

## A case report on oesophageal schwannoma: A rare occurrence

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### Abstract

Most tumorous lesions of the esophagus are esophageal cancers. Benign primary tumors of the esophagus are uncommon, and account for approximately 2% of all esophageal tumors. More than 80% of benign esophageal tumors are leiomyomas, with schwannomas being rare. A 55-year-old man visited our internal medicine department with complaints of palpitations and discomfort during swallowing. A chest computed tomography scan showed a lobulated tumor (5x4x2 cm) in the upper to middle mediastinum, with homogenous inner opacity, compressing the esophagus. Upper gastrointestinal endoscopy revealed a smooth-surfaced elevated lesion covered with normal mucosa, and a schwannoma was diagnosed based on the biopsy result. Histopathological examination revealed spindle-shaped cells in a fasciculated and disarrayed architecture and nuclei in a palisading pattern. Immunohistochemical studies revealed S100 protein positivity and the absence of staining for  $\alpha$  smooth muscle actin ( $\alpha$ SMA), CD34 and CD117, thereby establishing the diagnosis of benign schwannoma. Her postoperative course was uneventful and there has been no evidence of recurrence to date.

**Keywords:** Schwannoma, Leiomyoma, Mediastinum, Palisading, Submucosa

### Background

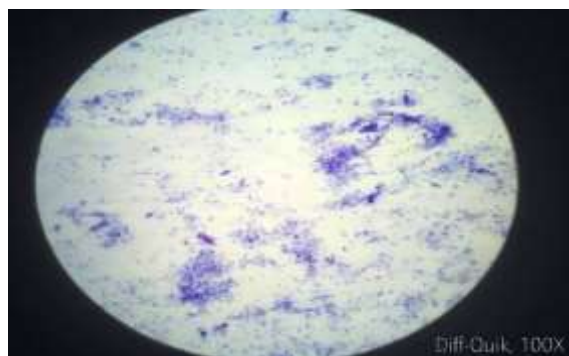
The incidence of benign primary tumors of the esophagus is low. Most are leiomyomas, and schwannomas are rare. We report a patient with schwannoma who was referred to us for evaluation of a mediastinal tumor. Detailed examination yielded a diagnosis of schwannoma arising from the esophageal submucosa and surgery was performed for excising the tumour.

### Case Presentation

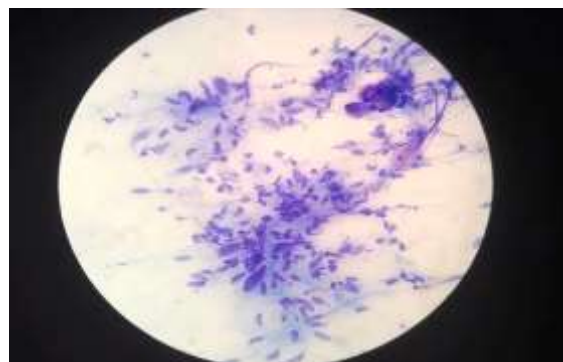
A 55-year-old man visited our hospital with complaints of palpitations and discomfort during swallowing. Her medical and familial histories were unremarkable. A frontal chest radiograph showed a smooth round mass, and a lateral radiograph showed a smooth mass slightly larger than 5 cm in diameter in the middle mediastinum between the trachea and the vertebral bodies. A chest computed tomography (CT) scan showed a lobulated tumor of size 5 × 4 × 2 cm in the upper to middle mediastinum, with a homogenous inner component, compressing the esophagus. Magnetic resonance imaging (MRI) of the chest revealed no invasion of surrounding organs. On imaging studies, a mesenchymal tumor such as gastrointestinal stromal tumor was suspected. Upper gastrointestinal endoscopy showed a smooth elevated lesion, 23 cm from the incisor teeth.

CT guided FNAC was performed which revealed spindle cells with ovoid to spindle shaped nuclei with pointed ends and bland chromatin in a fibrillary background.(Fig. 1, 2) Although there was no evidence of malignancy, the patient underwent surgery because of the large size of the tumor, dysphagia, palpitations caused by the tumor compressing the heart, and a suspicion of malignant potential. A schwannoma was diagnosed based on the biopsy result for the lesion. The

patient was placed in the left lateral position and underwent a mini thoracotomy via the fifth right intercostal space with thoracoscopic assistance. A mass of size 5 cm in diameter was found in the oesophageal wall which was resected and sent for histopathological examination.



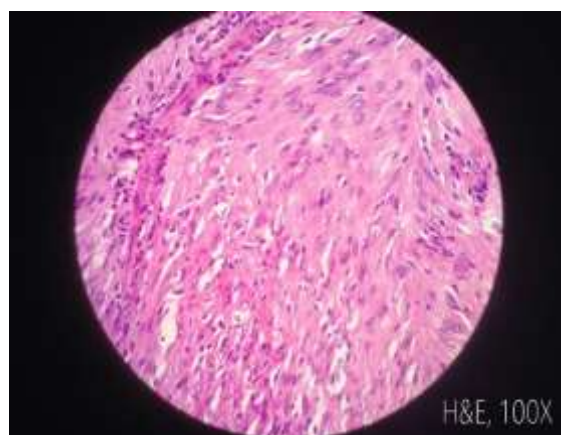
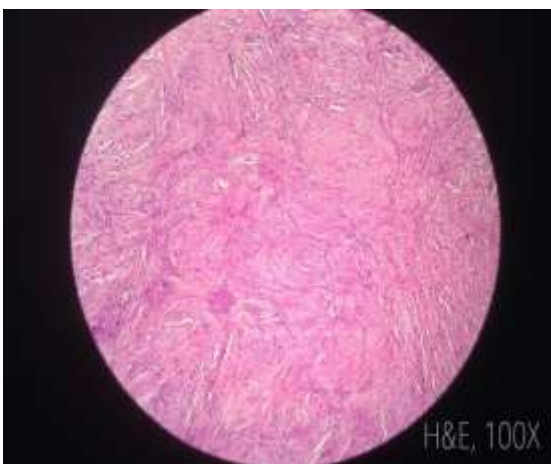
**Fig. 1: Spindle shaped cells in a fibrillary background (Diff-Quick, 100X)**



**Fig. 2: Higher magnification: showing spindle shaped cells with oval to spindle nuclei in a fibrillary background**



**Fig. 3:** Gross appearance of excised tumour: well encapsulated, grayish yellow, homogenous



**Fig. 4,5,6 & 7:** H & E images showing spindle shaped cells arranged in palisading fashion with both Antoni A & B areas and verocay bodies

### Histopathological Findings

**Grossly:** The tumor was well demarcated and elastic hard, had a globular appearance, and measured 5×4×2 cm. The cut surface was almost uniformly milky white. (Fig. 3)

Histopathological examination revealed spindle-shaped cells in a fasciculated and disarrayed architecture and nuclei in a palisading pattern. Two different patterns were recognized as Antoni A and Antoni B. Type A Antoni areas were quite cellular and composed of spindle cells arranged in palisading fashion with presence of verocay bodies. Type B areas were less cellular, sparsely situated. Mitoses were absent. (Fig. 4,5,6 &7)

Immunohistochemical studies revealed S100 protein positivity and the absence of staining for smooth muscle actin (SMA), establishing the diagnosis of benign schwannoma. (Figure 8,9) His postoperative course was uneventful and there has been no evidence of recurrence to date.

### Discussion

Most tumorous lesions of the esophagus are esophageal cancers. Benign primary tumors of the esophagus are uncommon, and account for approximately 2% of all esophageal tumors. More than

80% of benign esophageal tumors are leiomyomas, with Schwannomas being rare<sup>[1,2]</sup>. Schwannoma of the esophagus more frequently develops in women than in men and these tumors are often located in the upper and mid esophagus in the mid mediastinum. A preoperative diagnosis of this condition is difficult, and the definitive diagnosis is often established after resection<sup>[3]</sup>. A schwannoma that developed in the posterior mediastinum was reported<sup>[4]</sup>. Furthermore, a patient with a malignant schwannoma has also been reported, but such cases are extremely rare<sup>[5]</sup>.

Symptoms of this disease include dysphagia, dyspnea, and chest pain, and are likely to appear and worsen as the schwannoma increases in size<sup>[6]</sup>. Fluorodeoxyglucose positron emission tomography as well as CT and MRI are reportedly useful for the confirmation of mediastinal tumors<sup>[7]</sup>. However, diagnosing a schwannoma is difficult with only imaging studies. In our present case as well, the results from the biopsy obtained with esophagogastroendoscopy were necessary for establishing the diagnosis. Schwannoma is a submucosal tumor, and endoscopic ultrasonography-guided fine needle aspiration biopsy is reportedly useful for both diagnosis and management<sup>[8]</sup>.

In general, histological features of schwannoma include spindle-shaped tumor cells arranged in a palisading pattern or with loose cellularity in a reticular array. Immunohistochemical stainings for S100 protein,  $\alpha$ SMA, CD34 and CD117 are also useful<sup>[9]</sup>.

Chemotherapy and radiation therapy are ineffective such that surgical excision including the capsule is indicated for symptomatic cases and also for those with possibly malignant schwannoma. The use of enucleation with video-assisted thoracoscopic surgery is becoming common for small tumors ( $\leq 2$  cm). However, for large tumors ( $\geq 8$  cm) with broad areas adjacent to the esophageal muscular layer, the mucosal defect becomes extensive, and esophagectomy and esophagogastrostomy are thus usually performed<sup>[10]</sup>. In our present case, the tumor was approximately 5cm in diameter which was excised. His postoperative course was uneventful. She had neither postoperative stricture nor problems with eating and drinking. This favorable outcome confirmed the procedure to have been appropriate.

## Conclusions

Here, we report a relatively rare case of schwannoma of the esophagus, initially identified as a medium sized mediastinal tumor. The tumor was 5cm in diameter. The mass was excised and oesophageal wall repaired. Patient was followed up for a year and he developed no recurrence postoperatively and was doing well.

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