A Giant Upper Esophageal Leiomyoma: A Rare Case

BEF Wafa Hasan
AD Umesh Nabar
AD Jalila Adnan

Patient: Female, 55-year-old
Final Diagnosis: Esophageal leiomyoma
Symptoms: Cough
Medication: —
Clinical Procedure: —
Specialty: Surgery

Objective: Rare disease
Background: Benign esophageal neoplasms are uncommon, representing less than 10% of esophageal tumors. Esophageal leiomyomas are benign mesenchymal esophageal tumors, typically involving the distal two-thirds of the esophagus. However, upper esophageal leiomyoma is unusual. Signs and symptoms are nonspecific and similar to any esophageal lesion. The role of radiologic imaging in the diagnosis of leiomyoma is vital. Enucleation is the preferred surgical method for treatment of esophageal leiomyoma. In our patient, the presence of a large tumor necessitated esophageal resection and reconstruction.

Case Report: A previously healthy 55-year-old woman presented with a chronic productive cough, which did not improve with symptomatic treatment and was not associated with shortness of breath, fever, anorexia, or weight loss. A chest radiograph revealed a superiorly located posterior mediastinal mass, measuring 9.6×4.8×4.6 cm in maximum dimensions. The mass was further evaluated by contrast-enhanced computed tomography, which demonstrated a mass with stippled calcifications, consistent with esophageal leiomyoma. Surgical resection was successfully performed, and the patient had complete resolution of her symptoms. A follow-up visit revealed a postoperative complication of left vocal cord palsy. Speech-language therapy was recommended.

Conclusions: Esophageal leiomyoma can present solely with respiratory symptoms without any gastrointestinal involvement and can be misdiagnosed because of atypical symptoms, uncommon location, or atypical imaging features. Recognizing the imaging features and pathologic basis of esophageal leiomyoma is essential for detection, early diagnosis, and management. Endoscopic and radiologic tests are critical to differentiate esophageal leiomyoma from other esophageal lesions and to eliminate the risk of malignancy.

Keywords: Case Reports • Esophageal Neoplasms • Leiomyoma • Mediastinal Neoplasms

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/932430
Background

Benign esophageal neoplasms are uncommon and represent less than 10% of esophageal tumors [1]. Despite its rarity, esophageal leiomyoma is considered the most common benign esophageal tumor. This tumor is composed of a varying proportion of smooth muscle and fibrous tissue. It usually occurs in middle-aged patients and is twice as common in men [2]. The size of esophageal leiomyoma is variable but is usually less than 3 cm. Giant esophageal leiomyoma with a size greater than 10 cm is very rare [2,3]. Most patients are asymptomatic, and esophageal leiomyoma is discovered incidentally. Symptoms like dysphagia, retrosternal pain, and regurgitation are usually associated with large tumors [4].

The typical location for esophageal leiomyoma includes the distal two-thirds of the esophagus. However, involvement of the esophagus above the level of the aortic arch is unusual [2]. Herein, we present the case of a middle-aged woman with a giant upper esophageal leiomyoma presenting with chronic cough.

Case Report

We present the case of a 55-year-old woman who presented with a history of cough for 3 months. Her cough was productive of yellow sputum and did not contain any blood. She had not identified any aggravating or relieving factors of the cough. The patient used over-the-counter symptomatic treatments, which did not result in any improvement. The cough was not associated with chest pain, shortness of breath, fever, anorexia, or weight loss. Otherwise, the patient was healthy and there was no history of respiratory diseases, including asthma or allergies. There was no history of recent travel. She never smoked or drank alcohol. Her family and social history were unremarkable.

On examination, the vital signs were within normal limits. A chest examination revealed a bilateral vesicular breathing sound with no crepitations or wheezes. Auscultation of the precordium showed normal heart sounds with no murmurs. Additionally, basic laboratory investigations, including renal and hepatic profiles, were within the reference range.

Plain chest radiograph revealed a well-defined, oval-shaped soft-tissue mass located posteriorly in the superior mediastinum, causing narrowing and displacement of the trachea. The mass measured 9.6×4.8×4.6 cm in its maximum dimensions. Stippled calcifications were also noted within the mass (Figure 1).

Subsequently, the patient underwent an upper gastrointestinal study, which demonstrated a marked deviation of the upper thoracic esophagus. The narrowing caused by this lobulated mass, however, was minimal (Figure 2). Contrast-enhanced computed tomography (CT) confirmed the presence of a large soft-tissue mass in the posterior superior mediastinum extending from the level of C7 to T5 and arising from the esophagus. The trachea and major vessels were displaced anteriorly. The mass was abutting the medial wall of the aortic arch with no signs of invasion. Stippled intralesional calcifications were suggestive of leiomyoma (Figure 3).

Figure 1. Frontal and lateral chest radiograph shows a large superior posterior mediastinal mass.
In light of the aforementioned clinical and laboratory findings, the patient was scheduled for an Ivor Lewis procedure for esophageal resection. The surgery was performed without immediate postoperative complications. A histopathological examination revealed a well-encapsulated mass composed of interlacing fascicles of smooth muscle with focal whorl formation. Foci of cystic degeneration and calcification were noted in the mass. No active mitotic activity was noted. These findings were consistent with esophageal leiomyoma.

The patient was discharged on the fifth day after surgery. In the follow-up visits, the patient had complete resolution of her previous symptoms, but she developed a new symptom of hoarseness, which was attributed to left vocal cord palsy. She was referred to speech-language therapy.

**Discussion**

We present a rare case of a giant leiomyoma of the upper esophagus presenting with chronic cough. Most esophageal tumors are asymptomatic and discovered incidentally. Clinical symptoms arise when the tumor is large enough to compress the adjacent structures, causing dysphagia, retrosternal chest pain, or regurgitation. Unlike gastric leiomyoma, upper gastrointestinal bleeding rarely occurs [4]. Interestingly, in the present case, the patient presented solely with a chronic cough and with no history of gastrointestinal involvement.

While the present case included a solitary esophageal leiomyoma, it should be noted that 3% to 4% of patients have multiple leiomyomas [5]. Additionally, gynecologic leiomyomas can co-exist in certain genetic syndromes [3]. Radiologic imaging is vital in establishing the diagnosis, since some findings can be characteristic for leiomyoma. As in the present case, chest radiography can show a posterior mediastinal mass [6]. Upper gastrointestinal studies, the most frequently performed investigations for the evaluation of esophageal lesions, typically demonstrate a lesion with a smooth surface and normal mucosa, with upper and lower borders of the lesion forming acute or slightly obtuse angles with an adjacent esophageal wall [3].
Figure 3. A large soft-tissue attenuation mass arises from the upper third esophagus. Punctate calcifications are noted. The mass displaces the trachea and major vessels anteriorly. There are no signs of invasion.
In CT images, the leiomyoma appears as a homogenous mass in the esophageal wall, with no signs of invasion. Although differentiation from other esophageal tumors is difficult, the presence of calcifications is virtually pathognomonic of leiomyoma [3]. Regarding magnetic resonance imaging, the tumor has a signal intensity similar to or slightly higher than that of the chest wall on T2-weighted images. The tumor exhibits a homogenous enhancement [6].

Endoscopic ultrasound demonstrates high accuracy in the diagnosis of esophageal leiomyomas exhibiting a hypoechoic and homogenous mass with sharply demarcated margins in the muscular layer [7].

Gross pathological examination shows a firm, round, pink-tan, well-circumscribed, or lobulated mass with no necrosis or ulceration and a whorled appearance [3]. Histology and immunohistochemistry studies are important for definitive diagnosis [2]. Histologically, leiomyomas comprise bundles of interlacing smooth-muscle cells, well-demarcated by adjacent tissue or by a definitive connective-tissue capsule [8]. Leiomyoma is distinguished from esophageal gastrointestinal stromal tumor by negative CD117 and CD34 activity on immunohistochemistry [9].

Enucleation via thoracotomy or thoracoscopy is the preferred surgical method for treatment of esophageal leiomyoma, depending on the location and size of the tumor. In contrast, surgery is favored for giant esophageal leiomyoma esophageal resection and reconstruction [10]. Since malignant degeneration is exceedingly rare [5], enucleation or esophageal resection is advised for large or symptomatic tumors. However, surgical treatment of small, asymptomatic leiomyomas is controversial, as some authors recommend resection to rule out malignancy, while others advocate for observation [10].

Conclusions

Giant esophageal leiomyoma is a very rare tumor, particularly if it involves the upper thoracic esophagus. The tumor can present solely with respiratory symptoms without any gastrointestinal involvement. Esophageal leiomyomas can be misdiagnosed because of atypical symptoms, uncommon location, or atypical imaging features. Recognizing the imaging features and pathologic basis of esophageal leiomyoma is essential for their detection, early diagnosis, and management. The use of both endoscopic and radiologic tests is critical to differentiate esophageal leiomyoma from other esophageal lesions and to eliminate the risk of malignancy.

Department and Institution Where Work Was Done

Department of Radiology, Salmaniya Medical Complex, Manama, Bahrain.

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.