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Giant Esophageal Leiomyoma: Diagnostic and Therapeutic Challenges

Authors' Contribution:

Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABEF 1 **Hatem Elbawab**
BE 2 **Abdullah Fahad AlOtaibi**
BEF 2 **Ammar A. Binammar** 
BEF 2 **Dhuha N. Boumarah**
BEF 2 **Turki Muslih AlHarbi** 
BF 1 **Farouk T. AlReshaid** 
BF 1 **Zeead M. AlGhamdi** 

1 Thoracic Unit, Department of Surgery, King Fahd Hospital of the University, College of Medicine, Imam Abdulrahman Bin Faisal University, Dammam, Saudi Arabia
2 College of Medicine, Imam Abdulrahman Bin Faisal University, Dammam, Saudi Arabia

Corresponding Author: Hatem Elbawab, e-mail: hybawab@iau.edu.sa

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Patient: **Male, 24-year-old**
Final Diagnosis: **Giant esophageal leiomyoma**
Symptoms: **Shortness of breath • productive cough • dysphagia**
Medication: —
Clinical Procedure: —
Specialty: **Surgery**

Objective: **Challenging differential diagnosis**

Background: Leiomyoma is a rare, benign, esophageal tumor that does not often measure >10 cm. Here, we report a case of giant esophageal leiomyoma in a 24-year-old man.

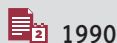
Case Report: A 24-year-old man who smoked and had primary hypertension and glucose-6-phosphate dehydrogenase deficiency presented with a history of shortness of breath and productive cough with yellowish sputum, a long history of dysphagia to solid food, and a weight loss of 7 kg over 2 months. A chest X-ray revealed a mediastinum with a width >8 cm. Computed tomography of the patient's chest revealed a multilobulated mass that originated from the upper and middle thoracic esophagus, caused severe narrowing of his esophageal lumen, and was compressing his trachea and right main bronchus.

Resection of the tumor was performed and, because of the large defect after the surgery and the mucosal necrosis, the patient underwent an Ivor-Lewis esophagectomy. His postoperative course was uneventful. He had no symptoms when he was seen in the outpatient clinic for follow-up and fully recovered.

Conclusions: Giant esophageal leiomyoma (GEL) is a rare oncological entity that presents several diagnostic and therapeutic challenges because of the scarcity of information in the medical literature on surgical management. The descriptions of techniques for surgical resection of GEL do not include ways to effectively perform subsequent reconstruction. The aim of the present paper was to contribute to this scant information by reporting our experience with performing an Ivor-Lewis esophagectomy to manage a case of GEL.

Keywords: **Esophageal Neoplasms • Esophagectomy • Ischemia**

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Background

Leiomyoma is the most common benign esophageal tumor, although it is still rare when compared to the prevalence of carcinoma. It is usually found as a single lesion in the middle or lower third of the esophagus [1]. Leiomyoma can occur in patients of any age, but the peak incidence is in the third to fifth decades of life. The ratio of cases in men and women is approximately 2: 1 [1,2]. Patients with leiomyoma typically have nonspecific symptoms that are not long-standing. Dysphagia, pain, and weight loss are the most common issues [1]. A small percentage of esophageal leiomyomas grow to be >10 cm and are known as giant esophageal leiomyomas (GELs) [3,4]. Most patients with GEL experience symptoms such as dysphagia and chest congestion. Surgical excision is the mainstay of management of GEL and is associated with a good prognosis and quality of life [4]. Herein, we report a case of GEL originating from the distal esophagus that resulted in respiratory symptoms and was successfully treated with an Ivor-Lewis esophagectomy.

Case Report

A 24-year-old man diagnosed with primary essential hypertension at age 14 years and who had glucose-6-phosphate dehydrogenase (G6PD) deficiency presented to the Emergency Department with a 3-day history of shortness of breath and a productive cough with yellowish sputum. He reported that the shortness of breath started suddenly and was continuous and progressive, unrelated to exertion, and relieved by coughing and expectoration of sputum. Since childhood, the patient had experienced chronic dysphagia when he ate solid food, which had worsened during the previous 2 months. During that same period, he also lost his appetite and unintentionally lost 7 kg of weight.

The patient had no history of fever, fatigue, orthopnea, or odynophagia. He had smoked 1 1/2 packs of cigarettes per day since he was 12 years old and smoked a shisha (a waterpipe for tobacco) since he was 7 years old. He also consumed alcohol occasionally. The patient's grandfather had a history of esophageal disease, which had been treated surgically 30 years ago, but neither the pathology nor the type of surgery had been documented.

Physical examination of the patient was unremarkable, except for mild conjunctival pallor. He was tachypneic, tachycardic, and hypertensive, with blood pressure as high as 177/130 mmHg.

A chest X-ray revealed a mediastinum with a width >8 cm (Figure 1). A lateral chest X-ray revealed a posterior mediastinal paraspinal mass. The differential diagnosis was esophageal

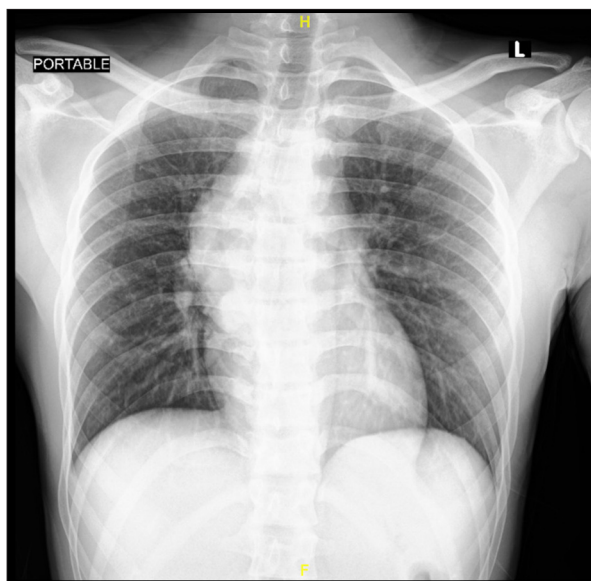


Figure 1. A posteroanterior chest X-ray showing widening of the mediastinum.

leiomyoma, lymphoma, and aortic dissection. CT of the chest revealed a multilobulated mass originating from the upper and middle thoracic esophagus that caused severe narrowing of the esophageal lumen and compression of the trachea and the right main bronchus (Figure 2). A barium swallow study showed narrowing of the mid-esophagus because of the compressive effect of the mass, and deviation of the structure to the right (Figure 3). Upper gastrointestinal endoscopy showed severe esophageal luminal narrowing with intact overlying mucosa (Figure 4). The patient's long history of dysphagia, his young age, and the size of the mass, together with the intact mucosa, favored GEL as his diagnosis.

The patient consented to enucleation of the GEL and a possible esophagectomy and stomach pull-up. The surgery was performed under general anesthesia as a right posterolateral thoracotomy through the fifth intercostal space. The initial exploration revealed a large mid-thoracic posterior mediastinal mass, which was hard and extended from the azygos vein superiorly to 10 cm above the right diaphragmatic cupola. The azygos vein was ligated and transected between 2 silk ties and ligating clips. Meticulous dissection was carried out with alternating sharp and blunt techniques to enucleate the tumor. The excised mass measured approximately 12×9 cm and was lobulated and encircled the esophageal mucosa in a horseshoe shape (Figure 5). After tumor enucleation, the mucosa exhibited ischemic changes with multiple tears. Therefore, an Ivor-Lewis esophagectomy was the optimal technique for restoration of gastrointestinal (GI) continuity. The thoracotomy incision was closed temporarily over a chest tube and the patient was placed in a supine position. A midline laparotomy incision was made in his abdomen and extensive dissection and ligation was performed of surrounding

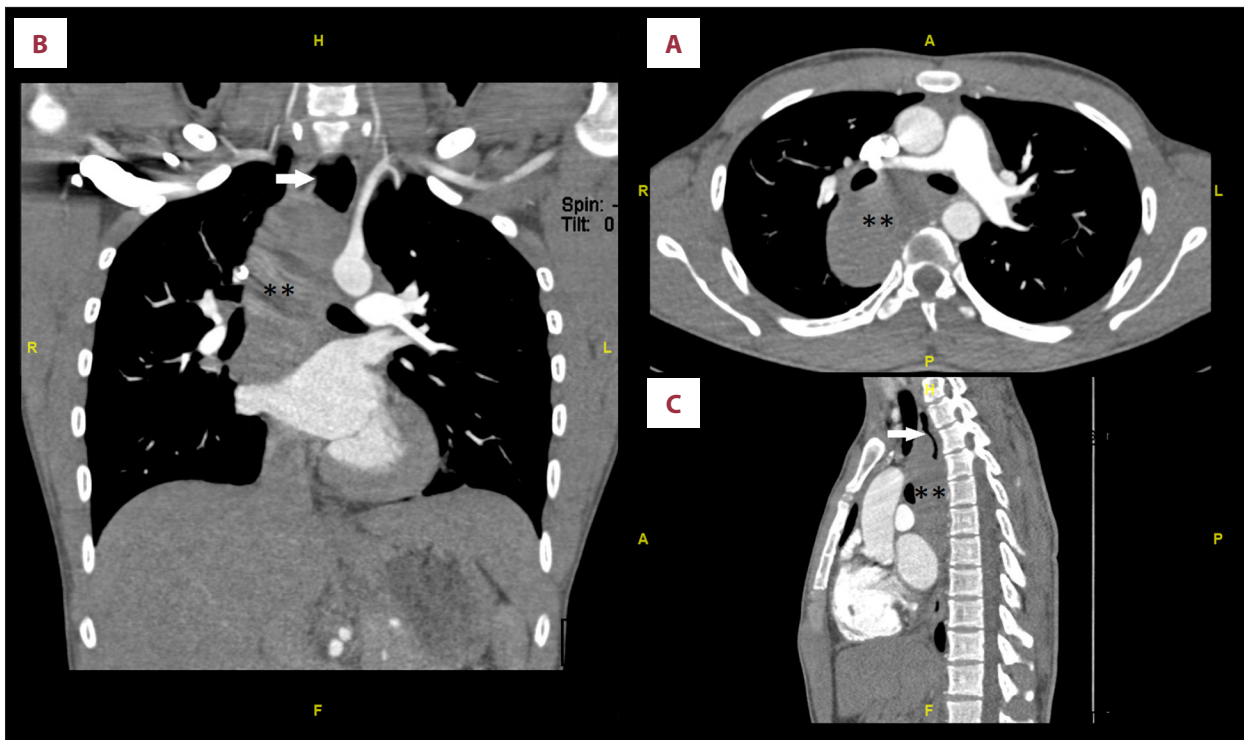


Figure 2. Axial computed tomography of the chest **A:** with contrast, **B:** with coronal reconstruction, and **C:** with sagittal reconstruction showing a large, posterior, mediastinal mass (double asterisk). There is severe narrowing of the esophageal lumen and a dilated superior esophageal segment (white arrow).

gastric vasculatures, preserving the right gastroepiploic artery. To facilitate reconstruction of the patient's GI tract, the lesser curvature was dissected so that his stomach would be shaped like a tube. Pyloroplasty and jejunostomy, using the Witzel technique, also were performed.

Before the patient's abdomen was closed, a gastric tube was placed in the thoracic cavity through the hiatal opening. Thereafter, the previous thoracotomy incision was opened again to access the thorax. The patient's esophagus was transected above the level of the azygos vein and then the caudal end was aligned with the gastric tube and end-to-end anastomosis was achieved using an EndoGIA™ stapler. To assess the integrity of the anastomosis, methylene blue was injected through the patient's nasogastric tube; there was no leak. The patient's thoracotomy wound then was closed after 2 chest tubes had been inserted and connected to an underwater seal. He was taken to the Intensive Care Unit, where he was sedated, intubated, and remained on a nasogastric tube.

The patient's postoperative course was uneventful. Two days later, he had significant clinical improvement and was extubated and transferred to a regular hospital ward. He was discharged 10 days after surgery when an upper gastrointestinal Gastrografin contrast study showed no leaks (Figure 6). The patient's pathology report confirmed the diagnosis of leiomyoma.

The mass measured approximately 12×9×5 cm, had no evidence of malignancy, and was composed of spindle cells with abundant cytoplasm and elongated nuclei. The nuclei had minimal atypia and very few mitotic figures. Immunohistochemical staining was positive for smooth muscle actin and caldesmon and negative for S100 protein, ALK-1, and B-catenin.

When the patient was seen in the clinic 1 week after discharge, he had no symptoms. A chest X-ray was taken, which was normal, and his stitches were removed. He was seen in the clinic again 2 weeks later and continued to be symptom-free. He was given an appointment for thoracic clinic follow-up 1 year later.

Discussion

Leiomyoma is considered to be the most common benign esophageal tumor. The muscularis of the esophagus is the anatomical origin of this tumor, and it is most commonly found in the middle and lower thirds of the esophagus. Patients with esophageal leiomyoma often have nonspecific symptoms [1]. Tumors >10 cm in diameter are known as GELs [4].

When present, the symptoms are usually not specific and are long-standing. As in the present case, dysphagia, pain, and weight loss are the most common symptoms [1]. Our patient

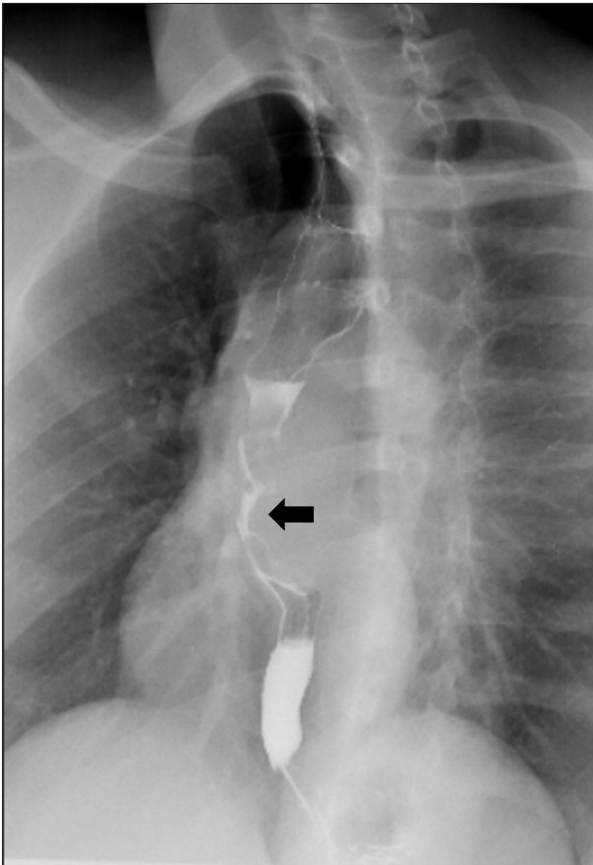


Figure 3. A preoperative barium swallow study showing a giant esophageal leiomyoma that is associated with mid-esophageal narrowing (black arrow) and deviation of the esophagus to the right.

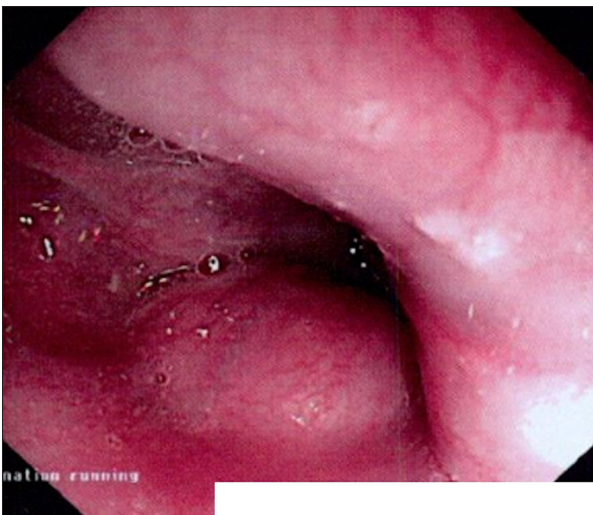


Figure 4. Esophageal endoscopy showing severe esophageal luminal narrowing with intact overlying mucosa.

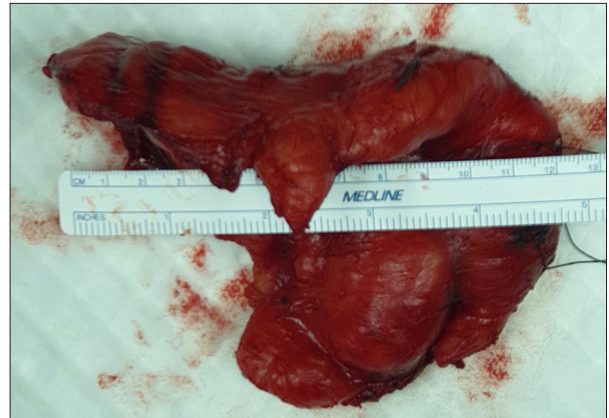


Figure 5. The excised mass measured 12x9 cm and was horseshoe-shaped.

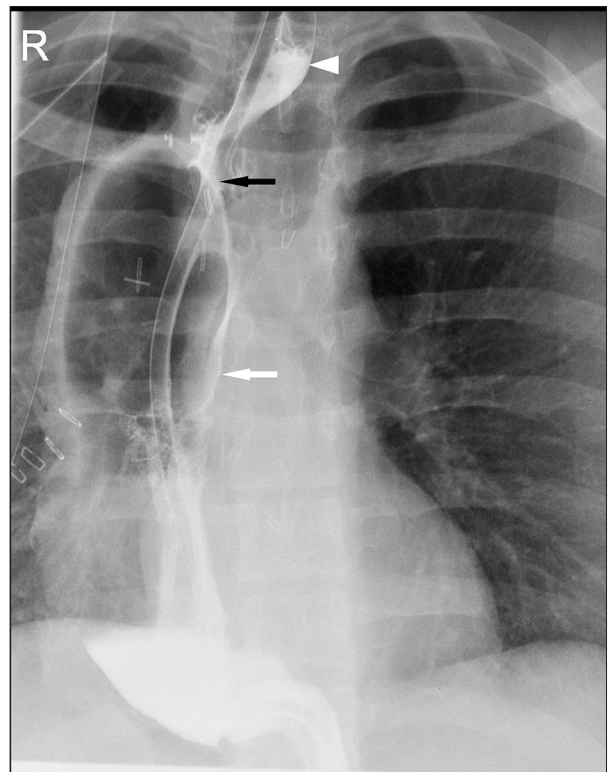


Figure 6. A postoperative upper gastrointestinal Gastrografin contrast study showing no leakage. The esophagus (arrowhead), esophago-gastric anastomosis (black arrow), and gastric tube (white arrow) are visible.

had shortness of breath because his 12x9x5 cm mass was compressing his trachea and right main bronchus. Compression of the trachea is uncommon and has been reported in a single case report [5]. Our patient's other symptoms were cough, epigastric pain, heartburn, regurgitation, and night sweats. Esophageal leiomyoma is usually located in the middle and lower thirds of the esophagus [1]. However, masses in the upper and middle esophagus are not uncommon.

Radiological investigations, such as barium swallow, CT scan, esophagoscopy, and endoscopic ultrasound (EUS), are considered to be useful for diagnosing esophageal leiomyoma [6]. A CT scan can show the size and location of a paraesophageal mass [7]. In the present case, the CT images revealed a mass originating from the upper and middle esophagus, accompanied by esophageal lumen narrowing and compression of adjacent airways. In addition, esophagoscopy showed a severely narrowed esophageal lumen with overlying intact mucosa. Seremetis et al considered these findings to be characteristic of esophageal leiomyoma [1]. The role of preoperative biopsy for esophageal leiomyoma is still debatable. Sun and colleagues suggested that it is not always necessary because of the risk of mucosal damage during enucleation and the probability that insufficient material will be obtained to make a diagnosis [8].

For assessment of esophagus leiomyoma, EUS is used to determine the exact position of the mass in relation to the esophageal wall as well as the mediastinum. Moreover, EUS is highly accurate for evaluating these types of lesions and it can distinguish cystic from solid esophagus submucosal masses [9,10]. To make the diagnosis and select treatment for esophageal leiomyoma, it is crucial to determine the depth of the tumor. This can be done with EUS, which can be used to visualize all layers of the esophageal wall, enabling the layer of the origin of a submucosal mass to be determined [1]. In the present case, however, EUS was not performed.

The standard surgical approach for esophageal leiomyoma is thoracotomy and enucleation without opening the mucosa. For GEL, however, the decision about whether to perform enucleation or esophagectomy is dependent on the surgeon's experience, the size and shape of the tumor, and the extent of adhesions between the tumor and the esophageal mucosa [11]. Esophageal resection may be necessary for several reasons, such as if mass is very large, the adhesions between the tumor and esophageal mucosa are extensive and tight, or there are irreparable tears in the mucosa [4,12]. Virgilio and colleagues recommend esophagectomy for any circumferential or horseshoe-shaped tumors [13]. In the present case, enucleation was attempted through a right posterolateral thoracotomy. Prior to surgery, however, we had anticipated a need for esophagectomy because of the horseshoe shape of the tumor, which was confirmed when we found multiple tears in the mucosa after enucleating the tumor.

Recently, video-assisted thoracoscopy has been used successfully by some centers for either enucleation of GEL or esophagectomy [14,15]. The size of a leiomyoma should not be the factor that determines which approach to perform. Open thoracotomy and minimally invasive surgery have been used to manage esophageal leiomyomas measuring from 8 cm to 17

cm and from 9 cm to 22.5 cm, respectively. The experience and technique of the surgeon play a very important role in determining the surgical approach [16].

Large tumors are believed to be more likely to harbor malignancy than smaller ones because of the established positive correlation between tumor size and number of genetic mutations [17]. In a study in which 7 cases of GEL were managed with partial esophagectomy and gastric pull-up, the authors concluded that partial esophagectomy was necessary to distinguish between benign and malignant neoplasms and should be considered when managing patients with GEL [12]. However, malignant transformation from leiomyoma to leiomyosarcoma reportedly is extremely rare, occurring in <0.2% of cases [18].

Several reconstruction techniques have been described in the literature, with gastric pull-up and colonic interposition being the most common. There is no consensus about the ideal method of reconstruction after esophagectomy secondary to GEL. The choice of esophageal conduit is largely dependent on the location of the lesion. Gastric pull-up surgery is widely accepted because the stomach is a well-vascularized organ and requires a single anastomosis. Reflux esophagitis, dumping syndrome, and stricture formation are known post-operative complications of gastric pull-up surgery [4]. To reduce the rate of gastroesophageal reflux after gastroplasty, it is advisable to leave >6 cm of the gastric tube below the diaphragm. However, some surgeons prefer to use the colon as an esophageal conduit instead of the stomach to avoid complications, especially for tumors located in the upper third of the esophagus [12]. Therefore, a multidisciplinary, patient-tailored approach, with consideration of the surgeon's expertise, is a necessity when managing similar cases. The prognosis is overwhelmingly favorable, and most patients with GEL recover completely with no problems or recurrences [2].

Conclusions

GEL is a rare oncological entity that presents several diagnostic and therapeutic challenges. Because of the rarity of this condition, studies in the literature addressing its surgical management are scarce. The aim of the present paper was to contribute to this limited documentation by reporting our experience managing a case of giant leiomyoma of the esophagus with an Ivor-Lewis esophagectomy.

Declaration of Figures' Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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