Giant leiomyoma of the esophagus: A case report and review of the literature

Dev özofagus leiomiyomu: Vaka sunumu ve literatürün gözden geçirilmesi

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Although leiomyoma is the most common esophageal mesenchymal neoplasm, it is a rare condition. Resection of the tumor is recommended in symptomatic patients, and observation is recommended in asymptomatic patients with small lesions. We discuss herein a patient admitted to our outpatient clinic for dyspepsia in whom a giant annular esophageal leiomyoma was diagnosed. Resection through thoracoabdominal approach is recommended instead of adjuvant treatment in similar sized tumors.

Key words: Leiomyoma, esophagus, endoscopic ultrasonography, computerized tomography, tru-cut biopsy

Leiomiyom nadir olmasına rağmen özofagusun en sık görülen mezenkimal tümörüdür. Semptomatik hastalarda rezeksiyon, asemptomatik ve küçük lezyonu olan hastalarda izlem önerilmektedir. Bu vaka sunumunda dispeptik yakınmaları nedeniyle polikliniğe başvuran ve dev annüler özofagus leiomiyom tanısı konan hasta tartışılmıştır. Benzer boyutlu tümörlerde adjuvan tedavi yerine torakoabdominal yaklaşım ile rezeksiyon uygun gözükmektedir.

Anahtar kelimeler: Leiomiyom, özofagus, endoskopik ultrasonografi, bilgisayarlı tomografi, tru-cut biyopsi

INTRODUCTION

Leiomyoma is the most common benign tumor of the esophagus (1). Less than 10% of esophageal tumors are benign (2-5), and more than half are leiomyomas. They arise from smooth muscle cells, and the incidence at autopsy is between 0.005% and 5.1% (6, 7). Middle-aged men are most frequently affected (2, 6, 8). Most patients have a single tumor (1, 2). The size of the esophageal leiomyoma may change, but a size of 1 to 29 cm has been defined in the literature (1, 5, 9). Esophageal leiomyomas have been reported as being smaller than 5 cm in 49%, 5 to 9 cm in 33.7%, 10 to 14 cm in 12.2%, 15 to 19 cm in 2.5%, and larger than 20 cm in 2.5% of the patients reported (9). Leiomyomas larger than 10 cm are defined as giant leiomyomas (1, 4). Dysphagia, retrosternal pain, and pyrosis are the most common symptoms (1-3). No relationship has been found between symptoms and size or location of the tumor (4-7). Here we describe a giant leiomyoma of the distal esophagus with concomitant dyspeptic symptoms.

CASE REPORT

A 52-year-old man was admitted to our hospital with dyspepsia and esophageal reflux. There was no nausea, vomiting, or weight loss. Results of a physical examination and standard laboratory tests were normal. A chest radiograph showed a mass in the right hemithorax, and a filling defect was apparent on esophagography. Computerized tomography (CT) scanning of the chest and abdomen revealed a giant calcified 110 x 99 mm mass at the distal esophagus invading the cardia and fundus of the stomach (Figure 1).

An upper gastrointestinal system endoscopy showed an indentation from the mass at the cardia with a normal mucosa. Results of a colonoscopic examination were normal. Endoscopic ultrasonography (EUS) showed a mass between the carina and the cardia, 9 to 15 cm in diameter (Figure 2). A transthoracic CT-guided Tru-cut biopsy was performed, after which a mild pneumothorax developed that spontaneously healed.

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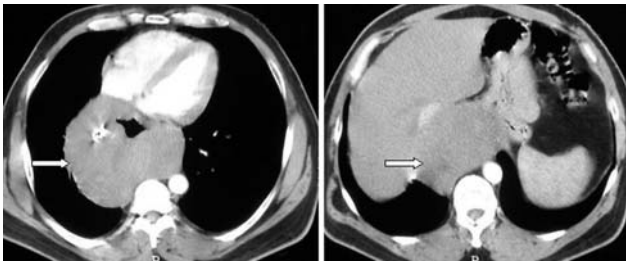

Figure 1. Computerized tomography scanning of the chest and abdomen showing a giant calcified 110 x 99 mm mass at the distal esophagus invading the cardia and fundus of the stomach (arrow).

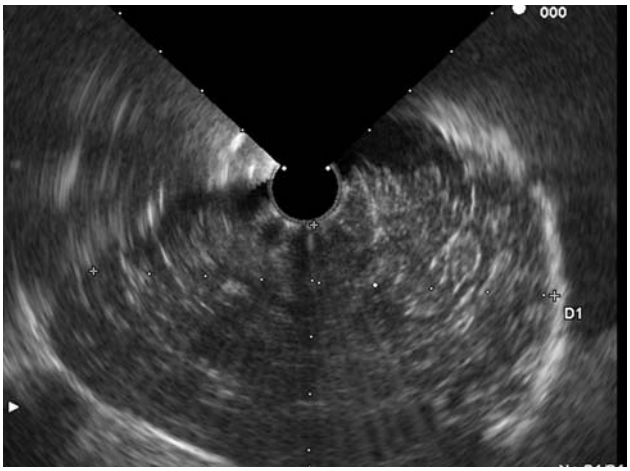

Figure 2. Endoscopic ultrasonography showing a nearly 9-15 cm mass between the carina and cardia.

Histopathologic examination revealed spindle cell proliferation, but no mitotic activity or cellular anaplasia was present. Immunohistochemical study with actin showed diffuse and strong positive staining but only a few cells stained positively with desmin.

A right thoracotomy and midline laparotomy were performed. During surgical exploration, a giant annular mass in the distal esophagus involving the cardia of the stomach was observed. The mass was resected completely with a distal esophagectomy and esophagogastrostomy. Macroscopic examination revealed a 17 x 9 x 10 cm mass weighing 1075 g (Figure 3). Microscopic examination revealed well-defined submucosal smooth muscle tissue without mitotic activity or necrosis. The patient was discharged six days after the operation. He has been followed for 25 months. A follow-up endoscopy showed mild erosive esophagitis at the anastomosis line.

DISCUSSION

Leiomyomas may occur in all parts of the esophagus, but 60% occur in the distal third, 30% in the middle, and 10% in the proximal esophagus (2, 7, 8). They are typically oval or spherical (2). These tumors often have an intramural location but some may be present near the esophageal diverticula or grow intramurally as a pedunculated polyp (1, 4). Leiomyomas grow slowly, and half of the patients are asymptomatic unless the tumor grows intramurally (1, 3). No direct correlation has been found between tumor size and symptom duration. Dysphasia is the most common symptom of leiomyomas followed by retrosternal pain (1-3, 10). Weight loss is a rare symptom, the cause of which is not clear. Although gastrointestinal bleeding is a common finding in gastric leiomyomas, esophageal leiomyomas rarely bleed, which may be because they do not ulcerate (3, 4).

Esophageal leiomyomas may present as a posterior mediastinal mass on chest radiograph (1-3) and may be seen as an incidental radiologic finding (5, 11). Barium swallow is the most commonly used radiologic test for esophageal lesions (2, 4). The finding on barium swallow is a smooth crescent-shaped filling defect in the contour of the esophageal lumen without a mucosal abnormality. The tumor is usually mobile during swallowing (3, 4).

On contrast-enhanced CT, leiomyoma shows weak homogeneous contrast enhancement and is difficult to differentiate from other esophageal tumors like neurofibromas, hemangiomas, or fibromas (1, 12, 13). Generally, T2 sequences on magnetic resonance imaging reveal isointense submucosal lesi-

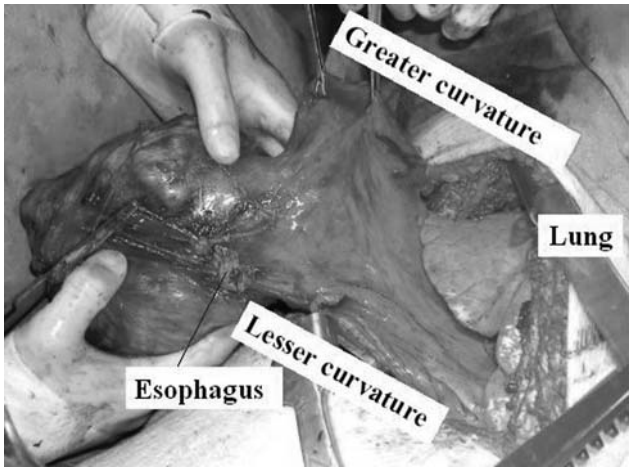

Figure 3. Intraoperative, macroscopic view of the 17 x 9 x 10 cm, 1075 g mass after resection.

ons and differ from esophageal carcinoma, in which a higher signal appears on T2 sequences (12). Esophagoscopy is also used for the differential diagnosis of esophageal leiomyoma and carcinoma. Because esophageal leiomyomas are submucosal lesions, conventional endoscopy will not lead to an accurate diagnosis (2, 14). Recent use of the EUS clearly reveals the five-layered structure of the gastrointestinal wall (14). On EUS, leiomyoma presents as a homogeneous and hypoechoic lesion with clear margins, surrounded by a hyperechoic area (14), which can easily be differentiated from a lipoma, cyst, or hemangioma in the esophageal wall. The diagnostic accuracy of EUS in gastrointestinal leiomyoma has been found to be superior to other imaging techniques (14). EUS also clearly shows the origin of the leiomyoma either as muscularis mucosa or muscularis propria. Recently, the EUS-guided Tru-cut biopsy has been used for diagnosis (15). We performed a CT-guided Tru-cut biopsy, which is not routinely used to diagnose submucosal lesions of the esophagus. Histologic and immunohistochemical examination is important in the definitive diagnosis of leiomyomas. Leiomyomas typically are strongly positive for desmin and SMA (smooth muscle actin) and are negative for CD 34 and c-kit mutations (4). In our patient, the presence of positive staining for desmin and SMA was compatible with the diagnosis of leiomyoma.

Surgical treatment of leiomyomas is controversial. However, resection of the tumor is recommended in symptomatic patients, while observation is recommended for asymptomatic patients with lesions smaller than 5 cm and when the preoperative workup has excluded malignancy (9). The surgical indications of these tumors include unremitting symptoms, increased tumor size, mucosal ulceration, histopathologic diagnosis, and facilitation of other surgical procedures (9). Malignant transformation in leiomyomas is rare, and needle aspiration biopsy usually does not accurately identify the nature of the lesion; therefore, malignancy can only be ruled out by resection. Malignant degeneration of the esophageal leiomyoma and malignant tumors occurring with leiomyomas have also been reported (16, 17). Some authors suggest that a leiomyoma should be removed when diagnosed even when asymptomatic because malignancy cannot otherwise be excluded (1, 4). However, because of the complications related to surgery, some authors do not suggest operation unless there is an evidence of malignancy (4).

The standard surgical approach is thoracotomy. The preferred surgical technique for leiomyomas is transthoracic enucleation without opening the mucosa (1, 2), which is easier, faster, and safer compared to resection (1, 4, 9). Esophageal resection is indicated for large tumors and tumors located at the gastroesophageal junction. Leiomyomas located at the proximal and middle third of the esophagus can be operated via a right thoracotomy (2). Surgical resection is performed by a transhiatal approach for tumors in the lower third of the esophagus. The video-assisted thoracoscopic approach with the intraoperative esophagoscopy is another alternative that facilitates the procedure. EUS-guided endoscopic resection, endoscopic laser ablation, and aspiration lumpectomy are less invasive (4). In our patient, because of the size and location of the tumor, it was resected using a thoracoabdominal approach, and a distal esophagectomy and esophagogastrostomy were performed.

In conclusion, diagnosis of esophageal leiomyomas requires both endoscopic and radiologic examinations. Currently, resection via thoracoabdominal approach is recommended for large tumors located at the distal esophagus.

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