

Case Report

Upper Esophageal Schwannoma: Rare Differential Diagnosis of Dysphagia

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ABSTRACT

Esophageal schwannomas are rare primary submucosal tumors, 45 cases have been reported so far. We herein report the 46th case of an esophageal schwannoma from Nepal. A 60-year-old woman presented with progressive dysphagia. Oesophago-Gastro-Duodenoscopy (OGD) showed a submucosal mass with mucosal puckering in the upper esophagus; Computed tomography (CT) of the chest showed an upper esophageal mass of size 8x7x6cm³ compressing the trachea. Bronchoscopy showed external compression of the mid trachea. The patient underwent three incision VATS esophagectomy. Histopathological examination and immunohistochemical (IHC) staining confirmed the diagnosis of schwannoma.

Keywords: Esophagus, Schwannoma, VATS, Esophagectomy.

Introduction

Esophageal schwannomas are rare primary submucosal tumors (ESMTs). Benign esophageal tumor accounts for approximately 2% of all esophageal tumors; more than 80% are leiomyomas.^{1,2} These ESMTs are difficult to diagnose preoperatively and the final diagnosis is often made after surgery and IHC staining.²

We here report a case of large upper esophageal schwannoma which was managed by video-assisted thoracoscopic surgery (VATS)-esophagectomy and a review of previously reported cases in the literature till date has been done.

Case Report

A 60 years old female was referred to our center with a complaint of chest discomfort and dysphagia for solid food for 7 months. Dysphagia was progressive, relieved by taking clear liquid. She gives no history of shortness of breath, her past medical and family history

was unremarkable. Oesophago-Gastro-Duodenoscopy (OGD) showed a submucosal mass bulging on the posterior esophageal wall with puckered mucosa located at 17 to 23 cm from the incisors (Figure 1). CT of the chest showed an upper esophageal mass, 8x 7x 6cm³ in size, compressing trachea with deviation towards the right, and multiple periesophageal lymphadenopathies: largest measuring 1.5x1cm² (Figure 2). Bronchoscopy showed external compression of the mid trachea.



Figure 1: View of tumor during Oesophago-Gastro-Duodenoscopy (OGD)

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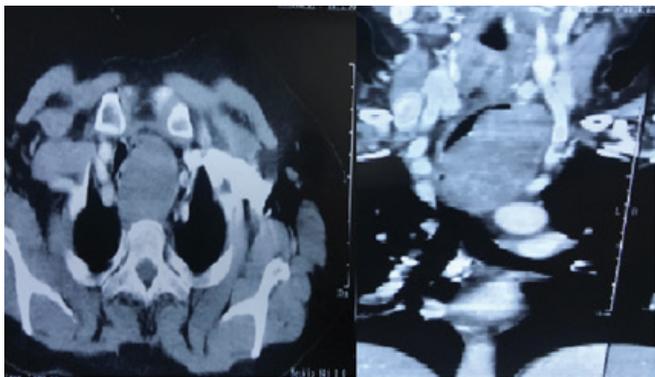


Figure 2: esophageal luminal compression due to mass on the esophagus as shown on CT Chest (Axial and Coronal view)

The patient was taken up for surgery with a provisional diagnosis of leiomyoma. Surgery was performed with single-lumen endotracheal tube insertion, in a semi-prone position. A 10mm camera port was inserted at 7th intercostal space (ICS) along with the midaxillary line and carbon dioxide (CO₂) pneumothorax was created at 8mm of Hg. Two 5mm working ports were placed at 5th ICS along the anterior axillary line, and 6th ICS along the posterior axillary line, respectively. On inspection, a lower margin of the tumor was visualized approximately 2cm above the azygos arch extending to the right thoracic apex. Enucleation of the tumor was attempted but due to dense adhesion and possible invasion of mucosa by the tumor, it was not possible. Hence, esophagectomy was decided. Complete thoracoscopic esophageal mobilization was done. The thoracic duct was ligated and ports were closed.

Patient was repositioned in supine, and an upper midline laparotomy was carried out. The stomach was mobilized over the right gastric and right gastro gastroepiploic vessels. Left-sided neck incision was made anterior to the left sternocleidomastoid muscle and the cervical esophagus was mobilized. The tumor was felt extending from the chest cavity to the lower part of the cervical esophagus. The esophagus was divided at the neck approximately 2 cm above the tumor and the esophagus along with the tumor was excised through laparotomy incision.

Gastric conduit of 5 cm in diameter was created and

pulled up through the bed of the previous esophagus to the left side of the neck. The gastroesophageal hand-sewn anastomosis was done in 4 layers, and anastomosis was wrapped with a pedicle of omentum. Feeding jejunostomy was performed. Incisions were closed. The postoperative period was uneventful, patient was discharged on the 12th postoperative day.

Specimen: There was a solid lobulated mass of size 8x7x6cm³ arising from the muscularis propria of the esophagus, extending from cervical to thoracic esophagus. Mass densely adhered to the mucosa (Figure.3).

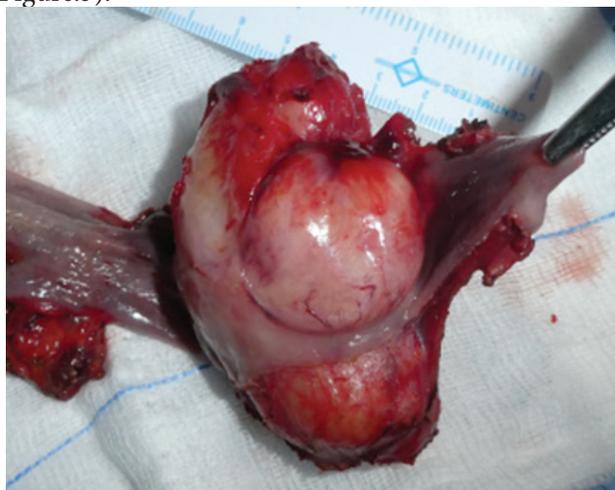


Figure 3: Gross surgical Specimen

Histopathology: Tumor showed neoplastic Schwann cells with a moderate amount of cytoplasm with ill-defined cell borders and vague nuclear palisading). Tumor cells strongly and diffusely expressed S-100 protein (Figure 4). The above findings confirmed the diagnosis of schwannoma.

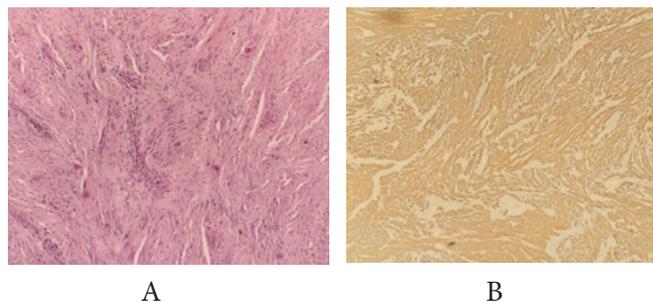


Figure 4: Photomicrograph of the specimen in A. H and E stain and B. immunohistochemistry for S-100 expression. (H&E stain, 20X)

Discussion

Esophageal schwannomas are extremely rare. Chaterlin and Fissore described the first esophageal schwannoma in 1967. A thorough review of literature in Pub Med showed only 30 cases till 2010 and 15 more cases from 2010 to date.^{2,3} Our case seems to be the 46th case. Esophageal schwannoma is commonly located in the upper and mid esophagus, more predominant in female and Asian population.^{4,5}

Patient with ESMTs remains asymptomatic for a long time. Symptoms depend on the location and size of the mass. Progressive dysphagia (53.7%), dyspnea (10.4%), cough (4.4%), chest pain (4.4%) and weight loss (4.4%) are most common features.⁵⁻⁸

Preoperative diagnosis is difficult due to the rarity of the disease, difficult anatomical location, and low yield on biopsy. Imaging modality includes X-ray, CT scan, magnetic resonance imaging (MRI), positron emission tomography (PET), esophagography, OGD, endoscopic ultrasound (EUS). EUS-guided fine-needle aspiration biopsy (EUS-FNAB) is useful for both pre-operative diagnosis and management of this disease, though EUS-FNAB may have several procedural risks: such as bleeding and infection. Mucosal trauma might cause difficulties in intra-operative enucleation of the tumor. Diagnostic accuracy of EUS-FNAB for SMTs is around 85.2 % as reported by Rong et al.^{9,10}

In general, histological features of schwannoma include spindle-shaped tumor cells arranged in a palisading pattern or with loose cellularity in a reticular array. Schwannoma stains are positive for S-100 and vimentin. Schwann cells stain negative for CD117, CD34, actin, and desmin differentiating it from the gastrointestinal stromal tumor (GIST) and leiomyomas.¹¹

Schwannomas are insensitive to chemotherapy and radiation therapy. Esophageal schwannoma is managed by endoscopic or surgical procedures. Smaller tumors up to 2cm are best managed by per-oral endoscopic tumor resection (POET) or VATS enucleation and larger tumors require VATS or Open enucleation.^{4,12,13} However, larger tumors where submucosal plane can't be achieved, esophagectomy either by minimally invasive or by open

approach is recommended.^{14,15}

In our case, the preoperative diagnosis couldn't be made because of the unavailability of EUS at our institution. An attempt for enucleation was done but due to dense adherent mucosa, we proceeded with VATS esophagectomy. For a large tumor, like in our case, located in the upper esophagus adjacent to the trachea and major vessels, VATS esophagectomy seems to be the ideal approach.

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