Leiomyoma of Esophagus

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Leiomyomas are rare benign esophageal neoplasms with an indolent clinical course. Symptoms mimic that of esophageal cancer. Esophagoscopy and endoscopic ultrasonography are the main diagnostic methods. Symptomatic and large leiomyomas should be treated surgically while small, asymptomatic lesions may be managed by regular follow up and repeated endoscopies. (Ann Thorac Cardiovasc Surg 2007; 13: 78–81)

Key words: leiomyoma, esophagus

Introduction

Benign tumors of the esophagus are rare lesions that constitute less than 1% of esophageal neoplasms. Nearly two thirds of benign esophageal tumors are leiomyomas; the others are mostly polyps and cysts.1,2) Leiomyomas are the commonest benign mesenchymal tumors of the esophagus. Most tumors described as leiomyomas and leiomyosarcomas in the older medical literature actually refer to gastrointestinal stromal tumors (GISTs).3) Leiomyoma is a term used only in relation to tumors of the esophagus. The significance of knowing about leiomyomas is due to the fact that it can mimic esophageal cancer and lead to diagnostic confusion. In this article, we review our experience with esophageal leiomyomas at our institute.

Patients and Methods

We retrospectively reviewed the medical records of patients diagnosed with esophageal tumors at the Tata Memorial Hospital from January 2003 to January 2005. About 1,500 patients were registered with esophageal tumor during the study period. Of these, 290 patients underwent some types of esophageal surgery. We found six patients with esophageal leiomyomas. The esophageal leiomyomas thus constituted 0.4% of all the esophageal tumors in this study. We recorded the presenting symptoms, investigations, management and outcome of these patients. The diagnostic workup included barium swallow, upper gastrointestinal endoscopy (UGI scopy), endoscopic ultrasound (EUS) and computed tomography (CT). Fine needle aspiration cytology (FNAC) or biopsy was done in cases where there was suspicion of malignancy. Of the six patients, we had histological diagnosis of leiomyoma in five patients. The sixth patient had classical features of leiomyoma on EUS and CT scan and was planned for regular follow up. We had no tissue diagnosis for this patient.

Results

Six patients were diagnosed with esophageal leiomyomas of which five were female. Their age [TK1] ranged from 34 to 65 years. Of the six patients, tissue diagnosis of leiomyoma (Fig. 1) was available for five patients. Dysphagia was the commonest presentation though nonulcer dyspepsia, retrosternal burning and epigastric pain were associated. One patient had symptoms of dyspnoea, cough and hoarseness related to a large mass (Figs. 2 and 3). Table 1 shows the clinical presentation and the details of the management of all the patients. Three patients required esophageal resection, while two patients underwent lo-
Leiomyoma of Esophagus


cal enucleation and one patient with a small, relatively asymptomatic leiomyoma was kept under close observation. All the operated patients had uneventful postoperative recoveries. One patient who had undergone total tran-

Discussion

Leiomyomas are the commonest benign mesenchymal tumors of the esophagus contributing about two-thirds of all benign lesions of the esophagus. Leiomyomas usually arise as intramural growths, most commonly along the distal two thirds of the esophagus. They are multiple in approximately 5% of patients.1,2,4,5)

Esophageal leiomyomas rarely cause symptoms when they are smaller than 5 cm in diameter. Large tumors can cause dysphagia, vague retrosternal discomfort, chest pain, esophageal obstruction, and regurgitation. Rarely, they can cause gastrointestinal bleeding, with erosion through the mucosa. Other than the nonspecific symptoms associated with esophageal leiomyomas, very few physical findings are usually noted. In extremely rare cases where severe esophageal obstruction is caused by a leiomyoma, weight loss and muscle wasting may be observed.6)

Histologically, leiomyomas comprise of bundles of interlacing smooth muscle cells, well-demarcated by adjacent tissue or by a definitive connective tissue capsule.

Fig. 1. Esophageal leiomyoma showing a submucosal fasciculated spindle cell tumor with cigar-shaped nuclei. (H&E × 200)

Fig. 2. Barium swallow showing smooth convex mass and distinct margins at the junction of mass and normal mucosa.

Fig. 3. Computed tomography (transverse section) of one of our patients showing large leiomyoma in the esophagus occupying the posterior mediastinum.

sthoracic esophagectomy (Fig. 4) had a minor anastomotic leak in the neck which healed with conservative manage-
ment. All patients are asymptomatic and recurrence-free at last follow up.
Table 1. Clinical presentation and management of patients

<table>
<thead>
<tr>
<th>Age/Sex</th>
<th>Symptoms</th>
<th>UGI scopy</th>
<th>EUS</th>
<th>CT scan</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>55/F</td>
<td>Dysphagia Gr III, dyspnoea, hoarseness, cough, mass in lower neck</td>
<td>Extrinsic compression on left side from cricopharynx to 24 cm</td>
<td>Not done</td>
<td>Well-defined mass in tracheo-esophageal groove extending from left lobe of thyroid to aortopulmonary window, size: 9×5 cm</td>
<td>Left thoracotomy-TTE</td>
</tr>
<tr>
<td>34/F</td>
<td>Dysphagia Gr II</td>
<td>Submucosal tumor 22–27 cm</td>
<td>Well-defined hypoechoic tumor with calcification</td>
<td>Well-defined soft tissue mass in M/3 esophagus causing obliteration of lumen with proximal dilatation, size: 5×3 cm</td>
<td>Right thoracotomy-myotomy with enucleation</td>
</tr>
<tr>
<td>65/F</td>
<td>Dysphagia Gr II</td>
<td>Submucosal lesion at 22–28 cm</td>
<td>Large encapsulated tumor pushing tracheobronchial tree</td>
<td>Mass in M/3 esophagus with luminal narrowing and proximal dilatation, size: 6×3.5 cm</td>
<td>THE</td>
</tr>
<tr>
<td>62/M</td>
<td>Nonulcer dyspepsia, Dysphagia Gr I</td>
<td>Submucosal tumor at cardia and hiatal sac with ulceration</td>
<td>Not done</td>
<td>Mass in L/3 esophagus, size: 6×3 cm</td>
<td>Left thoraco-abdominal approach—Extramucosal excision</td>
</tr>
<tr>
<td>48/F</td>
<td>Dysphagia Gr III, epigastric pain</td>
<td>Submucosal nodule</td>
<td>Submucosal nodule</td>
<td>Mass in L/3 esophagus involving posterior wall, size: 5×3 cm</td>
<td>Esophagogastrectomy</td>
</tr>
<tr>
<td>40/F</td>
<td>Retrosternal pain related to meal</td>
<td>Polypoidal submucosal lesion at 30 cm</td>
<td>Well-defined encapsulated lesion from muscularis mucosa</td>
<td>Mass lesion in lower esophagus, size: 3×3 cm</td>
<td>Observation with regular endoscopy</td>
</tr>
</tbody>
</table>

TTE, total transthoracic esophagectomy; THE, transhiatal esophagectomy; UGI scopy, upper gastrointestinal endoscopy; EUS, endoscopic ultrasound; CT, computed tomography.

They are composed of fascicles of spindle cells that tend to intersect with each other at varying angles. The tumor cells have blunt-ended elongated nuclei and show minimal atypia with few mitotic figures.7)

On barium swallow, the classic appearance is a smooth concave mass underlying the intact mucosa. Distinct sharp angles are seen at the junction of the tumor and normal tissue. At endoscopy, the lesions are identified as mobile submucosal masses. If a leiomyoma is suspected at esophagoscopy, a biopsy should not be performed as it would cause scarring at the biopsy site, which would hamper definitive extramucosal resection at surgery.4) However, an ulcerated growth should be biopsied to rule out malignancy. Endoesophageal ultrasonography is important in the diagnosis of leiomyomas, which demonstrate a homogeneous region of hypoechogeticity juxtaposed

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Fig. 4. Specimen of total esophagogastrectomy done for one of our patients showing a large leiomyoma in upper and middle third of esophagus.
with the overlying mucosa.8,9) Asymptomatic or smaller leiomyomas may be followed up periodically with regular barium swallow and endoscopy as they have a characteristic radiographic appearance, slow growth rate, and negligible risk of malignant transformation.2) Resection may be required to confirm the histopathological diagnosis in some cases. Surgical excision is recommended for symptomatic leiomyomas and those greater than 5 cm. Although a formal esophageal resection is not mandatory for leiomyomas. Frequently the large size and extent of the tumor (as in three of our patients) may preclude local enucleation.

Tumors of the middle third of the esophagus may be approached using a right thoracotomy. Tumors in the distal third of the esophagus may be resected through a left thoracoabdominal approach, transhiatally or by a left thoracotomy. For extramucosal excision or enucleation, the outer esophageal muscle is incised longitudinally. Careful dissection is done to separate and remove the leiomyoma from the underlying submucosa. If the mucosa is inadvertently opened during dissection, the underlying mucosa is reapproximated, followed by closure of the longitudinal muscle.10) Some authors have shown that large extramucosal defects may be left open without subsequent complications developing.

Segmental esophageal resection (esophagogastrectomy) may be indicated for giant leiomyomas of the cardia. Total transthoracic esophagectomy (as for one patient in our series) is rarely needed especially for very large leiomyomas involving long segment of the esophagus. While open surgery is the mainstay of therapy for leiomyomas, combined esophagoscopy and video-assisted resection (thoracoscopy) have been performed recently in six patients, without complication.11)

References