

Benign intramural schwannoma of the esophagus – case report

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Abstract

Schwannomas of the esophagus are rare peripheral nerve tumors. A 37-year-old woman with dysphagia was found to have an intramural tumor of the upper esophagus. The lesion was revealed on computed tomography. Endoscopic ultrasound biopsy was nondiagnostic. Through right thoracotomy, the mass was enucleated from the wall of the esophagus. Benign schwannoma was diagnosed only after immunological staining examination.

Key words: intramural oesophageal tumours, schwannoma, diagnosis, treatment.

Streszczenie

Nerwiaki ostonkowe rzadko umiejscawiają się śródściennie w przełyku. Kobieta, 37 lat, była diagnozowana z powodu zaburzeń w połykaniu. Tomografia komputerowa klatki piersiowej wykazała śródścienny, owalny guz górnego piersiowego odcinka przełyku. Biopsja guza techniką ultrasonografii endoskopowej nie pozwoliła na postawienie diagnozy. U pacjentki wykonano prawostronną torakotomię z enukleacją masy patologicznej ze ściany przełyku. Badanie histopatologiczne z zastosowaniem immunohistochemii wykazało łagodnego nerwiaka ostonkowego ściany przełyku.

Słowa kluczowe: guzy śródścienne przełyku, nerwiak, diagnostyka, leczenie.

Introduction

Benign primary intramural tumors of the esophagus are rare and account for approximately 2% of all esophageal tumors [1].

Most of them are esophageal leiomyomas; less frequent are schwannomas or malignant masses such as gastrointestinal stromal tumors (GIST). Endoscopic ultrasound biopsy or frozen section biopsy during the operation does not differentiate the tumors. Only immunohistochemical studies can allow proper diagnosis. We report a case of benign esophageal schwannoma causing progressive dysphagia in a 37-year-old woman.

Case report

A 37-year-old woman presented due to symptoms of chest pain during swallowing of solid food for 3 months. The physical examination was noncontributory. Computed tomography of the chest showed a soft tissue mass in the lumen of the upper esophagus (Fig. 1). Endoscopic ultrasound identified the mass at 19 cm from the incisors, but the biopsy was nondiagnostic. A provisional diagnosis of esophageal leiomyoma was made.

Bronchoscopy and pulmonary function test of the patient were normal. The right chest was opened through a right anterolateral thoracotomy at the fifth intercostal space. The esophagus was mobilized from the neck to the azygos vein.

The intramural tumor 3.5 × 3 × 3 cm was enucleated without erosion of the mucosa (Fig. 2). The esophageal wall allowed primary closure of the muscular layers, and the native esophagus was preserved.

Histopathological examination of the tumor revealed spindle-shaped cells in a fasciculated and disarrayed architecture and nuclei in a palisading pattern (Fig. 3). Immunohistochemically the tumor was positive for S100p and GFAP, and negative for CD34, CD117, SMA, desmin and H-caldesmon (Fig. 4). The diagnosis was benign schwannoma.

Postoperative recovery was uneventful, and there has been no evidence of recurrence to date.

Discussion

Esophageal schwannoma is uncommon and difficult to diagnose in preoperative investigations due to its similarity to other intramural esophageal tumors (leiomyoma,

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Fig. 1. Computed tomography scan showing concentric soft tissue lesion surrounding the esophagus

GIST, oesophageal cysts or cancer). These tumors more frequently develop in women and are often located in the upper and mid esophagus. Malignant schwannoma has also been reported [1, 2]. Symptoms of this tumor include dysphagia, dyspnea [1, 3], chest pain, and hematemesis, and appear when the tumor increases in size. Imaging studies (positron emission tomography, computed tomography, and magnetic resonance imaging) are useful for the confirmation of esophageal tumor [1, 4, 5]. Endoscopic ultrasonography-guided fine needle aspiration biopsy is not always useful for diagnosis, as in our cases [1]. The value of frozen section is limited, and it can lead to diagnostic pitfalls [6].

Final diagnosis is possible upon positive immunohistochemical studies for S-100 protein and negative staining for smooth muscle markers such as SMA, CD34, and CD117 [7].

Surgical excision of the tumor is the method of choice, because chemotherapy and radiotherapy are ineffective. The option of enucleation is recommended when: the tumor is well encapsulated, a clear margin is achievable, and there is sufficient redundant mucosa to close the defect [6].



Fig. 2. Enucleated esophageal schwannoma (3.5 × 3 × 3 cm)

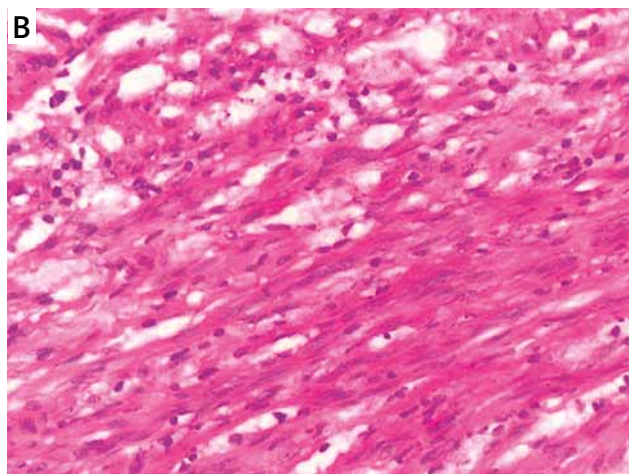
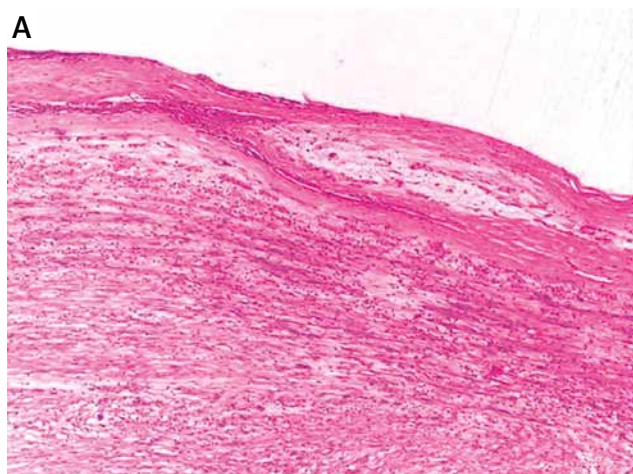


Fig. 3. Histology of the esophageal mass upon resection. Spindle-shaped cells in a fasciculated and disarrayed architecture and nuclei in a palisading pattern. A) ×40, B) ×200

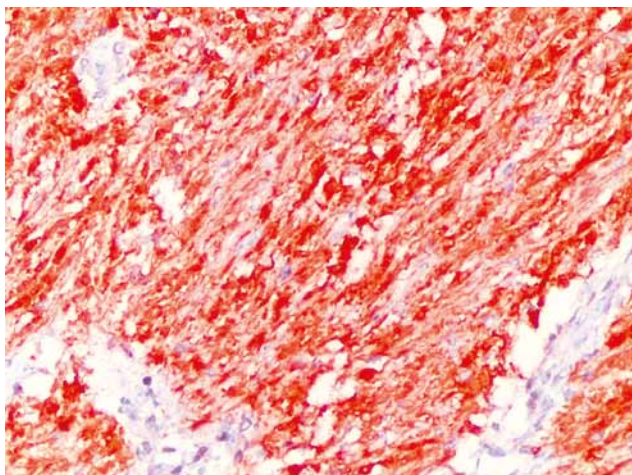


Fig. 4. Immunohistochemical studies revealed S100 protein positivity

In our case we removed the tumor without damage to the mucosa.

Enucleation with video-assisted thoracoscopic surgery (VATS) is possible for small tumors (≤ 2 cm). For large tumors (≤ 8 cm) with broad adjacent to the muscular layer and an extensive mucosal defect, esophagectomy with gastric pull-up is recommended [3, 7]. The long-term prognosis

after conservative resection of benign esophageal schwannoma is unknown.

Disclosure

Authors report no conflict of interest.

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