

Giant esophageal leiomyoma and cardiac compression

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ABSTRACT

Esophageal leiomyoma is rare diseases, it is often asymptomatic, and typically require only close surveillance except if became large and cause symptoms secondary to compression. We report a case with giant leiomyoma presented with chest pain and unstable blood pressure secondary to cardiac compression. It is usually managed by thoracotomy and surgical enucleation and possible resection.

Key Words: Giant esophageal leiomyoma, Esophagus.

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Introduction

Unlike esophageal carcinoma, esophageal leiomyoma is rare diseases, it is often asymptomatic, and typically require only close surveillance except if became large and cause symptoms secondary to compression. If the patient becomes symptomatic or the diagnosis uncertain then surgery indicated with excellent long-term outcome. The most common presenting symptoms are dysphagia, pain, and respiratory symptoms.¹ We report a case with giant leiomyoma who presented with chest pain and unstable blood pressure secondary to cardiac compression.

Case Report

A 58 years old female was admitted in King Khalid University Hospital with a history of chest pain, shortness of breath on exertion and uncontrolled blood pressure. The patient was known to have high blood pressure, diabetes mellitus. A chest X-ray was done, showed widened mediastinum, echocardiography showed normal examination with possible mediastinal mass compression right atrium. Ct scan of the chest was done which showed large 10.6 x11.4x14.3 cm heterogeneous mixed density lesion in the posterior mediastinum, with compression to heart and extended towards the left midline. Upper endoscopy was done which showed mild external

compression of distal esophagus. Since the patient was symptomatic and the lesion looks benign in the Ct scan, the decision was made to proceed with surgery. After preoperative evaluation, surgery was performed under general anesthesia with double lumen intubation, the patient was placed in the left lateral position. The thoracoscopy was performed but because of limited space secondary to the large size of the mass, it was decided for posterolateral thoracotomy. The mass was occupying most of the lower thoracic cavity pushing the heart and carina toward the midline. Dissection was started from distal esophagus by mobilizing the inferior pulmonary ligament then moving toward the groove between the pericardium and the mass, with blunt and sharp dissection of mass from subcarinal space, the mass excised from the muscular wall of the esophagus, Intraoperative endoscopy was done which showed intact mucosa and no evidence of perforation. The mass was sent a for frozen which came as spindle cell fibromyxoid neoplasm. Chest tube inserted and wound closed. The patient was sent to the recovery room in good condition. The patient recovered very well after surgery and was discharged in post-operative day five. Final pathology confirmed the diagnosis of leiomyoma which was arising from the esophagus. Immunohisto-chemical studies showed that the tumor cells were strongly positive for desmin and H-

caldemon. The tumor cells were negative for S100, neurofilament, CD34, EMA, CD117 and calretinin (Figure 2, 3)



Figure 1. CT Scan chest showing giant heterogeneous mass arising from esophagus and compressing right atrium.

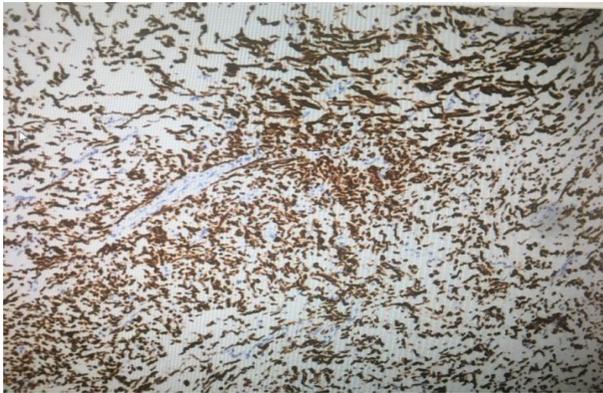


Figure 2. Histopathology of mass which was strongly positive desmin confirming the diagnosis of leiomyoma.

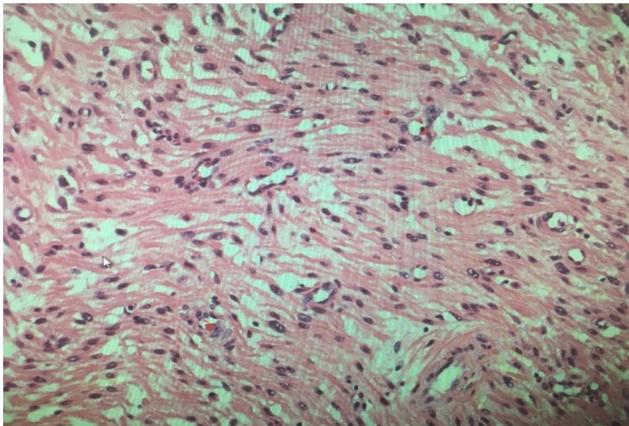


Figure 3. Histopathology of the mass showing spindle cells with multiple foci of degenerative myxoid changes consistent with leiomyoma.

Discussion

Esophageal leiomyomas is a rare esophageal benign tumor, arises from smooth muscles in the muscularis propria with incidence from 0.006 to 0.1 %.² The usual size of leiomyomas ranges from 2-5cm in size. Esophageal leiomyomas greater than 10 cm in diameter are generally described as giant leiomyomas.^{3,4} Most patients with giant leiomyoma are symptomatic. However, the common symptoms are dysphagia and chest pain. Respiratory symptoms such as dyspnea, recurrent respiratory infections, and cough can occur in about 10% of cases.⁵ Diagnosis of giant esophageal leiomyoma can be made by barium swallow, CT Chest with oral contrast, Endoscopy and Endoscopic ultrasound. Usually, a biopsy is not indicated since most of the patient symptomatic and CT imaging confirm the benign nature of the mass, it increases the risk for adhesions to the mucosa during healing and as a result, may complicate surgical enucleation, increasing the risk for violation of the mucosa at the time of resection .⁶

In the past, most of the patients with leiomyoma treated with open thoracotomy, gastro-esophagostomy or tumor enucleation. In recent years, many centers have gradually implemented minimally invasive surgeries for the treatment of esophageal leiomyoma, including resection or enucleation of esophageal leiomyoma by thoracoscopy, laparoscopy or DaVinci robot-assisted thoracoscopy.⁷⁻⁸ Those surgical approaches are ideal for small leiomyoma less than 5 cm in size. However, giant esophageal leiomyoma usually treated with Thoracotomy and enucleation or sometimes with esophageal resection.^{9, 10}

Conclusion

Giant esophageal leiomyoma is a very rare benign esophageal tumor that is usually symptomatic and presented with Dysphagia, respiratory and cardiac similar symptoms secondary to direct compression. The tumor usually managed by thoracotomy and surgical enucleation and possible resection.

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