

Case Report

GIANT ESOPHAGEAL LEIOMYOMA WITH MEGAESOPHAGUS A CASE REPORT

M.R. Fattahi,* A. Izadpanah,** P.V. Kumar***

Departments of *Internal Medicine, **Surgery and ***Pathology, Shiraz University of
Medical Sciences, Shiraz

ABSTRACT

A 48-year-old man with a giant leiomyoma of the esophagus accompanied by megaesophagus is reported in this paper. He presented with dysphagia and was treated successfully by means of an esophagegogastric resection. The literature is reviewed and the clinical and pathological aspects of this rare tumor are discussed.

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Key Words • Esophagus • leiomyoma • megaesophagus

Introduction

In comparison with stomach and intestine, benign esophageal tumors including hemangioma, papilloma, fibroma, schwannoma and leiomyoma are very rare. Leiomyoma is the most common benign tumor of the esophagus¹ and is seen about once in every 100 patients diagnosed as esophageal cancer.¹ Although a number of cases have been reported in the literature,^{1,9} only very few cases of esophageal leiomyoma are associated with megaesophagus. Herein, we report a second case that developed a secondary megaesophagus.

Case Report

A 48-year-old man was referred to Nemazi Hospital with the chief complaint of dysphagia of eight months duration. It was initially associated with solid foods and later with

liquid diets. He had a 7 Kg weight loss over the past six months. Physical examination revealed no abnormality. There was no organomegaly or lymphadenopathy. His Hb was 14.8gr%, ESR = 28 and other routine lab data were within normal range. Endoscopy was performed with difficult passage through the lower esophagus owing to a large protruding mass with smooth surface in the distal end of the esophagus. The mucosa was intact. There was no esophagitis or bleeding. Esophagography revealed a tortuous and dilated esophagus with lower esophageal obstruction (Fig. 1).

Clinically, a benign tumor was suspected and an esophagectomy by O'ringer method was performed. Gross examination revealed a huge tumor of 11 x 10 x 10 cm, in the lower esophagus with dilatation of the proximal parts (Fig. 2).

The histologic findings of the tumor revealed interlacing bundles of spindle-shaped cells of smooth muscle (Fig. 3). The diagnosis of leiomyoma was made, due to the absence of necrosis, pleomorphism and high nucleocytoplasmic ratio. The mitoses were few

Correspondence: A. Izadpanah M.D., Department of Surgery, Faghihi Hospital, P.O.Box:71345-1853, Tel/Fax: +98711-6247402, E-mail: fattahim@sums.ac.ir

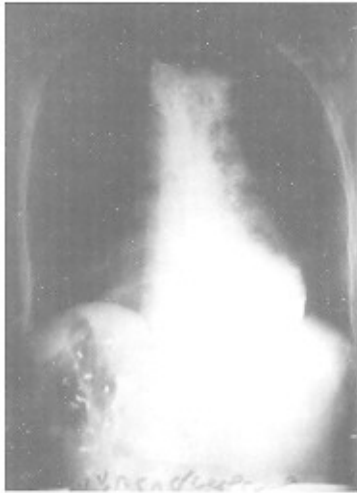


Figure 1: Esophagogram revealing severe narrowing of the distal esophagus without mucosal irregularity. The site of narrowing is compatible with that of the mass in the distal part of esophagus simulating achalasia.

in number and ranged from about 1-2 per 10 high power field. Immunohistochemical study revealed positive reaction to actin and desmin and negative reaction to EMA (Epithelial Membrane Antigen) and NSE (Neuron Specific Enolase). The post-operative period was uneventful and the patient was discharged.

Discussion

Benign tumors of the esophagus are rare, constituting only 0.5-0.8% of all esophageal neoplasms.¹ Approximately 60% of benign tumors of the esophagus are leiomyomas.^{10,11} They vary greatly in size, but seldom cause symptom when less than 5 cm in diameter.¹² Larger tumors cause dysphagia, vague retrosternal pressure and pain. Most reported leiomyomas have been asymptomatic and found incidentally at autopsy.

Giant leiomyomas are rarely reported, moreover association with megaesophagus has only been reported in one previous case.¹

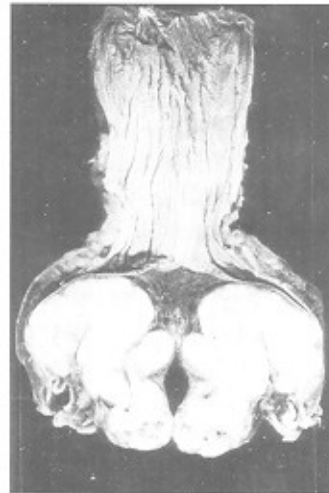


Figure 2: A well defined grayish white lobulated 11×10×10 cm tumor at the lower part of the esophagus.

Giant esophageal leiomyomas present a diagnostic and therapeutic challenge because of their size and the possibility of malignant behavior. The lower third of the esophagus is the most common site with the middle third being the next most commonly affected area.¹⁰ Esophageal leiomyomas more than 1000 grams are also rare.^{3,4} Several esophageal diseases, such as cancer, scleroderma, and peptic esophagitis and achalasia can produce varying

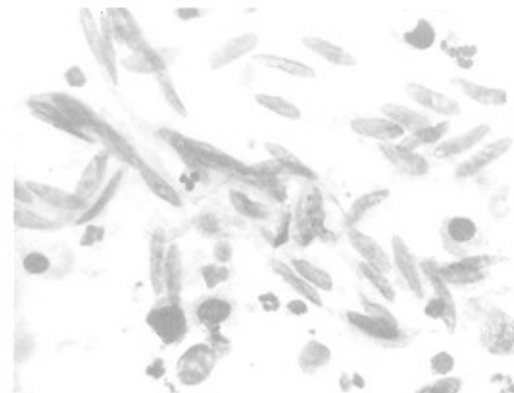


Figure 3: Microscopic examination revealing a cellular tumor composed of interlacing bundles of spindle-shaped cells without mitoses. (H&E×1200)

degrees of esophageal dilatation.¹ But a megaesophagus secondary to a leiomyoma is rare.¹ These tumors have no clear-cut gender preponderance¹² and are usually seen in patients with an age range of 20 to 70 years.¹¹ They may be multiple in 3% to 10% of patients.¹² Dysphagia is the main symptom and is seen in more than 50% of cases.¹²

Leiomyomas of the esophagus mainly grow intramurally. They may grow eccentrically toward the posterior mediastinum and occasionally they grow into the esophageal lumen.^{1,5} Radiographic studies are the main diagnostic tool as they also assist in appropriate treatment plan.¹¹ Occasionally, these tumors are found in plain chest X-ray.¹¹ Endoscopy is useful in the diagnosis of leiomyoma and to exclude the possibility of esophageal cancer.

Leiomyomas must be treated surgically, either enucleation (in cases of small tumors) or segmental resection (in cases of larger tumors). Histologically, these tumors may be mistaken for neurogenic tumors due to the presence of spindle shaped cells. However, the absence of Verocay bodies and palisading cells confirms the diagnosis of leiomyoma. Due to the lack of mitoses, necrosis and pleomorphism, we have ruled out the possibility of sarcoma in this case.

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