceral metastasis.

Histological criteria for sebaceous carcinoma are high mitotic activity, nuclear pleomorphism, lobular architecture and foamy vacuolization of the cytoplasm. Histologically poor prognosis indicators are poor differentiation, presence of lymphatic or vascular permeation, presence of pagetoid cells shown in histology and immunohistochemical staining.

Treatment of sebaceous cell carcinoma requires local resection, cryotherapy, topical chemotherapy, and radiotherapy. We did a wide surgical excision with removal of regional lymph node. There is diverse opinion regarding postoperative irradiation and chemotherapy. Metastasis has been reported to occur as late as five years after the initial diagnosis, lending support to the surveillance of patients with sebaceous carcinoma [10]. Multimodal therapy has been shown to improve both visual prognosis and survival.

CONCLUSION

Though extra ocular sebaceous cell carcinoma is rare aggressive tumor, it should be considered as

differential diagnosis in ulceroproliferative cutaneous swellings in regions where sebaceous glands are found. Histopathology plays a key role in diagnosis.

****

Author Contributions

Aftab Shaikh – 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
Rajesh Chincholkar – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Samarth Agarwal – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Aman Singh – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Arshad Khan – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published
Dhiraj Patil – 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. Nelson BR, Hamlet KR, Gillard M, Railan D, Johnson TM. Sebaceous carcinoma. J Am Acad Dermatol 1995 Jul;33(1):1–15; quiz 16–8.
2. Rao NA, Hidayat AA, McLean IW, Zimmerman LE. Sebaceous carcinomas of the ocular adnexa: A clinicopathologic study of 104 cases, with five-year follow-up data. Hum Pathol 1982 Feb;13(2):113–22.
3. Shields JA, Demirci H, Marr BP, Eagle RC Jr, Shields CL. Sebaceous carcinoma of the ocular region: a review. Surv Ophthalmol 2005 Mar-Apr;50(2):103–22.
4. Bhavarajua VM, Shamim SE, Naik VR, Shaari S. Sebaceous cell carcinoma of scalp - a rare presentation. Malays J Med Sci 2007 Jan;14(1):67–70.
5. Lazar AJ, Lyle S, Calonje E. Sebaceous neoplasia and Torre-Muir syndrome. Curr Diagn Pathol 2007 Aug;13(4):301–19.
6. Kass LG, Hornblass A. Sebaceous carcinoma of the ocular adnexa. Surv Ophthalmol 1989 May-Jun;33(6):477–90.
7. Pang P, Rodríguez-Sains RS. Ophthalmologic oncology: sebaceous carcinomas of the eyelids. J Dermatol Surg Oncol 1985 Mar;11(3):260–4.
8. Rao NA, Hidayat AA, McLean IW, Zimmerman LE. Sebaceous carcinomas of the ocular adnexa: A clinicopathologic study of 104 cases, with five-year follow-up data. Hum Pathol 1982 Feb;13(2):113–22.
9. Justi RA. Sebaceous carcinoma; report of case developing in area of radiodermatitis. AMA Arch Derm 1958 Feb;77(2):195–200.
10. Cohen PR, Kohn SR, Davis DA, Kurzrock R. Muir-Torre syndrome. Dermatol Clin 1995 Jan;13(1):79–89.
11. Schwartz RA, Torre DP. The Muir-Torre syndrome: a 25-year retrospect. J Am Acad Dermatol 1995 Jul;33(1):90–104.
12. Ghosh SK, Bandyopadhyay D, Gupta S, Chatterjee G, Ghosh A. Rapidly growing extraocular sebaceous carcinoma occurring during pregnancy: a case report. Dermatol Online J 2008 Aug 15;14(8):8.

ABOUT THE AUTHORS

Article citation: Shaikh A, Chincholkar R, Agarwal S, Singh A, Khan A, Patil D. Sebaceous cell carcinoma of axilla: A rare case report. Int J Case Rep Images 2016;7(5):350–353.

Aftab Shaikh is working in Department of General Surgery at Grant Government Medical College and Sir JJ, Group of Hospitals, Byculla, Mumbai.

Rajesh Chincholkar is working in Department of General Surgery at Grant Government Medical College and Sir JJ, Group of Hospitals, Byculla, Mumbai.

Samarth Agarwal is Lecturer in Surgery in Department of General Surgery at Grant Government Medical College, Mumbai, India. He earned MBBS degree from Topiwala National Medical College, Mumbai, India and MS General Surgery degree from Grant Government Medical College, Mumbai, India. He has published 10 research papers in national and international academic journals.
His research interests include minimally invasive laparoscopic, urological and bariartric surgeries.
He intends to pursue mch urology in future.

Aman Singh is working in Department of General Surgery at Grant Government Medical College and Sir JJ, Group of Hospitals, Byculla, Mumbai.

Arshad Khan is working in Department of General Surgery at Grant Government Medical College and Sir JJ, Group of Hospitals, Byculla, Mumbai.

Dhiraj Patil is working in Department of General Surgery at Grant Government Medical College and Sir JJ, Group of Hospitals, Byculla, Mumbai.