ESOPHAGEAL SCHWANNOMA

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Abstract: Benign tumors of the esophagus are rare, and almost all of them are leiomyomas. Esophageal schwannoma is extremely rare. We report a case of esophageal schwannoma in a 54-year-old Taiwanese man who was admitted due to dysphagia. Barium esophagogram revealed a protrusive smooth-marginated filling defect of about 3.6 cm in diameter with intact mucosa over the lower third of the esophagus. The submucosal tumor was removed via left thoracotomy with enucleation. There was no anatomic relationship between the tumor and the vagal nerve trunk. Histopathologic examination revealed interwined bundles of spindle cells with spiral-like proliferation. Immunohistochemical study was positive for S-100 protein. Esophageal schwannoma was diagnosed. The patient had no postoperative complications, and was healthy and free of tumor recurrence 16 months after operation.

Key words: Esophageal neoplasms; Immunohistochemistry; Male; Neurilemmoma


Benign tumors of the esophagus are rare, and almost all of them are leiomyomas. Schwannoma is extremely rare as an esophageal tumor. Most benign esophageal submucosal tumors are treated by resection due to progressive enlargement; the preoperative impression is usually a leiomyoma of the esophagus.1,2 A definitive diagnosis of esophageal schwannoma requires confirmation by histopathological, immunohistochemical or electroscopic studies.1 Here we report a case of esophageal schwannoma.

Case Report

A 54-year-old man was healthy except for a history of gastric ulcer for which he had received regular medical treatment in our hospital for several years. In November 1995, he underwent panendoscopy examination for follow-up of the gastric ulcer. A submucosal tumor 1.0 cm in size in the esophagus located at a distance of 35 cm from the incisor teeth was incidentally found. Esophagoscopy with biopsy was done and the results revealed chronic esophagitis without dysplastic change. Regular gastrointestinal outpatient department follow-up was performed thereafter. In 1996, the lesion was found to be slowly enlarging, and by 1999 it had doubled in size over a 4-year period. Because the patient had no clinical symptoms, he refused to undergo further invasive treatments. However, the lesion continued to grow and symptoms of dysphagia were noted during the next year. The patient was admitted for surgical treatment on September 25, 2001. At admission, the patient had no other digestive tract symptoms except dysphagia. There was no evidence of anemia or jaundice, and the thorax and abdomen were unremarkable at examination. Laboratory analysis, including hematological profile, biochemical profile and urinalysis, revealed no abnormalities. Tumor markers such as squamous cell carcinoma antigen were also within the normal range.

Endoscopy showed a 1.0-cm smooth and protruding submucosal lesion in the esophagus at a distance of 35 cm from the upper row of teeth. An endoscopy before this hospital admission in July 2001 showed obvious enlargement of the protruding submucosal lesion. A submucosal tumor of 4.0 cm in diameter was detected (Fig. 1A).

Before this admission, roentgenographic examination of the esophagus with barium swallow showed a smooth margined filling defect about 3.6 cm over the lower third of esophagus with intact mucosa, and computed tomography (CT) revealed a short segment of eccentric wall thickening, about 1.3 cm in maximal wall thickness, over the lower third of the esophagus (Fig. 1B).
Preoperative evaluation demonstrated an esophageal submucosal lesion, and a leiomyoma was strongly suspected. On September 27, 2001, left-side posterolateral thoracotomy was performed because the lesion was located over the lower third of the esophagus, about 3 cm above the esophago-gastric junction. The tumor was enucleated from the esophageal wall smoothly. There was no anatomic relationship between the tumor and vagus nerve trunk and trachea. The tumor measured 2.5 x 2.0 x 1.5 cm in size and was elastic and firm. The cut surface of the tumor was yellowish-white (Fig. 2).

Histologically, the tumor was composed of thin, densely cellular spindle-shape cells arranged in interlacing fascicles (Fig. 3A). The nuclei of the tumor cells showed elongation or undulation and distinct cytoplasm without conspicuous dysplastic change, necrosis, or mitosis. Schwannoma was diagnosed based on these pathological findings. This diagnosis was also supported by positive immunohistochemical staining for S-100 protein and negative staining for actin (Fig. 3B). The postoperative course was uneventful and the patient was alive and well with no tumor recurrence at 16 months after the operation.

Discussion

Schwannoma is the most common primary tumor of the mediastinum. However, esophageal schwannoma is quite rare. Chatelin and Fissore first reported this entity in 1967, and only 18 cases including the present case have been reported. Esophageal schwannoma has been previously reported in Taiwan, and this is the second reported case. Review of the previously reported cases indicated that the prognosis of esophageal schwannoma is generally excellent. There was a slight female predominance and the site of tumor origin was in the thoracic esophagus in most cases. The most common preoperative diagnoses in these patients were esophageal submucosal tumor, esophageal leiomyoma or leiomyosarcoma, cysts, or lipomas. This is probably because of the lack of awareness of schwannoma arising in the esophagus, due to its extremely low incidence.

Differentiation of schwannoma from other submucosal tumors is very difficult on preoperative
examination by esophagoscopy, esophagography or CT. Diagnosis of the tumor type requires histological examination, and the identification of neurogenic tumor cell origin by positive immunostaining for S-100 protein. If immunostaining for S-100 protein is positive, the diagnosis of esophageal schwannoma can be made with confidence.

Methods for performing surgical resection of these tumors include thoracotomy, thoracoscopy, and endoscopy. Thoracotomy with resection is the preferred therapy for esophageal tumors, and if a submucosal tumor of the esophagus has a diameter of 2 cm or less, removal can be accomplished endoscopically.6,7 Thoracoscopic resection of tumor8 has become increasingly common more recently because of its low degree of invasiveness and reduced pain at the surgical wound site.

The prognosis in patients with esophageal schwannoma is generally excellent although 1 case with local recurrence was reported.9 Early removal of submucosal esophageal tumors is indicated and long-term follow-up for recurrence after operation is important.

Fig. 3. A) Histological appearance of the tumor shows thin, densely cellular, spindle-shaped cells arranged in interlacing fascicles (hematoxylin and eosin, x 10). B) Positive immunohistochemical staining for S-100 protein showing hyalinization of a vessel (S-100 immunohistochemical stain, x 400).

References