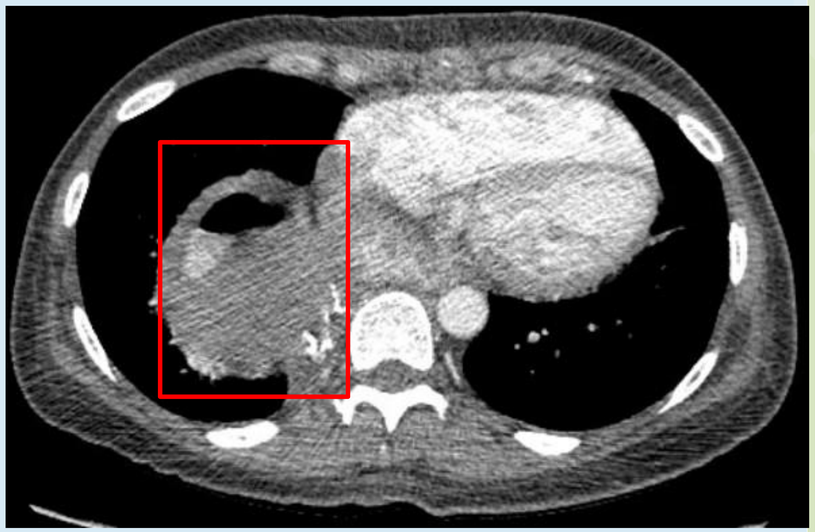
Leiomyomatosis presenting as Achalasia

Iman Waheed Khan, MD; Hira Hanif, MD; Shefali Amin, DO; Jesus M Salis Noain MD;
John Altomare, MD; Anthony Donato, MD, Nirav Shah, MD.
Reading Hospital, TowerHealth

Reading Hospital
TOWER HEALTH
Advancing Health. Transforming Lives.

INTRODUCTION

Leiomyomas represent less than 1 % of esophageal neoplasms yet constitute 67% of the benign esophageal tumors. They typically present as slow-growing espohageal masses causing slowly progressive dysphagia, however giant leiomyomas can cause more rapid symptoms. The mainstay of treatment for leiomyomas is resection or enucleation by conventional thoracotomy or thoracoscopy. However, for giant leiomyomas of esophagus, esophagectomy remains an option..



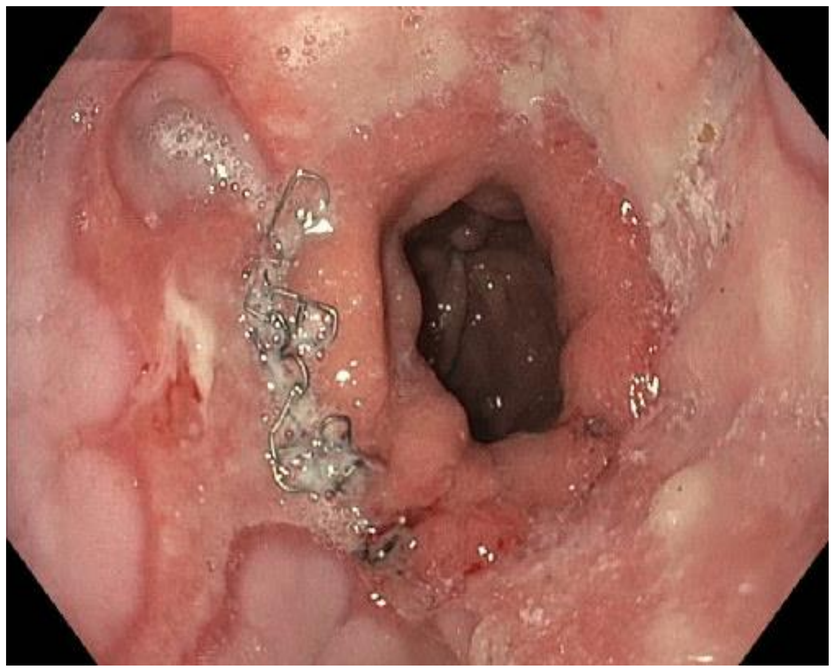
CT of the Chest highlighting marked wall thickening around GEJ/distal esophagus as well diffuse wall thickening of thoracic esophagus. Single wall measured up to 3 cm in thickness, with severe luminal narrowing and moderate dilatation proximal to it.

CASE DESCRIPTION

We present the case of a 37-year-old female with a past medical history of long-standing dysphagia reportedly diagnosed with achalasia on endoscopy 10 years ago in Barbados who presented with progressively worsening dysphagia and acid reflux. When she was diagnosed with achalasia 10 years ago it was recommended that she be managed conservatively and hence she did not undergo any definitive treatment. Over the past several years she had progressive worsening of her dysphagia to both solids and liquids and worsening acid reflux despite adhering to a soft and liquid diet, so she decided to come to the US for further evaluation. Upper endoscopy revealed a significantly dilated and tortuous distal esophagus, with tight and firm gastroesophageal junction (GEJ) and grade 4 candida esophagitis. A CT scan of the chest showed moderately dilated esophagus with severely thickened distal esophageal wall and diffuse wall thickening of the thoracic esophagus. These findings were consistent with achalasia and she was evaluated by cardiothoracic surgery and deemed an appropriate candidate for Heller myotomy with Dor fundoplication. During her procedure, it was found that the distal 15 cm of esophagus was invested with a massive circumferential ill-defined mass with a wall thickness of more than 3 cm. The procedure was converted to open distal esophagectomy with gastroesophageal anastomosis with feeding jejunostomy. Biopsy was consistent with leiomyomatosis. On her one-month post op follow up her swallowing had improved significantly, she was tolerating the general diet and the jejunal tube was removed.

DISCUSSION

Delayed diagnosis can lead to slow , then sometimes rapid growth of leiomyomas. Early intervention can allay the need for more extensive surgery as was needed here. Patients presenting with progressive dysphagia to solids must undergo endoscopic evaluation to rule out tumor or malignancy. If investigative techniques are inconclusive, CT scan and endoscopic ultrasound (EUS) should be considered as both can identify a mass arising within the esophageal wall.



EGD post distal esophagectomy with gastroesophageal anastomosis