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Case Report

Esophageal leiomyoma within an epinephric diverticulum ☆

Sameeta Kumari[a], *Muhammad Ibrahim Saeed*[b], *Faisal Waseem Ismail*[b,*], *Muhammad Bilal Ibrahim*[c]

[a] *Medical Graduate, Aga Khan University Hospital, Karachi, Pakistan*
[b] *Section of Gastroenterology, Department of Medicine, Aga Khan University Hospital, Karachi, Pakistan*
[c] *John H. Stroger Jr Hospital of Cook County, Chicago, IL, USA*

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ABSTRACT

Epinephric diverticula are distal esophageal pouches protruding from the epithelial lining of the esophagus while esophageal leiomyomas are benign smooth muscle lesions that constitute a significant percentage of all gastrointestinal leiomyomas. Epinephric diverticula and esophageal leiomyomas are common individually but their co-existence is rare. Moreover, they present asymptomatically but can occasionally present with complains of dysphagia and weight loss. In this paper, we present a 58-year-old Asian man with three months history of indigestion and progressive weight loss. Preoperatively, CT Scan with IV Contrast showed a large soft tissue mass appearing on the right distal esophageal wall, with its lumen communicating with the esophageal lumen, likely representing an epinephric diverticulum. Biopsy and immunohistochemistry stains confirmed the diagnosis of smooth muscle neoplasm, likely a leiomyoma. Later, the patient underwent a two-stage esophagectomy. The postoperative biopsy was consistent with the initial one: therefore, supporting the diagnosis of a leiomyoma. Postoperatively, the recovery remained uneventful.

Introduction

Esophageal diverticula and esophageal leiomyoma are benign pathologies of the esophagus; both present challenges in their respective diagnosis and management. Their co-existence is rare with only a limited number of documented cases [1]. Individually, epinephric diverticula comprise less than 15% of all esophageal diverticulas while esophageal leiomyomas account for almost 10% of all gastrointestinal leiomyomas [2,3].

Esophageal diverticula are characterized by a protusion or a pouch formed from the epithelial lining of the esophagus. One of the types of esophageal diverticula is epinephric diverticulum; they are noted to be found within 10 cm of the

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* Corresponding author.
E-mail address: Faisal.Ismail@aku.edu (F.W. Ismail).

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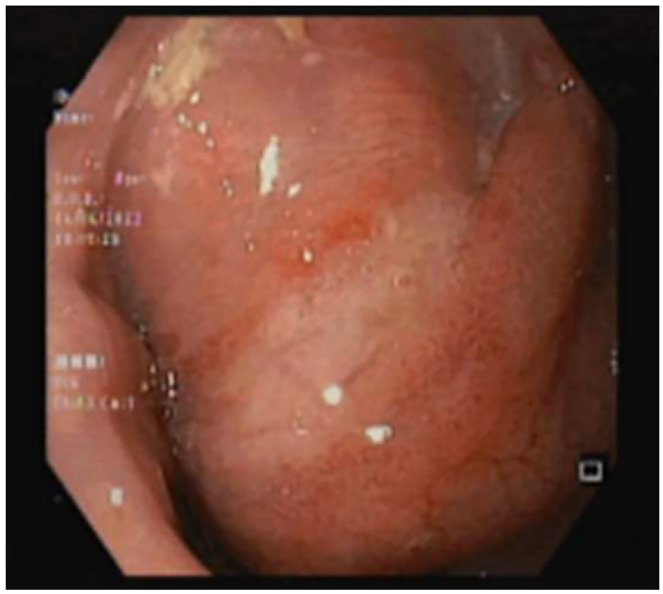

Fig. 1 – Paraesophageal hiatus hernia with subepithelial mass at GEJ.

gastroesophageal junction [4]. On the other hand, leiomyomas are common benign smooth muscle tumors; they predominately arise from the muscularis propria but can rarely involve the muscularis mucosa [5].

On presentation, many patients with epinephric diverticulum are asymptomatic; however, some complain of dysphagia, regurgitation and weight loss. Barium Swallow, computed tomographic (CT) Scan, and endoscopy help not only confirming the diagnosis of diverticulum; but also assist in ruling out any secondary diagnosis [6]. Diverticula can be treated diversely, ranging from being managed conservatively to complete local excision [6]; while leiomyomas can be treated by enucleation, traditional thoracotomy, or in more severe cases by esophagectomy [7]. Once resected, esophageal leiomyomas generally exhibit a low rate of recurrence [8].

Case report

A 58-year-old Asian man presented in the clinic with a long-standing history of rheumatoid arthritis for the past 2 decades. He presented with complains of indigestion, and a substantial weight loss of 6 kgs over the last 3 months. Initial laboratory tests included normocytic normochromic profile on complete blood counts sample with a hemoglobin of 11.9 g/dL, a hematocrit of 35.8%, and a platelet count of 130 × 10^9. These values were slightly below the lower end of normal limit. His liver function tests, prothrombin time, and creatinine, however, were all normal.

The patient was planned for upper endoscopy, which revealed a possible paraesophageal hiatus hernia with subepithelial mass at the gastroesophageal junction (GEJ) (Fig. 1). The examination also showed mild mucosal erythema in the body of the stomach. Biopsy was taken from both sites, which reported moderate acute inflammation with mild foveolar hyperplasia and no evidence of intestinal metaplasia or malignancy from the lesion. The stomach site reported mild inflammation only. CT Abdomen and Pelvis with intravenous contrast was performed. It demonstrated a large soft tissue mass appearing on the right distal esophageal wall with its lumen communicating with the esophageal lumen, likely representing a mass lesion within an epinephric diverticulum (Fig. 2). The mass measured approximately 61 × 48 mm in anterior-posterior and transverse dimensions. Moreover, the right lateral wall appeared significantly thickened measuring 37 mm, raising the possibility of a lesion within the diverticulum.

An endoscopic ultrasound (EUS), performed subsequently confirmed the diagnosis, revealing a large heterogenous subepithelial mass lesion of approximately 5 cm (Fig. 3). Biopsy was taken using a 22G fine needle biopsy (FNB); the microscopic examination showed linear cores and fragments of a spindle cell lesion mixed with blood. The spindle cells were arranged in bundles, had abundant eosinophilic cytoplasm and cigar shaped bland nuclei with occasional prominent nucleoli. However, there were no signs of cytological atypia, mitosis, or necrosis. Immunohistochemical stains led to the diagnosis of a smooth muscle neoplasm, most likely leiomyoma. The tumor tested positive for ASMA and Caldesmon, and negative for DOG-1 (Fig. 4).

Subsequently, the patient underwent a 2-stage esophagectomy. Intraoperative findings included a 5 × 4 cm hard mass at the distal esophagus, with no gross ascites or any other tumor deposits. The postoperative time was uneventful, and the patient was discharged on 7th postoperative day. During his follow-up after 1 month, he remained asymptomatic. The final biopsy confirmed the diagnosis of Leiomyoma with no evidence of malignancy (Fig. 5), as the histopathology report and immunohistochemical stains remained the same on testing. Moreover, stains also showed that the tumor was positive for Desmin and negative for CD117 and CD34.

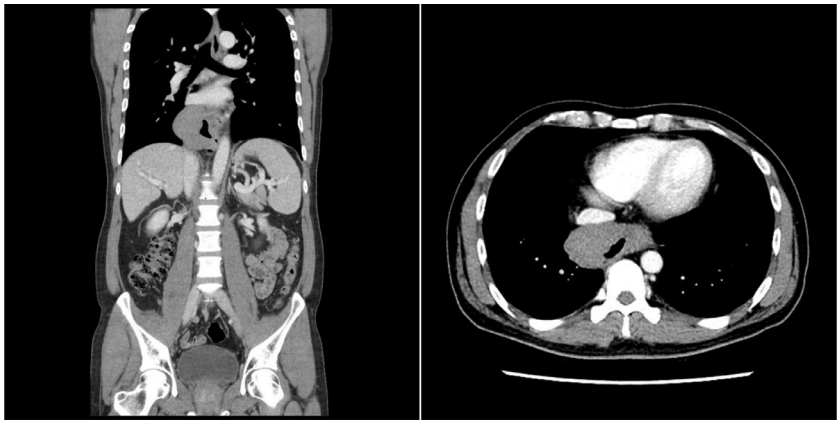

Fig. 2 – CT scan images.

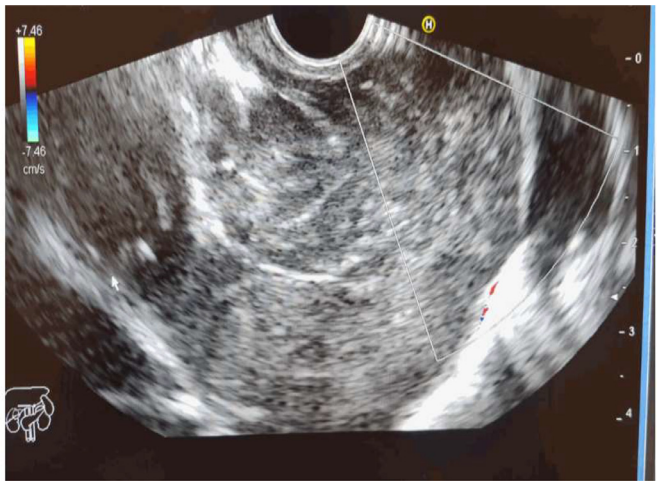

Fig. 3 – EUS image.

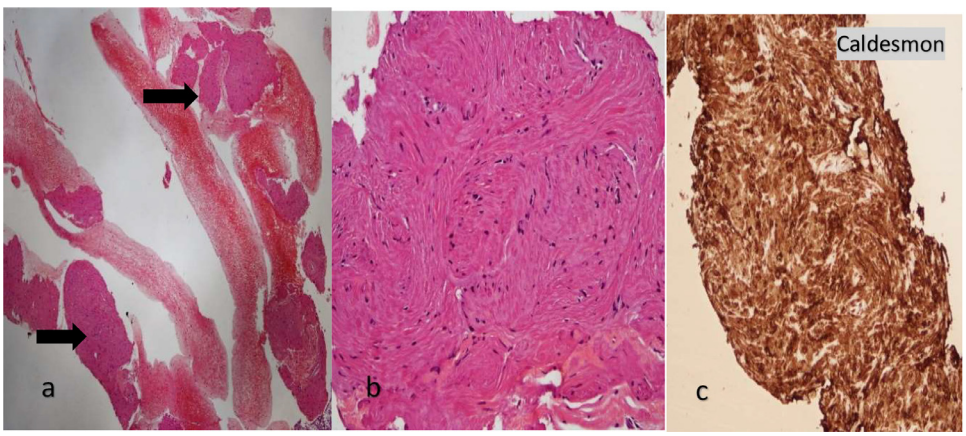

Fig. 4 – H&E sections of endoscopic ultrasound (EUS) guided core biopsy of the esophageal lesion. Low power (A, 4X) shows multiple fragments and linear cores of tissue (→) mixed with blood. The high power (B, 20X) shows interlacing bundles of smooth muscle cells, having abundant eosinophilic cytoplasm and spindle shaped nuclei without any atypia or mitoses. Diffuse positivity of tumor cells by immunohistochemical stain Caldesmon (Fig. 1c) and negative markers for gastrointestinal stromal tumor (DOG-1 & CD117, not shown here) confirms the smooth muscle nature of this spindle cell tumor. A diagnosis of leiomyoma was suggested.

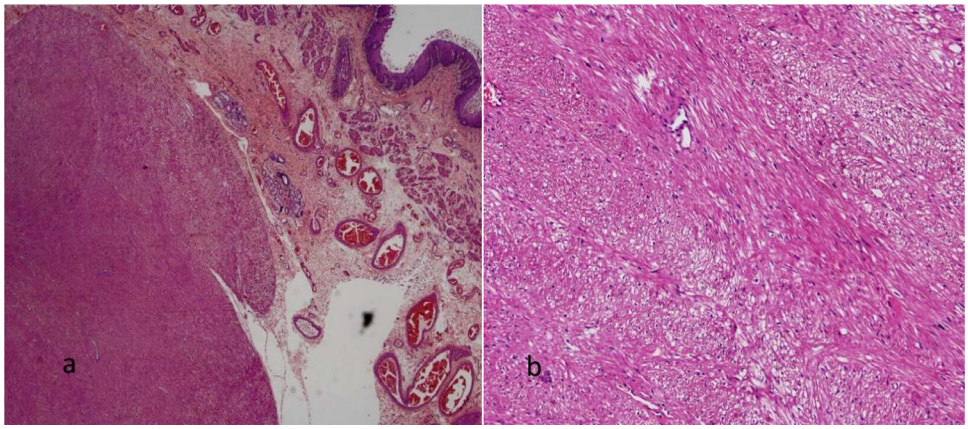

Fig. 5 – H&E sections of resected esophageal lesion. Low power (A, 4X) shows a circumscribed spindle cell tumor, centered in the submucosa and muscularis propria of esophageal wall. The high power (B, 20X) shows smooth muscle tumor of similar morphology as seen in the EUS-guided biopsy. There are no malignant features in the tumor. Immunohistochemical profile is also identical to the biopsy sample. These features are consistent with the diagnosis of Leiomyoma.

Discussion

Esophageal diverticula are characterized by a pouch or a protrusion formed from the epithelial lining of the esophagus. They can be categorized according to the layers of the esophagus involved as well as based on their location. Depending on the layers, true diverticula encompass all layers of the esophagus, while false diverticula bulge through the mucosal and submucosal layers of the esophagus only [9]. Classifying based on their location includes near the pharyngoesophageal area that is, Zenker's diverticulum, in the middle of esophagus, or in the distal esophagus that is, epinephric diverticulum as in our patient [6]. Epinephric diverticula may resemble a paraesophageal hiatal hernia as both can appear as a spherical pouch below the assumed gastroesophageal junction [10]. However, on CT scans, hiatal hernia are thick walled and associated with widening of the esophageal hiatus, thus differentiating it from epinephric diverticulum [11].

The incidence of cancer within a diverticulum is observed at rates of 0.3%-0.7% for pharyngoesophageal, 1.8% for midesophageal, and 0.6% for epinephric diverticula [12]. Benign tumors like leiomyoma are even more infrequent. Moreover, the potential for malignant transformation in a diverticulum is associated with stagnation and should always be a significant consideration when managing a long-standing diverticulum [1]. The definitive surgical management is diverticulectomy by interventional endoscopy; although, it can be treated conservatively [6,13]. However, if they are accompanied by benign lesions, management becomes different. Larger lesions often necessitate partial esophagectomy, while smaller lesions can easily be treated with simple enucleation by traditional thoracotomy or a video-assisted thoracoscopy [7]. On the other hand, hiatal hernias are commonly treated with laparoscopic or robotic repair methods [14].

Leiomyomas are the most prevalent nonmalignant tumors of the esophagus, accounting for approximately 60%-70% of cases [15]. Among these, they are reported to impact the distal third esophagus in 60%, the middle third in 30%, and the upper third in 10% of cases [5]. Moreover, they generally exhibit a low rate of recurrence once resected [8]. The preoperative diagnosis, however, of the leiomyoma is uncertain, as the clinical symptoms and presentation during standard diagnostic evaluation are nonspecific, and similar to some other malignant lesions of the esophagus [7]. Grossly, leiomyomas appear as firm and encapsulated with a smooth or nodular surface. Histologically, they are composed of interwoven smooth muscle cells and present as eosinophilic on hematoxylin-eosin staining. Moreover, these tumors exhibit a lack of blood vessels and an absence of mitotic activity. Immunohistochemistry and leiomyomas have some common features [16]. They are positive for desmin and smooth muscle actin and negative for CD117 and CD34 [16]. Most of these features are similar to the benign lesion in our case thus confirming leiomyoma within the epinephric diverticulum.

Conclusion

In conclusion, epinephric diverticulum in combination with a leiomyoma is a rare finding and an underlying lesion must be excluded in patients diagnosed with esophageal diverticulum before undergoing any intervention. Moreover, the surgical approach also varies when patients are diagnosed with lesions within the diverticulum, in contrast to cases of isolated diverticulum pathology.

Authors contribution

Sameeta Kumari did literature search, and initial write-up of case summary, introduction, report, and discussion, and is responsible for integrity of research. Muhammad Ibrahim Saeed

conceived the idea and edited the manuscript. Faisal Waseem Ismail and Muhammad Bilal Ibrahim did the review and final approval of the manuscript.

Patient consent

The authors confirm that written informed consent has been obtained from the patient, and, he has given approval for this information to be published in this case report.

REFERENCES

[1] Chowdhry M, Spyratou C, Lorenzi B, Kadirkamanathan S, Charalabopoulos A. Association between oesophageal diverticula and leiomyomas: a report of two cases. Case Rep Gastrointest Med 2016;2016:6832535.

[2] Fasano NC, Levine MS, Rubesin SE, Redfern RO, Laufer I. Epiphrenic diverticulum: clinical and radiographic findings in 27 patients. Dysphagia 2003;18(1):9–15.

[3] Linde EM, DiMaio DJ. Solitary esophageal leiomyoma with eosinophilic infiltrate: case report and review of the literature. Dis Esophagus 2011;24(1) E5-7.

[4] Matsumoto H, Kubota H, Higashida M, Manabe N, Haruma K, Hirai T. Esophageal epiphrenic diverticulum associated with diffuse esophageal spasm. Int J Surg Case Rep 2015;13:79–83.

[5] Mujawar P, Pawar T, Chavan RN. Video assisted thoracoscopic surgical enucleation of a giant esophageal leiomyoma presenting with persistent cough. Case Rep Surg 2016;2016:e7453259.

[6] Herbella FAM, Patti MG. Modern pathophysiology and treatment of esophageal diverticula. Langenbecks Arch Surg 2012;397(1):29–35.

[7] De Giacomo T, Bruschini P, Arcieri S, Ruberto F, Venuta F, Diso D, et al. Partial oesophagectomy for giant leiomyoma of the oesophagus: report of 7 cases. Eur J Cardiothorac Surg 2015;47(1):143–5.

[8] Tsai SJ. Benign esophageal lesions: endoscopic and pathologic features. WJG 2015;21(4):1091.

[9] Yam J, Baldwin D, Ahmad SA. Esophageal diverticula. StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023. [cited 2023 September 12]. Available from: http://www.ncbi.nlm.nih.gov/books/NBK532858/.

[10] Holihan J. A rare true epiphrenic diverticulum in a patient with achalasia, misdiagnosed as a paraesophageal hiatal hernia. Ann Clin Case Rep 2019;4:1609.

[11] Fergus VC, Epiphrenic diverticulum mimicking hiatal hernia (Case 6) - Pearls and Pitfalls in Abdominal Imaging [Internet]. [cited 2024 January 1]. Available from: https://www.cambridge.org/core/books/abs/pearls-and-pitfalls-in-abdominal-imaging/epiphrenic-diverticulum-mimicking-hiatal-hernia/B9339B57EE67EDE911A24CD5C52F73D2. (accessed November 5, 2011).

[12] Herbella F a M, Dubecz A, Patti MG. Esophageal diverticula and cancer. Dis Esophagus 2012;25(2):153–8.

[13] Lakhani DA, Hadi YB, Smith M. Epiphrenic diverticulum. Clin Gastroenterol Hepatol 2021;19(8):e75–6.

[14] Ma L, Luo H, Kou S, Gao Z, Bai D, Qin X, et al. Robotic versus laparoscopic surgery for hiatal hernia repair: a systematic literature review and meta-analysis. J Robot Surg 2023;17(5):1879–90.

[15] Rijcken E, Kersting CM, Senninger N, Bruewer M. Esophageal resection for giant leiomyoma: report of two cases and a review of the literature. Langenbecks Arch Surg 2009;394(4):623–9.

[16] Miettinen M, Sarlomo-Rikala M, Sobin LH, Lasota J. Esophageal stromal tumors: a clinicopathologic, immunohistochemical, and molecular genetic study of 17 cases and comparison with esophageal leiomyomas and leiomyosarcomas. Am J Surg Pathol 2000;24(2):211–22.