CASE REPORT

# A case report of large cervical oesophageal schwannoma: Importance of preoperative diagnosis in influencing surgical approach

**Wan Najmi Wan Daud, MMed Surg[1], Shazril Imran Shaukat, FRCR[2], Ahmad Khadri Awang, MMed Surg[3]**

[1]Department of General Surgery, KPJ Penang Specialist Hospital, Malaysia, [2]Advanced Medical and Dental Institute, Universiti Sains Malaysia, Malaysia, [3]Department of Cardiothoracic Surgery, KPJ Penang Specialist Hospital, Malaysia

## SUMMARY

**We present two cases of cervical oesophageal submucosal polypoidal growth from oesophagogastroduodenoscopy. Tunnelled biopsy was performed and subsequent immunohistochemistry was confirmatory of benign oesophageal schwannoma. Endoscopic ultrasound and contrasted computed tomography (CT) neck and thorax further formed preoperative diagnostic workup. Tumour enucleation was performed via left cervical incision for the first case. The second patient underwent thoracoscopic mobilisation, laparotomy, stomach pull up and cervical oesophageal resection. Both of them remain well till date.**

## INTRODUCTION

Schwannomas are relatively rare, with many case reports arising in Asia. Different surgical modalities could be considered depending on disease pathology, of which, the malignant lesion would require a more radical surgery than a benign tumour. Knowing definite histology upfront can be challenging hence, therefore, it poses a daunting task for surgeons to outline the best management strategies.

## CASE PRESENTATION

*Case 1*

A 31-year-old lady presented with dysphagia of 3 months duration. Upper endoscopy demonstrated a polypoidal tumour at 18 cm from upper incisor (Figure 1). Contrasted CT showed a well-circumscribed mildly enhancing homogenous tumour at the level between T2 to T3 thoracic spine measuring around 2.1 - 4.1 - 4.4 cm in diameter (Figure 2). A minor compressive effect was seen on the adjacent common carotid and brachiocephalic vessels. An endoscopic ultrasound further characterised the tumour being likely arising from the submucosal layer at the posterior wall. Initial biopsy showed spindle cell tumour of the oesophagus with occasional mitosis seen.

Subsequent immunohistochemistry confirmed benign oesophageal schwannoma with S-100 protein which was intense and diffusely positive. CD34 was weak and less intense. CD117, DOG1, smooth muscle actin, desmin, caldesmon, ALK1, STAT6, CKAE1/AE3 and LCA were all negative. The proliferative index estimation by Ki-67 was low; 1%.

Tumour enucleation was performed through 6 cm left anterolateral cervical incision. Oesophagotomy was closed in two layers with Vicryl 3/0 sutures. Nasogastric feeding was commenced on the next day until the oral gastrograffin study performed on the third post operative day. She was discharged on the next day.

*Case 2:* A 41-year-old man who had odynophagia of 3 days duration. He apparently had dysphagia to solid food in the last 2 years. Oesophagogastroduodenoscopy showed large oesophageal submucosal polypoidal growth with undigested food from 18 to 25 cm from upper incisor. PET-CT scan showed a well circumscribed supracarinal tumour from T1 to T5 vertebral body measured 4.0 cm (AP) -5.2 cm (W) - 6.8 cm (CC). There were also enlarged paraoesophageal and supraclavicular lymph nodes measured 19 mm and 15 mm respectively (with significant FDG tracer uptake). Immunohistochemistry confirmed the preoperative diagnosis of oesophageal schwannoma following a biopsy. Further management was discussed in regional Upper GI Surgery multidisciplinary teams to choose the appropriate surgical approach. Thoracoscopic mobilisation, laparotomy, gastric pull up, nodes dissection and cervical oesophageal resection were performed. Single layer hand sewn oesophagogastric anastomosis using Vicryl 3/0 sutures were done. He developed anastomotic leak and vocal cord palsy. No exploration done as he was not septic. Enteral feeding was prescribed using a nasojejunal tube at home along with vocal cord strengthening exercises. He recovered completely both for anastomotic leak and vocal cord palsy after 6 months. After 3 years of surgery, he is currently in good health.

## DISCUSSION

Schwannomas of gastrointestinal tract commonly occur in the stomach followed by small bowel, rectum and rarely oesophagus. Gastrointestinal schwannomas are mostly benign, but there is only single-digit number of malignant oesophageal schwannoma cases reported. It commonly occurs in the upper thoracic segment of oesophagus and up to date, less than 50 cases of oesophageal schwannoma were reported in English literature.

Essentially, a definitive diagnosis can only be established through histology combined with immunohistochemistry,

*This article was accepted: 19 May 2024*
*Corresponding Author: Wan Najmi Wan Daud*
*Email: drnajmi@yahoo.com*

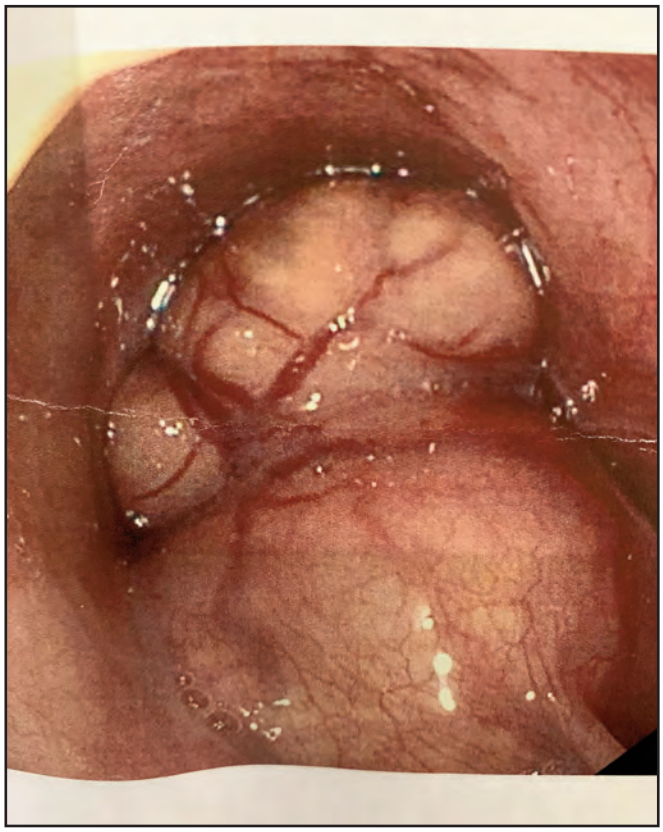


**Fig. 1:**



**Fig. 2:**

the latter being confirmatory. Dysphagia is the commonest presentation and rarely other symptoms like respiratory distress from airway compression, pneumonia and occasionally incidental finding from radiographs.

Oesophageal schwannoma is one of the differential diagnoses of oesophageal spindle cell tumours. In current practice, often an initial biopsy is done with immunohistochemistry being requested upon clinician's request. Hence, it is important not to assume all spindle cell tumours are malignant as this may influence surgical management. Furthermore, radiologically these tumours are indistinguishable. Establishing a diagnosis before resection is important as this may favour less invasive treatment.

Contrast-enhanced computed tomography scan (CECT) is the current imaging of choice and perhaps has replaced barium swallow in most centres. In contrast, positron emission tomography scan (PET) though widely used to stage oesophageal cancers, does not seem to provide any extra information to clinicians in any stage of the perioperative workup (1,4). Oesophageal schwannoma elicits high FDG uptake similar to malignant lesion despite being benign, hence making PET-CT, not a useful investigation to accurately distinguish between a benign or a malignant oesophageal lesion.

Flexible esophagogastroduodenoscopy allows the endoscopist to examine the oesophageal lumen particularly any mucosal involvement. One of the biopsy techniques for submucosal tumour as proposed by few authors is tunnelled biopsy. Commonly superficial bites lead to unsatisfactory reports. On the other hand, an imaging-guided biopsy is an alternative option if deemed feasible in terms of tumour size, location, relations to vascular structures and no other contraindications. We embarked on the former technique because more tumour tissue can be obtained under direct vision and haemostasis can be secured confidently.

Endoscopic ultrasound (EUS) is getting more popular especially to demonstrate and delineate the tumour origin, size and probability of its invasiveness[1,5]. Besides, it is also used as a part of the staging procedure to look for any lymph node enlargement and able to demonstrate layers of tumour involvement. In recent advances, tissue sampling can be obtained through EUS-fine needle aspiration (EUS-FNA), fine needle biopsy or trucut biopsy[3]. Diagnostic accuracy with EUS-FNA was reported at 85.2%. Tumour size of >2 cm , gastric tumour, large needle size, a high number of needle passes and the presence of onsite cytopathologist were favourable factors to obtain satisfactory tissue sampling.

Immunohistochemical (IHC) staining emerges as the utmost informative results in the histology report. S100 is positive in all cases of oesophageal schwannoma, suggesting nerve sheath origin. These stains characterise soft tissue tumour accurately with a huge impact on the preoperative plan. GIST is one of the main differentials of oesophageal schwannoma. CD117 and DOG1 which are confirmatory for GIST are negative for schwannoma. GIST patients with a massive or complex tumour where tyrosine kinase can be used to downsize GIST and minimise the extent of organs' resection

i.e. oesophagectomy for oesophagus or pancreaticoduodenectomy for duodenal tumour. Similarly, lymphoma of the oesophagus although it is rare, it responds well to chemotherapy which would preclude the patients from any major oesophageal resection. For oesophageal spindle cell sarcoma, radiotherapy would have a role as neoadjuvant or adjuvant even as a primary modality in selected cases, with durable long-term outcome, have been reported.

Complete surgical excision is the gold standard in treatment of oesophageal schwannomas and the prognosis of patients is excellent since these lesions are mostly benign. There have been various reports over the years discussing the optimal surgical treatments for oesophageal schwannoma from minimally invasive procedures to major resections. Several authors had reported a successful feasibility of endoscopic removal for small well-demarcated lesions. It can either be done by polypectomy, endoscopic mucosal resection (EMR) or endoscopic submucosal dissection (ESD) for lesions smaller than 15 mm in different parts of the oesophagus. On the other hand, a more aggressive approach through oesophagectomy has been proposed to get the adequate margins especially for bigger tumours or when anticipating some difficulty to close oesophagotomy securely.

Essentially, if the preoperative diagnosis has been confirmed, enucleation perhaps is the procedure of choice even for bigger tumour up to 87 mm[1,2]. The logic reason for this is because achieving clear margins and tumour recurrence are never a significant issue in benign tumours. Tumour enucleation is quite technically feasible because oesophageal schwannoma does not usually involve all layers of the oesophageal wall and is typically limited to the submucosa. Serious morbidities of oesophagectomy such as anastomotic leak, chylothorax and conduit necrosis are not worthy for small benign tumours. Nonetheless, when there is suspicion of malignancy, such as tumour larger than 10 cm, biopsy with mitotic figures, oesophageal muscle invasion or cellular atypia; oesophagectomy is worth to consider preventing recurrences or lymph node metastases.

Whilst cervical incision is the preferred approach for cervical oesophageal tumour, right thoracotomy or thoracoscopy is the procedure of choice for thoracic tumours. Tumour of the second case was bigger, adjacent to carina with significant enlarged nodes hence raised suspicion of malignancy. Since PET-CT scan is not always conclusive, we believe it was necessary to perform resection than enucleation to get clear margins with complete lymphadenectomy. Unfortunately, he developed anastomotic leak and likely traction nerve injury. Having more experienced-trained surgeons and with recent advances in minimally invasive surgery, has led to more cases being operated through video-assisted thoracoscopic surgery (VATS). A recent trend is to combine with endoscopic assistance to locate the tumour or with intraluminal dissection. Laterality (position) of the tumour is important for the surgeon to determine the side of thoracoscopic access. Generally left thoracoscopic approach is limited by the aortic arch and main bronchus. Nonetheless, Nakano et al from Japan had reported novel endoscopic assisted left thoracoscopic enucleation for large mid-thoracic schwannoma without any major morbidities[5]. More evidence with a better quality of trials are needed to determine whether minimally invasive procedures can be safely recommended as the standard procedure especially for benign thoracic tumours.

## CONCLUSION

Establishing preoperative diagnosis in submucosal oesophageal tumours is imperative to outline subsequent management particularly in selecting surgical approach and anaesthetic preparation.

We embarked a comprehensive preoperative workup for both patients which include endoscopy with good tissue sampling technique, CT scan and endoscopic ultrasound. The role of immunohistochemistry is definitive in establishing a diagnosis for submucosal oesophageal spindle cell tumours preoperatively.

Enucleation can be considered in a small benign oesophageal schwannomas though larger tumours may require oesophagectomy.

## REFERENCES

1. Zhu L, Li W, Zhu Z, Chai Y.Benign esophageal Schwannoma : a case report and review of literature. Niger J Clin Pract 2019; 22: 731-3.
2. Sachez RE, Ruben C, Alexandra RL, Emmanuel CJ, Jorge HR, Jesus MM. Esophageal schwannomas: A rarity beneath esophageal tumors a case report. Int J Surg Case Rep 2019; 58: 220-3.
3. Moon JS. Endoscopic ultrasound guided fine needle aspiration in submucosal lesion. Clin Endos 2012; 45: 117-23.
4. Jeon HW, Kim KS, Hyun KY, Park JK. Enucleation of giant esophageal schwannoma of the upper thoracic esophagus : reports of two cases. World J Surg Oncol 2014; 12: 39.
5. Onodera Y, Nakano T, Takeyama D, Maruyama S, Taniyama Y,Sakurai T. Combined thoracoscopic and endoscopic surgery for a large esophageal schwannoma. World J Gastroenterol 2017; 23(46): 8256-60.