Giant leiomyoma of the esophagus

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Abstract

Leiomyoma is the most common benign tumour found in the esophagus but it is, however, a rare neoplasm; in fact of all esophageal tumours, benign tumours account for fewer than 10%, of which 4% are leiomyomas. Leiomyomas should be removed when diagnosed, even if asymptomatic, because malignancy cannot otherwise be excluded and symptoms are likely to develop if treatment is delayed or omitted. Enucleation of esophageal leiomyoma is a safe and effective procedure. We report a case of symptomatic giant anular leiomyoma of the distal esophagus, compressing the trachea and the descending aorta, resected after right thoracotomy. © 2002 Elsevier Science B.V. All rights reserved.

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1. Introduction

Leiomyoma is the most common benign tumour of the esophagus. It is, however, a rare condition with an overall incidence of 8–43/10000 in autopsy series [1,2]. It is found mainly in the lower and middle thirds of the esophagus and, in most cases is a single lesion. About half the patients with leiomyoma are asymptomatic; when symptoms do occur they comprise dysphagia, unspecific retrosternal pain, heartburn and, occasionally, weight loss [3]. Diagnosis is usually made on esophagogram, chest X-ray, computerized tomography (CT) and magnetic resonance (MR) scan. Recently transesophageal echo probe has been widely used for preoperative diagnosis [4]. Leiomyomas should be removed when diagnosed, even if asymptomatic because malignancy cannot otherwise be excluded and symptoms are likely to develop if treatment is delayed or omitted [1].

We describe a case of symptomatic giant anular leiomyoma of the distal esophagus, compressing the trachea and the descending aorta, resected after right thoracotomy.

2. Case report

A 42-year-old man was admitted to our department because of a posterior mediastinal mass disclosed by chest X-ray for acute bronchitis.

Esophagogram showed a stricture of the distal thoracic esophagus which was compressed and displaced to the left side.

Chest CT scan confirmed the posterior mediastinal paraesophageal ovoidal mass, 15 cm in diameter, extending to the diaphragm (Fig. 1).

This mass caused hypoventilation and atelectasias of the right lower pulmonary lobe, because of compression and displaced the inferior supradiaphragmatic vena cava, inferior right pulmonary vein and descending aorta without infiltration. Esophagoscopy was negative; preoperative bronchoscopy was not performed because of patient’s refusal; we performed intraoperative bronchoscopy, confirming tracheal compression. Transthoracic CT guided fine needle aspiration biopsy was suggestive for leiomyoma; spirometry was normal (FEV1 = 3.75–92% pred.; FVC = 4.88–98% pred.; FEV1/FVC = 77)

Right thoracotomy was performed; the lower pulmonary lobe and lower vein were uplifted by the esophageal tumor which was separated from the lung. As the neoplasm involved the esophagus circumference, it was necessary to dissect the lesion in three parts to allow resection. It was also necessary to open the esophageal wall which was then sutured by simple interrupted stitches and protected by a pedunculate pleural flap; no part of the esophagus was excised; frozen section excluded malignancy.

Histologic examinations revealed fibroleiomyomatous tissue (9 × 15 cm) with cellular areas and coagulative necrosis (<10%), without cellular anaplasia; mitosis was <1 × 10 HPF. The immunohistochemical algorithm was: smooth muscle actina + , CD 34 − , C-Kit − , proliferation

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index/Mib 1:1–2%. The immunomorphologic examination revealed esophageal leiomyoma (Fig. 2).

An esophagogram performed 6 days after surgery showed no motor alterations and the patient was discharged 10 days after surgery.

3. Discussion

Leiomyoma is the most common benign tumour found in the esophagus but is nonetheless rare. Of all esophageal tumours, benign tumours account for fewer than 10%, of which 4% are leiomyomas [1,3]. These tumours are usually found in middle age and are almost twice as common in men; they are located mainly in the lower and middle thirds of the esophagus and, in the majority of cases, are single lesions [1]. They may undergo cystic degeneration; calcification occurs infrequently and malignant change is rare. However, malignant degeneration or leiomyoma concomitant with malignant neoplasms have been described [5,6].

Most leiomyoma are intramural but some present as intraluminal pedunculate polyps in or adjacent to esophageal diverticula [7,8]. About half the patients affected by leiomyoma are asymptomatic; when symptoms do occur, they are fairly long duration and include retrosternal pain and dysphagia. Bleeding may also occur when the mucosa over the tumour becomes ulcerated [1].

Esophageal leiomyoma appears on chest X-ray as a posterior mediastinal mass; esophagography, in which the tumour is seen as intramural, eccentric smoothly elevated, sessile lesion varying in size, is the most reliable form of diagnosis. Even where contrast medium is administered, CT
reveals a lesion which demonstrates homogeneous low or iso attenuation, while T2-weighted MR imaging displays a slightly hyperintense lesion [2]. Recently endoscopic ultrasonography has been used in the evaluation of leiomyoma of the esophagus and is very accurate in visualizing this lesion and differentiating cystic from solid submucosal esophageal masses. In addition, the test can establish the exact location of the mass in relation to the esophageal wall and mediastinum.

One hand, according to Hennessy et al. [1], leiomyoma should be removed when diagnosed, even if asymptomatic, because malignancy cannot otherwise be excluded and symptoms are likely to develop if treatment is delayed or omitted; on the other hand, according to Pearson et al. [9], although malignancy can only be ruled out by excision and histologic examination, it seems unreasonable to remove all small asymptomatic leiomyomas.

The affected segment of the esophagus is exposed at thoracotomy; the stretched fibres of esophageal muscle over the tumour are incised and the tumour enucleated. It is seldom necessary to open the esophageal lumen; the muscular wall is re-sutured after tumour resection. Sometimes after removal of a large tumour a significant muscular defect remains. This can be left without causing concern provided the esophageal mucosa is intact [1]. Enucleation of esophageal leiomyoma is a safe and effective operation. The video assisted thoracoscopic approach, combined with intraoperative esophagoscopy, when possible, facilitates the procedure with the added advantage of shortening hospital stay. The muscle layer of the esophagus should be approximated to avoid decreasing the propulsive activity of the esophageal body. This may improve the long-term outcome of the operation by preserving the acid-clearing mechanism of the esophagus and reducing the incidence of postoperative reflux esophagitis [10].

References