



Esophageal Schwannoma with Unusual Endoscopic Ultrasound Imaging Findings

Fu-Chieh Hsu¹, Tsai-Wang Huang²

¹Department of Surgery, Tri-Service General Hospital, National Defense Medical Center, ²Division of Thoracic Surgery, Department of Surgery, Tri-Service General Hospital, National Defense Medical Center, Taipei, Taiwan

Esophageal schwannomas are a subset of rare tumors that are difficult to differentiate from other esophageal tumors preoperatively. On endoscopic ultrasound (EUS), esophageal schwannomas generally appear as well-demarcated masses that are heterogeneous, hypoechoic, or mixed-echogenic in nature. We present a case of esophageal schwannoma with atypical EUS findings. EUS revealed two lobulated tumors that originated from the muscularis propria and characterized by heterogeneous hypoechoic features and poor blood flow signals. Right-sided video-assisted thoracoscopic surgery with esophageal tumor enucleation was performed. Immunostaining finally confirmed the diagnosis of an esophageal schwannoma. Esophageal schwannomas tend to show heterogeneous hypoechoic or mixed-echogenic features, but they rarely present with two lobulated masses.

Key words: Esophageal tumors, esophageal schwannoma, endoscopic ultrasound, video-assisted thoracoscopic surgery

INTRODUCTION

Leiomyomas account for the majority of benign esophageal tumors.¹ Of them, the esophageal schwannomas are rare² and therefore challenging to differentiate from other tumors, such as leiomyosarcomas, leiomyomas, and mediastinal tumors.³ Here, we describe the case of an 82-year-old woman who presented with a bulging subepithelial lesion located proximal to the esophagogastric junction, which was detected on esophagoscopy. Endoscopic ultrasound (EUS) revealed an atypical image of the esophageal tumor, which was eventually diagnosed as an esophageal schwannoma after surgical enucleation. The tumor presented as two lobulated masses from the muscularis propria with heterogeneous hypoechogenicity.

CASE REPORT

An 82-year-old female presented with dysphagia and poor appetite for approximately 1 month. The patient denied any comorbidity, and there were no significant findings in her medical history or family history. Laboratory investigation only revealed a high cholesterol level and normal tumor marker

levels, including carcinoembryonic antigen and squamous cell carcinoma antigen levels. Upper gastrointestinal (GI) endoscopy revealed a bulging subepithelial lesion located 40 cm from the incisors and proximal to the esophagogastric junction [Figure 1]. EUS (Olympus GF-UE260) revealed two lobulated tumors derived from the muscularis propria with heterogeneous hypoechoic features and poor blood flow signals [Figure 2]. Contrast-enhanced computed tomography (CT) of the chest revealed a mildly lobulated homogeneous soft tissue mass with a circumscribed margin located over the posterolateral aspect of the lower-third part of the esophagus [Figure 3]. Physical examination, whole-abdominal sonography, and other examination modalities revealed no abnormal findings.

Right-sided video-assisted thoracoscopic surgery with esophageal tumor enucleation was performed. The tumor over the lower third of the esophagus adhered to the aorta and mediastinum, and it was barely possible to dissect it from the esophageal muscular layer. A 5.5 cm × 3.5 cm × 3.5 cm well-encapsulated tumor with a yellowish cut surface and a hard elastic texture was successfully resected [Figure 4a and b]. Two different components were used for frozen sectioning.

Microscopic examination revealed proliferative spindle cells arranged in a whorled pattern and focal lymphoid tissues [Figure 5].

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Corresponding Author: Dr. Tsai-Wang Huang,

Division of Thoracic Surgery, Department of Surgery, Tri-Service General Hospital, National Defense Medical Center, No. 325, Sec. 2, Chenggong Rd., Neihu Dist., Taipei 114, Taiwan. Tel: +886-2-87927167; Fax: +886-2-87927403.

E-mail: chi-wang@yahoo.com.tw

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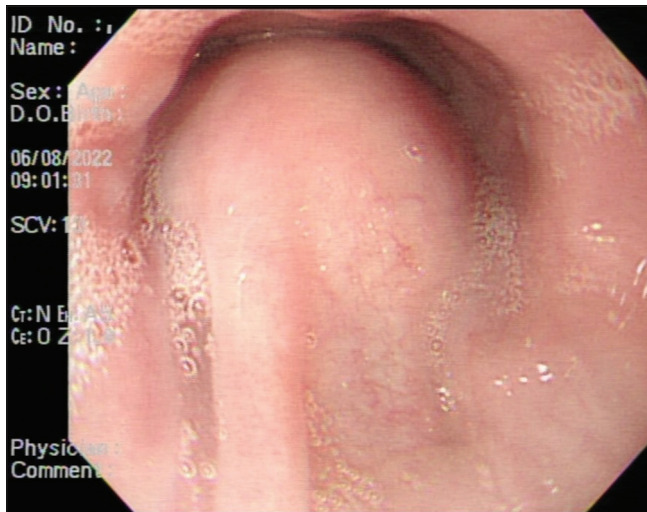


Figure 1: Upper gastrointestinal endoscopy revealing a bulging subepithelial lesion located 40 cm from the incisors and proximal to the esophagogastric junction

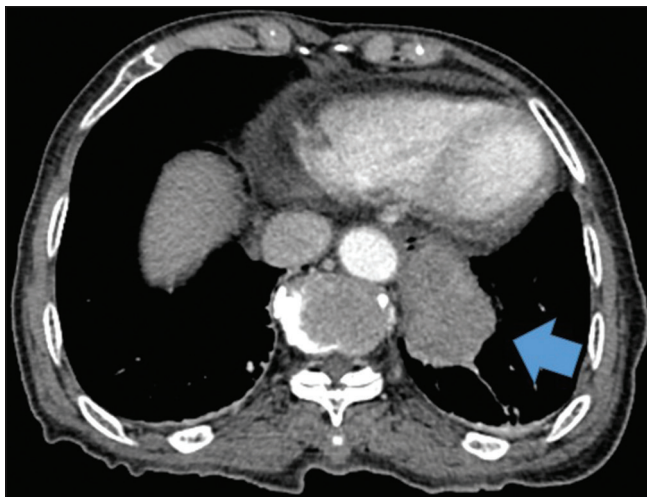


Figure 3: Contrast-enhanced computed tomography of the chest depicting a mildly lobulated homogeneous soft-tissue mass (blue arrow) with a circumscribed margin located over the posterolateral aspect of the lower third of the esophagus

Histopathological examination revealed a tumor arising from the esophageal muscular layer and the formation of peripheral lymphoid follicle. An esophageal schwannoma was diagnosed based on the appearance of tumor cells, which were positive on immunostaining for S-100 and Ki-67 and negative for CD117, DOG-1, and desmin. The postoperative course was uneventful, and the contrast CT of the chest 3 months later showed only the retained surgical clips over the lower third of the esophagus. There has been no evidence of recurrence to date.

DISCUSSION

Approximately 2% of all esophageal tumors are benign, and most of them are leiomyomas.² Esophageal schwannomas

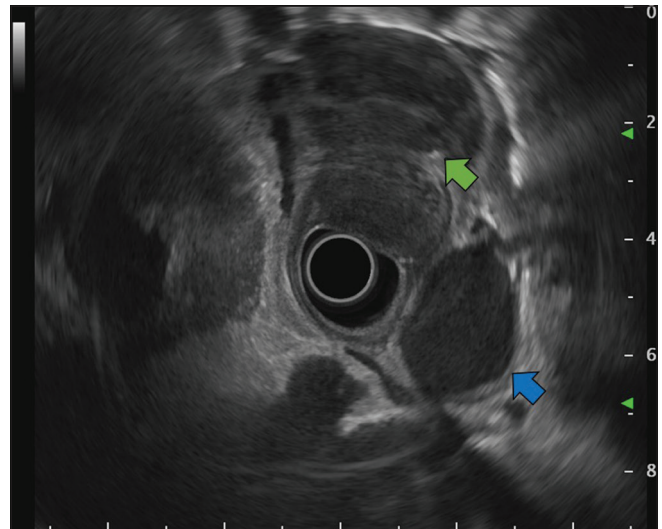


Figure 2: Endoscopic ultrasound revealing two lobulated tumors (green and blue arrows) derived from the muscularis propria showing heterogeneous hypoechoic features and poor blood flow signals

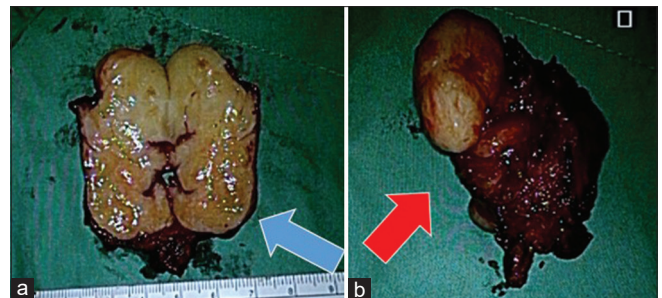


Figure 4: (a and b) A 5.5 cm × 3.5 cm × 3.5 cm well-encapsulated tumor (blue and red arrows) with a yellowish cut surface and hard elastic texture was resected

are extremely rare and predominantly develop in women in their 50s. As most patients are asymptomatic, the tumors are diagnosed incidentally.⁴ Dysphagia is the most common presenting symptom, but dyspnea, chest pain, epigastric pain, and hemoptysis may also occur.^{1,4} GI schwannomas originate from Schwann cells of the axon of Auerbach's plexus or, less frequently, of Meissner's plexus. These tumors can span the entire thoracic esophagus but usually occur in the upper thoracic segment.^{4,5} The present patient had a tumor in the lower third of the esophagus. Because accurate diagnosis during routine endoscopy was challenging, EUS was performed. All submucosal lesions >1 cm in size are assessed by EUS.⁶ EUS can distinguish the specific layer of the GI tract wall, determine the layer from which the lesion is derived, and even acquire cells or tissues for histopathology using EUS fine-needle aspiration biopsy. Su *et al.*⁷ reported that GI schwannomas generally present as heterogeneous hypoechoic or mixed-echogenic masses originating from the muscularis propria with clear

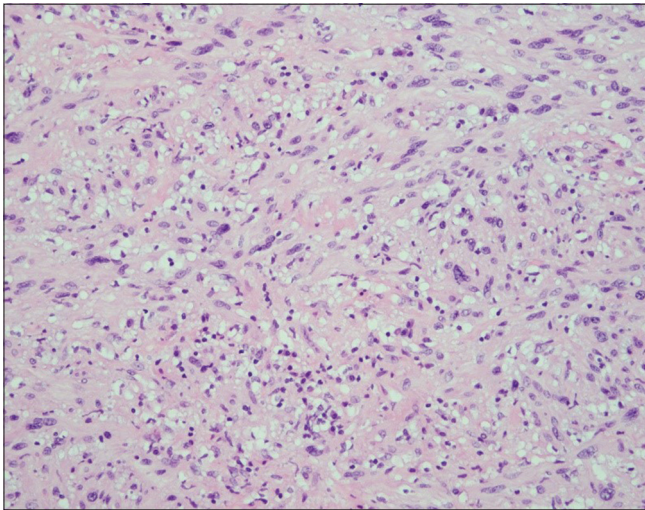


Figure 5: Microscopic examination results revealed proliferative spindle cells arranged in a whorled pattern and focal lymphoid tissues

boundaries and poor blood flow signals. In addition, EUS real-time tissue elastography can help distinguish between benign and malignant tumors as the hardness of tissues are evaluated using a 5-point scoring system.⁸ However, biopsies should be avoided in probable esophageal leiomyomas or GI stromal tumors (GISTs) because postbiopsy scarring may hinder surgical enucleation.⁹ As a result, establishing a preoperative definitive tissue diagnosis is challenging. On CT scans, esophageal schwannomas display a homogeneous denseness, while leiomyomas often show calcification. In contrast, leiomyosarcomas, being malignant, are more heterogeneous. Urgent surgical resection is recommended for the removal of symptomatic lesions, lesions that have increased in size during follow-up, or premalignant lesions (GIST size >2 cm).^{1,10} Video-assisted thoracic surgery for enucleation is preferable because of the shorter length of hospital stay, less postoperative pain, and fewer postoperative complications.

CONCLUSION

Esophageal schwannomas show a homogeneous density, whereas leiomyomas often show calcification on CT scans. Esophageal schwannomas show heterogeneous hypoechoic or mixed-echogenic features, and originate from the muscularis propria. They rarely present with two lobulated masses as seen in the present case.

Ethical approval

The study has been approved by an ethics committee: (TSGHIRB No: A202215169).

Declaration of patient consent

The authors certify that they have obtained all appropriate

patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Data availability statement

The data that support the findings of this study are available from the corresponding author, Huang TW, upon reasonable request.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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