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Esophageal Leiomyomatosis Combined With Intrathoracic Stomach and Gastric Volvulus

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ABSTRACT
Case Report: A 42-year-old female presented with long-standing symptoms suggestive of gastroesophageal reflux disease improved after proton pump inhibitor treatment. An upper endoscopy revealed an intrathoracic position of the stomach (type 4 hiatal hernia) with no mucosal abnormality. Barium swallow demonstrated gastric herniation with gastric volvulus without stenosis. A computed tomographic scan confirmed the intrathoracic location of the stomach associated with thickening and edema of the gastric wall due to gastric volvulus, but no evidence of malignancy. The patient was scheduled for laparoscopic gastric repositioning with anterior hemifundoplication. Due to the incidental intraoperative finding of a large distal esophageal tumor (frozen section: esophageal leiomyomatosis), the operation was converted to conventional distal esophagectomy and proximal gastrectomy with reconstruction using a Merendino procedure. Final histology revealed extensive circumferential leiomyomatosis of the distal esophagus with a diameter of 10 cm. Esophageal leiomyomatosis is an extremely rare pathological finding with <100 cases reported in the literature.

Key Words: Esophageal leiomyomatosis, Merendino procedure, Upside-down stomach, Intrathoracic stomach.

INTRODUCTION
Benign tumors of the esophagus are rare. Leiomyoma, occurring mostly as a solitary lesion, is the most common benign tumor, accounting for only 0.4% of all esophageal neoplasms.1 Most of the reported cases (<100) have occurred in young patients in association with hereditary disease (Alport syndrome) or in combination with other visceral leiomyomatosis.2 Usually, these patients present with dysphagia and respiratory symptoms at a young age.

We report the case of a patient who was scheduled for laparoscopic gastric repositioning and anterior hemifundoplication due to an intrathoracic stomach. The procedure was converted to conventional distal esophagectomy with proximal gastrectomy after the intraoperative discovery of a large distal esophageal tumor.

METHODS
Patient’s Data
A 42-year-old female with no relevant past medical history presented to our outpatient clinic complaining of long-standing symptoms suggestive of reflux disease that were relieved by proton pump inhibitor treatment (40 mg omeprazole daily). No history of dysphagia or retrosternal discomfort and no history of familial Alport syndrome were reported. Past surgical history included appendectomy at the age of 5 years and transvaginal conization. A physical examination did not reveal any abnormalities.

Diagnostic Tests
The results of all blood tests were within normal ranges. Upper endoscopy showed an intrathoracic stomach (type 4 hiatus hernia) with the possibility of gastric volvulus, but no mucosal abnormalities or stenosis. The barium swal-
low demonstrated herniation of the stomach associated with organo-mesenteric volvulus (Figures 1 and 2). Contrast-enhanced computed tomographic (CT) scan of the chest and abdomen confirmed the presence of an intrathoracic stomach associated with thickening and edema of the gastric wall most likely due to gastric volvulus (Figure 3). The patient was scheduled for laparoscopic gastric repositioning and anterior hemifundoplication.

Figure 1. Barium swallow study showing intrathoracic stomach (upside-down-stomach).

Figure 2. Barium swallow study showing intrathoracic stomach with organo-mesenteric volvulus.

Figure 3. Computed tomography of the chest: axial slices demonstrating intrathoracic stomach with thickening of the wall. Note the orally administered contrast medium located in the stomach.
RESULTS

Operative Treatment

Initially, a laparoscopic approach using a 5-trocar technique was performed. After gas insufflation and inspection of the abdominal cavity, we noticed a 1-cm to 1.5-cm liver lesion (segment 3) that was not described by preoperative CT scan. Then, we explored the hiatus. A large hiatal defect was present with more than 2/3 of the stomach herniated through it. The liver lesion was resected (wedge resection) by using a Harmonic knife. A frozen section revealed focal nodular hyperplasia.

Furthermore, we focused on the hiatus where we started with the repositioning of the stomach. The mobilization of the hernial sac was done successfully all around the hiatus apart from the left side. At this point, a paraesophageal mass lesion with a diameter of approximately 1.5 cm was detected. Macroscopically, we thought the tumor could be an enlarged lymph node or GIST. It was removed and sent for histopathology, which revealed the presence of a mesenchymal tumor.

We tried to continue the procedure laparoscopically under gastroscopic control with the aim of tumor excision. Due to the large tumor size (macroscopically >10 cm along the distal esophagus), we preferred to convert the procedure to laparotomy.

The hiatus was opened using the Harmonic knife for proper mobilization of the bulky esophagus. The decision was made to do proximal gastrectomy with distal esophagectomy and reconstruction using either the Merendino procedure or gastric pull up with esophagogastrostomy (Figure 4). Due to the young age of the patient and poor quality of life caused by intractable reflux, we decided to perform the Merendino procedure. After the esophagus was resected 2 cm above the proximal tumor edge, the proximal part of the stomach was resected using a linear stapler.

The reconstruction was done after preparation of a jejunal limb 15 cm to 20 cm in length, 30 cm to 40 cm away from the Treitz ligament. It was pulled up in retrocolic retrogastric fashion. An isoperistaltic esophagojejunostomy was performed using a circular stapler, followed by manual jejunogastrostomy and jejunoojejunostomy. An anterior hiatalplasty was performed using nonabsorbable suture material with jejunophrenicopexy and gastropexy to the left crus (Figure 4). The procedure was finished by creation of a catheter jejunostomy for postoperative feeding.

Histology

The final histopathology report confirmed a circumferential leiomyomatosis of muscularis propria of the lower esophagus 10.5 cm in length. The mucosa was intact with intestinal metaplasia at the distal esophagus and cardia. No evidence of malignancy was found (Figures 5 and 6).

Postoperative Course

The postoperative course was uneventful apart from dyspnea caused by left-sided pleural effusion. It was resolved by pleural catheter insertion. The patient was informed about her diagnosis of leiomyomatosis that needs regular follow-up for early detection of possible recurrence. She was discharged on the tenth postoperative day with antiemetic therapy and PPI.

The patient was seen at our unit 6 weeks after surgery. She had no complaints regarding reflux disease, and she is tolerating a normal diet well. The feeding jejunostomy was removed, and the daily dose of PPI was reduced to 20 mg of omeprazole. Two weeks later, this medication was discontinued.
Esophageal leiomyomatosis is a rare benign neoplastic condition in which proliferation of the smooth muscle layer of the esophagus leads to circumferential thickening. Usually, the distal part of the esophagus is affected. This correlates with the increased amount of muscle mass in this part. Though in 35% of cases, there may be diffuse involvement of the whole esophagus, in 80% of cases it extends to the proximal part of the stomach. 5,4 Histologically, this condition is characterized by circumferential proliferation of smooth muscle layer with minimal cellular atypia, but without mitosis or vascular invasion (Figures 5 and 6).

The first publication about this entity was done by Hall in 1916. 5 He described this condition in a 17-year-old girl who had a history of dysphagia: until now, <100 cases have been reported. Esophageal leiomyomatosis tends to occur in young adults and in children. The average age of presentation is 11 years. It is twice as common in males as in females.

Regarding the clinical presentation, most patients present with dysphagia that is usually progressive and longstanding over many years. It is important to differentiate it from that occurring in patients with achalasia in which dysphagia is also longstanding and progressive, but it tends to occur in adolescents. Dysphagia caused by malignancy does not have such a longstanding course. 6 Other digestive symptoms include vomiting, retrosternal pain, dyspepsia, and weight loss. Those patients may also present with respiratory symptoms like chest infection and dyspnea. 7 This condition can occur sporadically, or it may be associated with hereditary diseases like Alport syndrome, characterized by deafness, cataracts, and hematuria. It may present as isolated esophageal leiomyomatosis or in association with other visceral forms of leiomyomatosis like rectal, tracheobronchial, and others.

The preoperative diagnosis of this condition is difficult. CT scan, barium study, and endoscopic ultrasound (EUS) are the mainstay of diagnosis. The CT scan usually shows circumferential thickening of the esophageal wall that extends to the cardia. This feature differentiates esophageal leiomyomatosis from achalasia. 8 Barium swallow findings are similar to those for achalasia. It is not easy to differentiate these 2 entities, though the narrowed segment in achalasia is usually shorter than in leiomyomatosis. EUS is a promising tool for diagnostic accuracy in this pathology. Upper endoscopy also is helpful because it may show irregularity of the wall due to submucosal lesions that are mostly covered by normal mucosa. Chest radiography may show widening of the mediastinum.

Treatment of esophageal leiomyomatosis is surgical. A total or subtotal esophagectomy including proximal gastrectomy must be performed depending on the extent of the disease. The esophagus can be replaced either by colon or by stomach. Each procedure has its advantages and disadvantages in terms of operative difficulties and postoperative short- and long-term morbidities. Regarding our patient, who was already suffering from reflux disease, we preferred to use the Merindino procedure after distal esophagectomy with proximal gastrectomy. This is a more effective reconstruction for preventing reflux symptoms. 9 The Merendino technique was introduced in 1955.
by Merendino and Dillard\textsuperscript{10} as an option for reconstruction after gastroesophageal junction tumors.

\textbf{CONCLUSION}

A surgeon has to be aware of intraoperative incidental findings, ie, converting laparoscopic procedure and performing conventional resection and reconstruction with minimal morbidity. We are convinced that in this case Merendino’s procedure was the best reconstructive option, because our patient was already suffering from reflux symptoms.

\textbf{References:}

1. Seremetis MG, Lyons WS, deGuzman VC, et al. Leiomyoma of the esophagus. An analysis of 838 cases. \textit{Cancer}. 1976;38:2166–2177.

2. Lonsdale RN, Roberts PF, Vaughan R, et al. Familial oesophageal leiomyomatosis and nephropathy. \textit{Histopathology}. 1992;20:127–133.

3. Federici S, Ceccarelli P, Bernardi F et al. Esophageal leiomyomatosis in children: report of a case and review of the literature. \textit{Eur J Pediatr Surg.} 1998;8:358–363.

4. Bourque M, Spigland N, Bensoussan A, et al. Esophageal leiomyoma in children: two case reports and review of the literature. \textit{J Pediatr Surg.} 1989;24:1103–1107.

5. Hall A. A case of diffuse fibromyoma of the esophagus, causing dysphagia and death. \textit{Q J Med.} 1916;9:409–428.

6. Sidhu R, Sood BP, Kalra N, et al. Imaging features of esophageal leiomyomatosis: a case report. \textit{Clin Imaging}. 2002;26:293–295.

7. Leborgne J, Le Neel JC, Heloury Y et al. Diffuse esophageal leiomyomatosis. \textit{Apropos of 5 cases with two familial cases. [in French] Chirurgie}. 1989;115:277–286.

8. Levine MS, Buck JL, Pantongrag-Brown L. Esophageal Leiomyomatosis. \textit{Radiology}. 1996;199:533–536.

9. Hölscher AH, Vallböhmer D, Gutschow C, Bollschweiler E. Reflux esophagitis, high-grade neoplasia, and early Barrett’s carcinoma—what is the place of the Merendino procedure? \textit{Lancet.} 2009;374:417–424.

10. Merendino KA, Dillard DH. The concept of sphincter substitution by an interposed jejunal segment for anatomic and physiologic abnormalities at the esophagogastroduodenal junction; with special reference to reflux esophagitis, cardiospasm and esophageal varices. \textit{Ann Surg.} 1955;142:486–506.