Lymphangioma of esophagus presenting with dysphagia: A case report

Dharmesh H Kaswala, Jonathan Faiwiszewski, Kunar Grover, Kiran Rao, Michael Demyen

New Jersey Medical School, The University Hospital, UMDNJ, Newark, NJ, USA

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INTRODUCTION

Lymphangioma of the esophagus is a benign vascular tumor that rarely occurs in the upper gastrointestinal tract. To date, only 15 cases of this tumor have been reported in the literature. Depending on the specific histologic features, we divide lymphangiomas into three groups based on Wegener's classification: simple, cavernous and cystic. Most lymphangiomas are found in the neck (75%) and axilla (20%), with the remainder most commonly arising in the mediastinum, bone and retroperitoneum; liver and spleen involvement is rare and is associated with a worse prognosis. Herein, we report a rare case of lymphangioma of the esophagus and our experience with endoscopic resection of this lesion.

CASE REPORT

A 61-year-old Hispanic man with a past medical history of gastroesophageal reflux disease was referred for evaluation of a polypoid lesion in the mid-esophagus, which was noted on prior endoscopic exam for dysphagia to solids and liquids. An EGD (Esophagogastroduodenoscopy) done by his gastroenterologist a year earlier demonstrated what appeared to be a submucosal polypoid lesion in the mid-esophagus. Previous biopsies were non-diagnostic, showing inflammatory mucosa only. The patient denied any weight loss/blood in stool, chest pain, fevers, hemoptysis, nausea, vomiting, diarrhea or hematochezia. There was no family history of gastrointestinal cancer and the patient denied any history of smoking, alcohol or IV drug use.
The physical examination was unremarkable. The patient was placed on proton pump inhibitor and was referred for endoscopic ultrasound evaluation.

EGD showed a 1 cm submucosal-appearing lesion located at 25 cm from the incisors. It was round, with normal appearing overlying mucosa, and was easily compressed with cold forceps investigation. No mucosal breaks or ulcers were noted in the esophagus. The stomach showed linear gastritis and the duodenum was unremarkable. EUS (Endoscopic ultrasound) demonstrated a 13 mm × 5 mm anechoic lesion arising from the second echo layer corresponding to the sub-mucosal space. Doppler examination revealed no flow through this lesion. Next, using cold standard forceps, multiple biopsies were taken in a “bite and bite” tunneling method with obliteration of the lesion and expression of a small amount of sero-sanguinous fluid.

Histology findings showed cavernous lymphangioma of the esophagus. The patients’ symptoms of dysphagia were improved after removal of the lesion.

**DISCUSSION**

Benign tumors of the esophagus are rare, constituting less than 1% of all lesions of the esophagus. Approximately 2/3 of the benign esophageal tumors are leiomyomas, while the rest are various polypoid lesions or cysts. Lymphangiomas are benign lesions that arise from the cystic dilatation of hamartomas in deep lymphatic tissue. They are rarely found in the gastrointestinal tract (1%), and the esophagus is the most unusual location for this lesion. The first case of lymphangioma of the esophagus was reported on autopsy by Schmidt[5] and endoscopically by Brady,[6] and, to the best of our knowledge, only 15 cases have been reported in the literature.

The presentation of lymphangioma in the esophagus is markedly varied. It has been reported that there is a higher incidence of lymphangioma of the esophagus during childhood, but it can occur at any age.[7–9] They may range from (smallest lesion seen) up to 5 cm in length. Patients may be asymptomatic or may have overt complaints. The
The most common symptoms are dysphagia, heartburn, post-prandial vomiting and epigastric pain, although some patients may complain of odynophagia or even mid-sternal chest pain. Patients with chest pain are especially clinically challenging as a review of the literature shows that many of these patients have esophagitis, hiatal hernia or gastric ulcers, making it difficult to determine the etiology of the complaint.

The diagnosis of lymphangioma is usually made on histopathology [Figures 1 and 2]. The masses are composed of enlarged channels lined by lymphatic endothelial cells surrounded by a loose myxoid stroma. On gross examination, they are usually pale and smooth, with a multicystic cut surface and a clear yellowish fluid exudate.

The common tools to establish the diagnosis are esophagram and endoscopy. On endoscopy, lymphangiomas appear as a translucent, yellowish, easily compressible mass lesion that are usually less than 5 mm. Standard endoscopic biopsies are often normal as the lesions are submucosal. Esophageal ultrasound may the most useful tool for distinguishing a lymphangioma from leiomyoma or other sub-mucosal tumors because it aids in differentiating sub-mucosal lesions from lesions that arise from the muscular layers of the esophagus.

The treatment of esophageal lymphangioma is conservative in asymptomatic patients. For symptomatic patients, response to surgical or endoscopic removal of these tumors is excellent. Safe endoscopic resection of esophageal lymphangioma of less than 25 mm in diameter has been reported by Suwa and associates. Kuramoto and associates have stated that lymphangiomas less than 20 mm in diameter can be treated by endoscopic resection. However, a large tumor with symptoms or a lesion suspicious for malignancy requires surgical resection. Determining the extent of the tumor through the esophageal wall may be difficult during resection as the lymphangioma is soft and compressible. Yoshida and colleagues suggested that the combination of operative endoscopy and surgery is helpful to confirm the location of the tumor. No cases of malignant transformation have been described in the literature. Radiotherapy appears to be inadequate as the sole method of treatment.

**CONCLUSION**

Lymphangioma of the esophagus is a rare sub-mucosal tumor, generally small and benign in nature. Endoscopic ultrasound is an excellent tool for diagnosis of these lesions. Our experience reaffirms that endoscopic excision of such lesions is safe and appears to be without major complications.

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**REFERENCES**