Annals of Clinical Case Reports

Case Summary
Published: 22 Feb, 2019

A Rare True Epiphrenic Diverticulum in a Patient with Achalasia, Misdiagnosed as a Paraesophageal Hiatal Hernia

*Julie Holihan[1], Craig Floyd[2] and Farzaneh Banki[1,2]**

[1]Department of Surgery, McGovern Medical School at UTHealth, USA

[2]Memorial Hermann Southeast Esophageal Disease Center, Texas, USA

Abstract

Epiphrenic diverticula are pulsion/false diverticula caused by outflow resistance at the level of the gastroesophageal junction and esophageal motility disorders. We present a large epiphrenic diverticulum with clinical and pathological features of a true diverticulum in a patient with achalasia, which was misdiagnosed as a hiatal hernia.

Clinical Summary

A 67-year-old female who had undergone a transabdominal Heller myotomy for achalasia in 1973 presented with a 2-year history of progressive dysphagia to solids and liquids, regurgitation, cough, pneumonia and weight loss of 10 lbs. Chest Computed Tomography (CT) without oral contrast at an outside hospital showed a 10 cm pouch filled with debris above the left hemidiaphragm, diagnosed as a paraesophageal hiatal hernia. The patient was treated with proton pump inhibitors without response. She was then referred by a pulmonologist to thoracic surgery for respiratory symptoms attributed to aspiration caused by gastroesophageal reflux disease and a hiatal hernia.

The videoesophagram at our center showed a dilated esophagus with absence of peristaltic contractions. The distal esophagus was angulated and was connected to a 10 cm pouch in the left hemithorax. The narrowing in the distal esophagus above the pouch resembled the gastroesophageal junction. The pouch was round and filled with debris. The stomach was decompressed and was entirely located in the abdomen. The pouch in the chest mimicked a large paraesophageal hiatal hernia as shown in Figure 1. CT scan with oral contrast showed a pouch filled with debris above the left hemidiaphragm, shown in Figure 2A,2B. Upper endoscopy revealed that the distal esophagus had transformed into a large, round pouch filled with copious fermented food, shown in Figure 2C. A repeat endoscopy after 3 days of clear liquid diet, under general anesthesia to protect from aspiration, revealed a large pouch and no evidence of hiatal hernia, shown in Figure 2D. There were no dual lumens, one into the diverticulum and one into the stomach, as are usually seen in a false/pulsion diverticulum. Biopsies of the cavity showed squamous mucosa. A motility catheter placed under endoscopic guidance showed absence of esophageal contractions and failure of relaxation of the lower esophageal sphincter consistent with achalasia. Because of the history of prior Heller myotomy via a midline laparotomy and potential adhesions, and the size of the diverticulum, the procedure was planned as a thoracotomy, diverticulectomy and possible partial fundoplication.

A left posterolateral thoracotomy in the 7th intercostal space revealed a distended diverticulum with retained contents, shown in Figure 3A. A nasogastric tube was passed for decompression. There were dense adhesions between the diverticulum and the pericardium, left lower lobe, aorta and diaphragm which were divided. The diverticulum was a large pouch in continuation with the distal esophagus and seemed to contain all esophageal layers. There were dense adhesions between the esophagus and aorta which continued beyond the gastroesophageal junction possibly as the result of prior myotomy, but there was no visible plane of myotomy in the chest. The adhesions were divided and a completion Heller myotomy was performed distal to the diverticulum, extending beyond the gastroesophageal junction. The diverticulectomy was performed over a 48 French bougie using an endo GIA stapler and the staple line was imbricated with interrupted stitches. As the result of adhesions at the level of gastroesophageal junction in the abdomen, no partial fundoplication was performed. Endoscopy showed an easy access into the stomach, no retained food in the esophagus and no evidence of leak. Pathology showed presence of all three layers, including mucosa, muscular layer an adventitia, which confirmed the presence of a true diverticulum in figures Figure 3B,3C.

OPEN ACCESS

Correspondence:
Farzaneh Banki, Department of Surgery, McGovern Medical School at UTHealth, Memorial Hermann Southeast Esophageal Disease Center, 11800 Astoria Blvd, Houston, TX 77089, USA,
E-mail: Farzaneh.Banki@uth.tmc.edu

Received Date: *27 Jan 2019*
Accepted Date: *20 Feb 2019*
Published Date: *22 Feb 2019*

Citation:
Holihan J, Floyd C, Banki F. A Rare True Epiphrenic Diverticulum in a Patient with Achalasia, Misdiagnosed as a Paraesophageal Hiatal Hernia. Ann Clin Case Rep. 2019; 4: 1609.

ISSN: 2474-1655

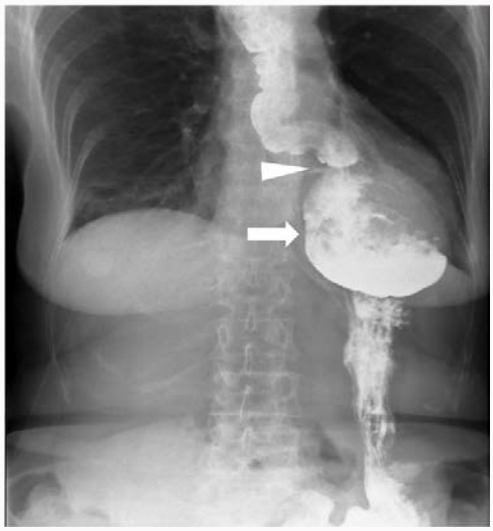

Figure 1: The barium esophagram shows a misread gastroesophageal junction (arrowhead) above a true epiphrenic diverticulum (arrow).

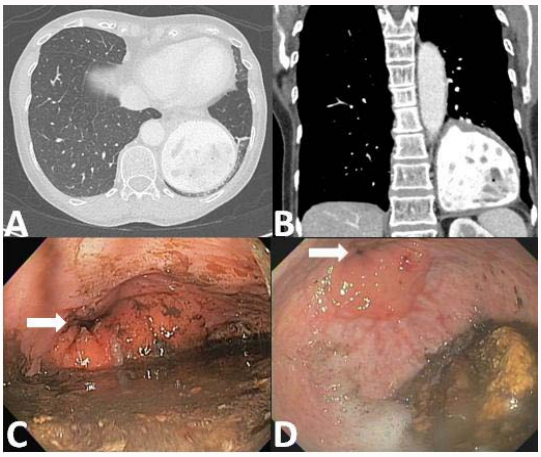

Figure 2: A,B) Axial and coronal views of the CT scan showing a 10 cm pouch filled with debris. C) Initial upper endoscopy showing a large pouch in the distal esophagus filled with food and fluid with poor visibility and a connection with the stomach at the level of the gastroesophageal junction (arrow). D) Repeat endoscopy after 3 days of clear liquid diet and under general anesthesia shows a clearer view of the esophageal pouch with squamous mucosa, only one exit inferior to the pouch via the gastroesophageal junction (arrow) and no evidence of hiatal hernia.

Postoperative video esophagram showed obliteration of the pouch, an easy passage of contrast into the stomach and no leak. The patient was started on a clear liquid diet on postoperative day one and was discharged home on a full liquid diet for two weeks. She was seen in our clinic on postoperative day 21, doing well, tolerating PO intake and had gained 2 lbs. She was instructed to have soft diet and advance to regular diet as tolerated. She will be followed in three months and one year after surgery in our clinic.

Discussion

Epiphrenic diverticula are false diverticula composed of esophageal mucosa, caused by a hypertensive or non-relaxing lower esophageal sphincter and associated motility disorders [1-3]. In contrast, true diverticula are composed of all esophageal layers and are usually associated with mediastinal inflammatory processes such as tuberculosis [4].

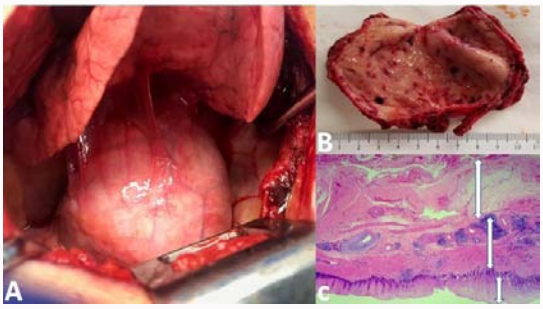

Figure 3: A) Intraoperative view of a large epiphrenic diverticulum with adhesions to the left lower lobe. B) Resected 10 cm epiphrenic diverticulum with 5 mm thickness. The external surfaces were congested and indurated. The mucosa showed a lobulated architecture with focal hemorrhagic stippling. C) Pathology slide shows a true diverticulum with adventitia (top arrow), muscular layer (middle arrow), and squamous mucosa (bottom arrow).

Symptoms associated with an esophageal diverticulum could be similar to a hiatal hernia [2]. In our case, an unusual presentation of a true diverticulum as a round pouch below the presumed gastroesophageal junction, resembling a hiatal hernia, made the diagnosis more complex. The anatomical location which was thought to be the gastroesophageal junction was probably caused by the combination of distal esophageal angulation and an outflow resistance created by the distended diverticulum filled with retained contents, creating a proximal a high-pressure zone. In parallel, the presence of the diverticulum, a marker of outflow resistance is probably the result of an incomplete myotomy and/or scar tissue at the level of gastroesophageal junction, creating a distal high-pressure zone. The presence of all three esophageal layers in the pathology specimen which proves the presence of a true diverticulum, confirms that the diverticulum did not consist of an out pouching of the mucosa usually seen in patients with epiphrenic diverticulum, but more likely because of gradual dilation of the distal esophagus with all its three layers, possibly as the result of an incomplete Heller myotomy. The rare presentation increases the diagnostic complexity, hence the reason for misdiagnosis as a paraesophageal hiatal hernia.

This case emphasizes the importance of a thorough assessment of patients with esophageal disease. Particularly a true diverticulum without evidence of an inflammatory process in the mediastinum, an unusual location of the presumed gastroesophageal junction, and the transformation of the distal esophagus into a large true diverticulum mandated a comprehensive evaluation. The use of complementary diagnostic tools allowed correct diagnosis and treatment of a rare true epiphrenic diverticulum, misdiagnosed as a hiatal hernia, which was treated *via* left thoracotomy, diverticulectomy and completion Heller myotomy with good outcome.

References

1. Cattaneo, Stephen M. Pearson's thoracic & esophageal surgery, 3rd ed. Ann Surg. 2008;248(6):1103-4.
2. Brandeis AE, Singhal S, Lee TH, Mittal SK. Surgical management of epiphrenic diverticulum: A single-center experience and brief review of literature. Am J Surg. 2018;216(2):280-5.
3. Zaninotto G, Portale G, Costantini M, Zanatta L, Salvador R, Ruol A. Therapeutic strategies for epiphrenic diverticula: systematic review. World J Surg. 2011;35(7):1447-53.
4. Herbella FA, Patti MG. Modern pathophysiology and treatment of esophageal diverticula. Langenbecks Arch Surg. 2012;397(1):29-35.