Abstract: Vagal schwannoma are extremely rare neoplasm. Occur between 3rd and 6th decades of life. Both sex equally affected. Most often presents as painless, slow growing, lateral neck mass. Hoarseness, pain, or cough may be the presenting complaints. Paroxysmal cough may be produced on palpating the mass-clinical sign unique of vagal nerve schwannoma. It displace the carotid arteries anteriorly and medially, jugular vein laterally and posteriorly. These swellings are mobile transversely but not vertically. Imaging plays a central role in diagnosing vagal nerve neoplasm and in particular Magnetic Resonance imaging (MRI) has become the routine imaging study for these tumours. Treatment is complete excision with preservation of neural pathway. We are describing here such a rare case of cervical Vagal schwannoma in a 21 years old female, presented with a swelling in the right side of neck of 3 years duration, she had classical paroxysmal cough on palpation. MRI neck showed features of right Vagal schwannoma. FNAC reported as benign spindle cell neoplasm. Per-operatively, 5x7 cm tumor situated antero-laterally to right carotid sheath arising from vagus nerve, great vessels displaced medially. Enucleation of the tumor was done. post-operative period was uneventful with normal voice. Histo-pathology was suggestive of Schwannoma.

Keyword: ‘Schwannoma’, ‘vagal nerve tumors’,

Introduction:
Schwannoma arising from the vagus is an uncommon (2–5%) benign nerve tumour. This tumour most often presents as a slow growing asymptomatic solitary neck mass, which rarely undergoes malignant transformation. Investigations such as FNAC have low specificity. The carotid artery and internal jugular vein may be displaced antero-laterally. Diagnosis is based on clinical suspicion and confirmation obtained by means of surgical pathology. Schwannomas of the vagus nerve must be differentiated from the carotid body and glomus vagale tumors because the distinction may influence treatment planning. Surgical excision is the treatment of choice for vagal schwannoma, with recurrence being rare. We describe a case of vagal schwannoma in a 21 years female who presented to our unit recently.
Schwannomas are rare benign tumors that originate from the Schwann cells of the nerve sheath. Schwann cells are neural crest-derived glial cells that are responsible for providing myelin insulation to peripheral nervous system axons; about one third occur in the head and neck region. Clinically, they present as asymptomatic slow-growing lateral neck masses that can be palpated along the medial border of the sternocleidomastoid muscle. Mostly asymptomatic, hoarseness may be present. Occasionally, a paradoxical cough may be produced on palpating the mass. This is a clinical sign, unique to vagal schwannoma. Presence of this sign, associated with a mass located along the medial border of the sternocleidomastoid muscle, should make clinicians suspicious of vagal nerve sheath tumors. Pre-operative diagnosis of schwannoma is difficult because many vagal schwannomas do not present with neurological deficits and several differential diagnoses for tumors of the neck may be considered, including paraganglioma, branchial cleft cyst, malignant lymphoma, metastatic cervical lymphadenopathy, carotid body tumor and glomus tumor. Furthermore, due to their rarity, these tumours are often not even taken into consideration in the differential diagnosis.

In terms of preoperative investigations, FNAC, US, and radiographic imaging with CT or MRI are usually performed. However, schwannomas are frequently difficult to characterize on FNAC. Liu et al. reported that the accuracy of FNAC was only 20%. In our case, FNAC was inconclusive on the first time, and even with incision biopsy it was misdiagnosed as tuberculous pathology. On noncontrast CT, it was reported that schwannomas were typically hypodense versus muscle; with contrast, these lesions tended to show some peripheral enhancement. On the other hand, MRI consistently identifies these lesions on both T1- and T2-weighted imaging. T1-weighted images display low signal intensity, and T2-weighted images show high intensity. Hirano et al. reported that MRI was useful for the diagnosis and peripheral hyperintense rim with central low intensity on enhanced T1 images of MRI. The relationship between the schwannoma and its nerve of origin can be better appreciated with MRI than CT. In addition, MRI appears to be the investigation of choice for diagnosis and identification of nerve of origin. These results indicate that MRI is most sensitive and specific in the diagnosis of schwannoma. The MRI appearance is considered quite typical and may lead to suspicion of the diagnosis pre-operatively as the cervical vagal schwannoma frequently appears as a well-circumscribed mass lying between the internal jugular vein and the carotid artery. As reported by Furukawa et al., MRI findings are also useful in providing a preoperative estimation of the nerve of origin of the schwannomas and to differentiate pre-operatively between schwannoma of the vagus nerve and schwannoma of the cervical sympathetic chain. The vagal schwannomas, in fact, displace the internal jugular vein laterally and the carotid artery medially, whereas schwannomas from the cervical sympathetic chain displace both the carotid artery and jugular vein without separating them. In our case, the criteria of Furukawa et al. were present. Treatment of vagal nerve tumours is complete surgical excision. At surgery, these tumours appear as yellowish-white, well-circumscribed masses. Dissection of the tumour from the vagus with preservation of the neural pathway should be the primary aim of surgical treatment for these tumours.
Incomplete treatment, such as open biopsy, should be avoided, since it makes definitive excision of the tumour much more difficult. If it is impossible to find an adequate plane and is technically difficult to preserve the integrity of the nerve trunk, the involved segment may be resected and an end-to-end anastomosis performed using microsurgical techniques. This type of procedure often results in definitive vocal cord paralysis.

The reported incidence of pre-operative vocal cord paralysis is about 12%, but hoarseness is almost always present following surgery. Therefore, pre-operative assessment of vocal cord mobility should be strongly recommended. Although it is very rare, clinicians should bear in mind the possibility of a nerve sheath tumour in the presence of a neck mass. Pre-operative suspicion is very important, because the patient, and the patient’s family, should be informed about the possible post-operative neurological complications, as far as concerns post-operative vocal cord palsy, an incidence of 85% has been reported. Furthermore, since vagal schwannomas are almost invariably benign in nature, a conservative approach should always be considered in first instance. In the presence of post-operative vocal cord palsy, aggressive voice therapy, for vocal cord compensation, should be started soon after surgery. Surgical excision is the treatment of choice, but slow growth and the noninvasive nature of schwannomas of the neck also allow an observational approach. The preferred method of removing a schwannoma is intracapsular enucleation. Complications are usually transient and in most cases do not require treatment. According to the study by Valentino et al., intracapsular enucleation while preserving the nerve fibers preserved its function by more than 30% when compared to complete tumor resection. These results suggested that intracapsular enucleation was an effective and feasible method for preserving the neurological functions. Conclusion:

Cervical schwannomas are rare neck tumors that are not widely discussed in the core surgical literature. Surgeons who evaluate neck masses need to be aware of the diagnostic work-up, surgical treatment, and likely complications of this pathology. In addition, treatments assuring the preservation of neurological functions are needed, since surgical resection may cause significant nerve damage unlike other tumors.

References: