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Schwannoma of the Esophagus: Computed Tomography and Endosonographic Findings of a Special Type of Schwannoma

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Schwannomas of the digestive tract are uncommon benign tumors seldom located in the esophagus. We present a case of intramural esophageal schwannoma with histopathological correlation evaluated by computed tomography and endoscopic ultrasonography. The imaging findings are described and other relevant features of schwannomas of the esophagus are discussed.

Key words: Computed tomography; endoscopic ultrasonography; esophageal neoplasms; schwannomas

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Schwannomas of the esophagus have been reported sporadically in the medical literature. As far as we know, only 21 cases have been described. While some cases seem to be schwannomas of the conventional type arising from the vagus nerve (2, 18), other reports describe intramural tumors bearing no recognizable relationship with any nerve trunk. It has been proposed that the latter could represent a distinct pathological entity (1, 16).

Case Report

A 48-year-old woman presented with a 2-year history of progressive dysphagia, worse for solids, with no other clinical complaints. Upper gastrointestinal endoscopy disclosed a submucosal lesion 24–28 cm from the incisors. The patient was referred to our institution. Computed tomography (CT) showed a homogeneous soft-tissue mass between the middle and lower third of the posterior wall of the esophagus, with smooth, well-defined contours (Fig. 1). After intravenous contrast administration, no significant enhancement was seen (Fig. 2). The lesion produced dilatation of the proximal esophagus. There were no other associated findings.

Endoscopic ultrasonography (EUS) revealed a well-defined, hypoechoic, slightly heterogeneous solid lesion (Fig. 3) that seemed to arise from the

muscularis propria of the esophagus, measuring 6 × 5 cm and replacing the submucosa. No regional lymph nodes were seen. Surgical enucleation was performed. The tumor was intramural and did not have any relationship with the vagus nerve or its branches. On gross inspection, a 9 cm, well-circumscribed, compact, yellow rubbery mass was seen (Fig. 4). A mesenchymal spindle cell neoplasm, well delineated but lacking a true capsule, with a prominent peripheral lymphoid cuff was seen microscopically (Fig. 5) composed of interlacing



Fig. 1. CT before intravenous contrast administration. A homogeneous soft-tissue mass with regular contours was observed involving the posterior esophageal wall.

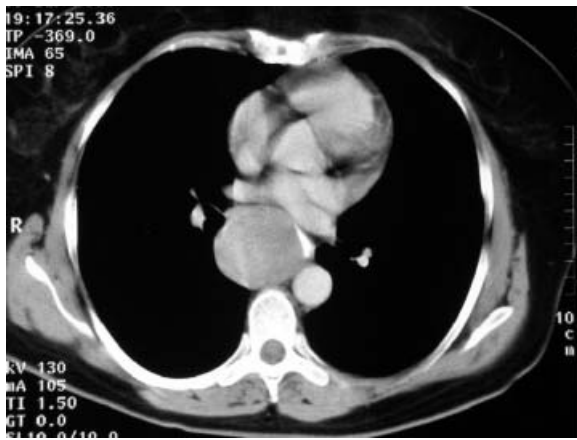


Fig. 2. Contrast-enhanced CT. The tumor did not show significant enhancement.

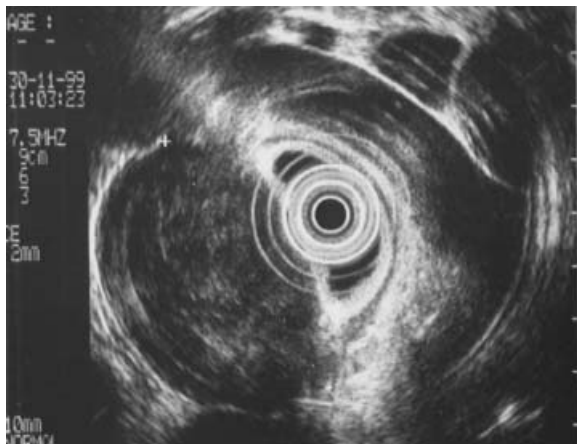


Fig. 3. Endoscopic ultrasonography. This sonogram shows a well-defined, hypoechoic, slightly heterogeneous solid lesion in the esophageal submucosa.

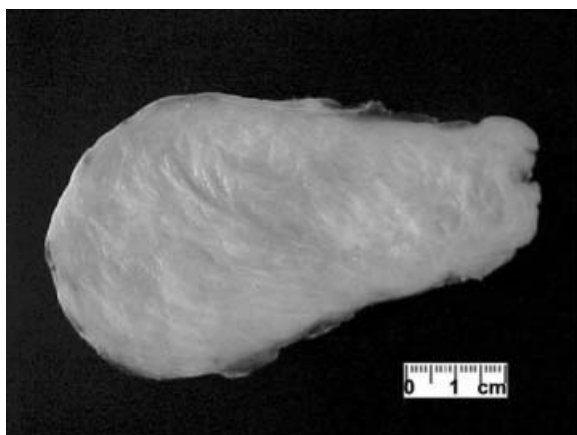


Fig. 4. Grossly, the tumor was a 9cm, well delimited, yellow rubbery mass, with no hemorrhage or necrosis.

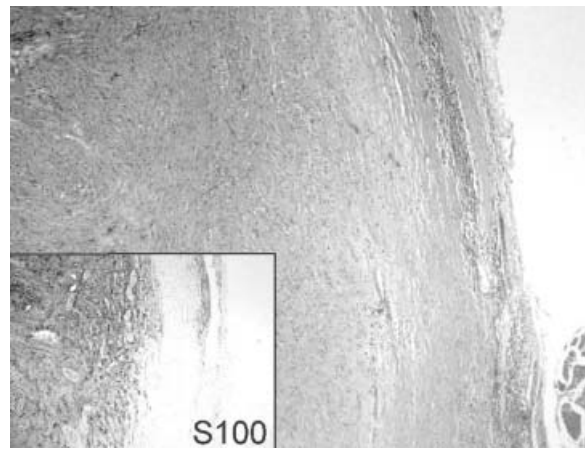


Fig. 5. Microscopically, the neoplasm lacked a capsule, was composed of fascicles of spindle cells and had a prominent peripheral lymphoid cuff (H&E, 50 $\times$). The cells were strongly positive for S100 protein (inset).

bundles of spindle cells, wavy collagen strands, thin-walled vessels, and interspersed inflammatory cells. No mitotic activity was found.

Immunohistochemically, the tumor showed diffuse and strong positivity for S100 protein, while CD34, CD117, actin, and desmin were negative. These morphological features were identical to those previously reported as digestive tract schwannomas (DTS) (1, 16).

Three years after surgery, the patient is well, with no evidence of recurrent disease.

Discussion

Esophageal schwannomas can be separated into two different types. Some are conventional, originating from the vagus nerve or its branches and extend into the esophageal wall. Others, less common, and unique to the digestive tract, are probably derived from Schwann cells of the neural plexus of the digestive tract. They are mostly intramural tumors, but may also present as intraluminal polypoid lesions.

The majority of schwannomas of the digestive tract (DTS) occur in the stomach. Pathological differences compared with schwannomas of peripheral nerves are the absence of capsule, lack of nuclear palisading, and the presence of a peripheral lymphoid cuff (10).

Clinical presentation of esophageal schwannomas depends on tumor location, growth pattern, and size. Dysphagia is the predominant symptom, secondary to narrowing of the esophageal lumen. Rare manifestations include cough and dyspnoea, associated with schwannomas of the cervical

esophagus (4), and acute hemorrhage (2). Few cases have been detected incidentally while asymptomatic (5, 8).

It is not possible to establish a definitive and correct preoperative diagnosis using current imaging techniques. Esophageal schwannomas have imaging findings similar to other more frequent mesenchymal esophageal tumors, such as leiomyomas, leiomyosarcomas (9), and others.

A chest radiogram can reveal an abnormal enlargement of the mediastinum (4, 5). On barium studies, most of these tumors appear as rounded filling defects with smooth margins (3, 6, 17), often causing narrowing and displacement of the esophageal lumen (4, 18).

Esophageal schwannomas have been barely characterized on CT, particularly in regard to post-contrast behavior (6). This technique allows some distinction from other more frequent lesions. Both cysts and lipomas have different densities; leiomyomas usually show enhancement and sometimes calcifications, and leiomyosarcomas tend to be heterogeneous (9). Neurofibromas and gastrointestinal stromal tumors (GIST) remain in the differential diagnosis.

EUS features of esophageal schwannomas are also non-specific. Cysts and lipomas have typical appearances. Malignant tumors, namely malignant GIST, can be recognized by the presence of irregular or lobulated extraluminal margins, cystic spaces, and lymph nodes with malignant features (spherical, hypoechoic, sharply demarcated, size >1 cm) (12, 14, 15). Further distinction can be achieved by EUS-guided fine needle aspiration biopsy (EUS-FNA). Although the definitive diagnosis of schwannomas is made by histological examination, requiring an appropriated immunohistochemical study, EUS-FNA may disclose its neural origin (7, 11).

The treatment of DTS should be total excision, which can be achieved by surgical enucleation or by endoscopic removal in small lesions (13). The prognosis after complete resection is excellent.

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