Otolaryngology online journal

ISSN: 2250-0359 **Volume 4 Issue 4 2014**

Cervical Vagal Schwannoma and cable grafting of vagus nerve: A Rare Case Report and Review of Literature

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Abstract:

Schwannoma or Nerve sheath tumors arising from the cervical vagus nerve are extremely rare. Usually presents as asymptomatic neck swelling in the age group of 3rd and 4th decades with no sex predilection. Very rarely presents with hoarseness of voice due to nerve compression. The diagnosis can be done by radio logically on CECT and MRI. Surgical excision with preservation of nerve function when ever feasible is the ideal treatment. Here we discuss a case of cervical vagal schwannoma in a 36 year old male which was excised en block and with sacrifice of nerve and nerve repaired by cable graft using great auricular nerve. The presentation, diagnosis and difficulty encountered in capsular dissection are discussed along with option for reconstruction of the sacrificed vagal nerve is discussed.

Key words: Cervical vagus nerve, Schwannoma, Excision, Nerve grafting

Introduction:

Schwannomas are the benign tumors arising from the nerve sheath. Approximately 25-40% [1] of all cases of schwannoma occurs in the head and neck region[1]. Schwannomas of head and neck have predilection of involvement in following sites: eighth cranial nerve (most common), followed by cranial nerve IX, XI, XII, the sympathetic chain, cervial plexus and brachial plexus[2-5]. Parapharyngeal site is the most common site of tumor origin (31%) followed by neck (23%), skull base (19%), sinonasal cavity (15%), middle ear (8%), tongue, posterior pharynx (4%) [6].

In contrast, cervical vagal nerve schwannoma is extremely rare[7].

3rd to 5th decades are most commonly involved age group with no sex predominance. Slowly progressive painless swelling in the upper lateral of the neck is the usual mode of presentation, but very rarely preoperative compression symptoms like hoarseness of voice may be seen. MRI is the radiological investigation of choice to diagnose and planning for surgery. Trans-cervical approach with enucleation and dissection of tumor off vagus nerve and preservation of nerve trunk is the treatment of choice. But is it feasible in all vagal nerve schwannoma? We report a case of cervical vagal nerve schwannoma in which it was not possible to shell out the tumor from the nerve even under microscope. So enbloc removal of mass along with sacrifice of vagal nerve was done for which cable nerve graft using great auricular nerve was done.

Case report:

A 38 year male presented to our outpatient department with complaints of slowly progressive painless swelling in right upper neck for two years. There was no history of hoarseness of voice, recurrent cough, difficulty in swallowing, nasal regurgitation and associated pain over the swelling.
On examination there was a single 2/2 cm firm, mobile, non tender, non fluctuant, non compressible nontrancluscent, pulsatile (transmitted pulsations) swelling just 1 cm below the angle of mandible in front of upper 1/3rd of anterior border of right sternocleidomastoid. The swelling was mobile in transverse plane but non mobile in vertical plane. Carotids were palpable anterior to swelling (fig 1).

Contrast enhanced computed tomography of neck (CECT) showed relatively homogenous, non enhancing soft tissue lesion in right carotid triangle region displacing carotid artery anteromedially and internal jugular vein (IJV) posterolaterally(fig 2). MRI with gadolinium of neck showed well defined, homogenous, hyper intense lesion in right carotid triangle with similar displacement of the great vessels. CT angiography showed displacement of common carotid artery anteromedially and posterolateral displacement of internal jugular vein by the mass. Fig 3.

Fine needle aspiration cytology (FNAC) was suggestive of spindle cell tumor. Excision of the mass in the neck was done under general anesthesia. Six cm trancervical incision was made in the right side of neck, 2 finger breadth below the angle of mandible. Intra-operatively a 3X3 cm well encapsulated, yellowish white, ovoid shaped mass found between common carotid artery and internal jugular vein. The internal jugular vein was pushed posterolaterally and was compressed with mass but could be separated. With meticulous dissection the mass was found to be arising from cervical

part of vagus nerve. An attempt at sub capsular dissection using microscope was made to enucleate the tumor and preserve vagus nerve. The vagus nerve was stretched and thinned was integrated with tumor capsule on dorsal side. It was difficult to separate fascicles of nerve from the tumor capsule, so part of the nerve was resected at both the ends to deliver the tumour ‘en bloc'.

Approx 3.5 cm segment of vagus nerve was deficient. An attempt to cable graft using great auricular nerve was made. Post operatively there was restricted right true cord mobility with no features suggestive of aspiration at two month follow up (fig 5).

. Histopathological examination showed encapsulated tumor showing hypo cellular and hyper cellular areas. Tumor cells were spindle shaped, having elongated kinky nucleus and inconspicuous nucleoli with moderate scant cytoplasm. Occasional verocay body formation was seen. Neoplastic cells revealed nuclear and cytoplasmic immunopositivity with S-100 and were negative for smooth musle actin(SMA) and diagnosis of schwannoma was confirmed.

Discussion:

Schwannomas are slow-growing, well encapsulated, solitary tumours arising from nerve sheath of any cranial or spinal nerve that has a sheath (optic and the olfactory nerves do not have the schwann cell sheath). Schwannoma was first established as a pathological entity by Verocay in 1908 who later called it neurinoma in 1910 [8]. Later the term neurilemmoma was coined by Stout in 1935 [9]. Approximately 25–45% of all the reported neurilemmomas occur in the head and neck and most of them are in the eighth nerve[1]. The most common site of the extracranial schwannomas in the head and neck region is the parapharyngeal space[8, 9]. Other reported sites of origin of cervical schwannomas are the cranial nerves IX–XII, the sympathetic chain, the cervical plexus, and the brachial plexus. But schwannoma arising from cervical part of vagus nerve is very rare.

The diagnosis is very difficult as most of them presents as asymptomatic slow-growing lateral neck swellings along the medial border of stenocleidomastoid without any neurological deficits. Cervical lymphadenopathy, malignant lymphoma, carotid body tumor, branchial cyst, as well as aneurysm of neck have to be considered in the differential diagnosis. Rarely, when vagal schwannoma is symptomatic, hoarseness is the most common presenting symptom and paroxysmal cough may be produced on palpating the mass. This sign is unique to vagal schwannoma and clinches the diagnosis. However our patient did not have hoarseness nor had paroxysmal cough on palpation of swelling (10).

Radiological imaging with CECT and MRI play an important role in the diagnosis and help to correlate the relation of tumor to surrounding vascular structure, nerves and muscles. On CECT vagal schwannoma typically appear as homogenous non enhacing lesion, rarely a peripheral enhancement with typical displacement of carotid artery anteromedially and IJV posteriorly.(11)). MRI appearance of vagal schwannoma is unique and may suggest preoperative diagnosis in absence of

FNAC report. On MRI, vagal schwannoma seen as well circumscribed ovoid enhancing mass lying between Jugular vein and carotid artery appearing hypo intense on T1 weighted and hyper intense on T2 wieghted image . According to Furukawa et al. the site and the way the major neck vessels are displaced could give further clues on the nerve of origin of the schwannoma in tha carotid space. Vagal schwannomas typically separate the internal jugular vein laterally and carotid arteries medially but do not usually widen the carotid bifurcation. On the contrary, sympathetic chain schwannomas displace both carotid and IJV anteriorly without separating them. The splaying of the carotid bifurcation is usually more prominent in carotid body tumours. This is called the "lyre sign," which, along with significant contrast enhancement, is rarely associated with schwannoma. (13)

Schwannomas of neck have been classified into 4 types based on macroscopic nature. In type 1, the nerve of origin can be identified from the tumor. In type 2, normal nerve passes on the surface of the tumour. In type 3 nerve fibers are slightly dilated on tumor surface. In type four, tumor surface is covered by thinned out nerve fibers (14). The nerve can be easily dissected off tumor in types I and II. It may still be possible to preserve few functioning nerve fibers in type III. However it may not be possible to preserve functioning nerve fibers off tumor in type IV. In our case we could not find plane between nerve fibers and tumor, so the vagus nerve was sacrificed and nerve anastamosis with great auricular nerve was done.

Treatment of cervical vagal nerve schwannoma is complete surgical excision. The primary aim of surgery should be to dissect the tumor of the vagus nerve and preserve the nerve trunk. Fujino(2000) described the intracapsular enucleation technique for vagus nerve tumors, since then it has become the standard surgical method. Careful intra-capsular dissection of the vagal schwannoma and an attempt to shell out the tumor from the nerve should be made in all cases. According to Valentino et al intra capsular enucleation while preserving the nerve fibers preserved the nerve function by more than 30%. However, when it is not possible to separate the vagal nerve fibers from the tumor technically (as in our case), en bloc resection of the tumor with sacrifice of the nerve should be done.

Does tumor reoccur with intra capsular ennuleation? According to Zbären, et al., there was no significant difference in the recurrence rate between the total tumor resection including nerve fibers and the intra capsular enucleation. An attempt to reconstruct the nerve should be made and end-to-end anastomosis or cable nerve grafting (as in our case) performed using microsurgical techniques (10). The post operative results of nerve anastomosis are uncertain. Post operatively hoarseness and homolateral vocal cord paralysis is seen in almost all cases. (15).

Since our case is type four schwannoma the nerve was sacrificed to resect the tumour and both cut ends were repaired using Greater auricular nerve as cable graft. Post operatively there was hoarseness and ipsilateral cord paralysis for which counseling was done preoperatively. However we found quick recovery of voice quality with speech therapy in 3 weeks.

Conclusion:

Vagal nerve schwannoma should be considered in the differential diagnosis of lateral neck mass and when associated with hoarseness and cough on palpation. MRI features suggest diagnosis and also can predict the nerve of origin of tumor. Preservation of integrity of vagal nerve fibers and complete excision of tumor is treatment of choice.

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Figures:

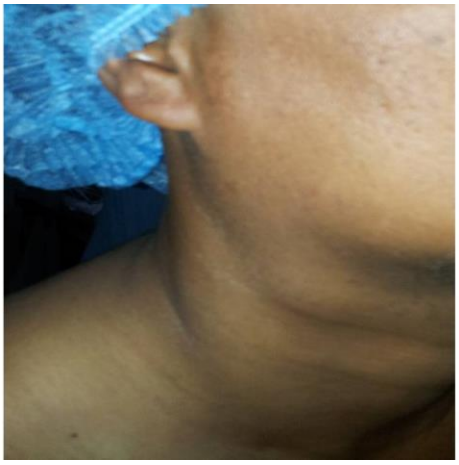
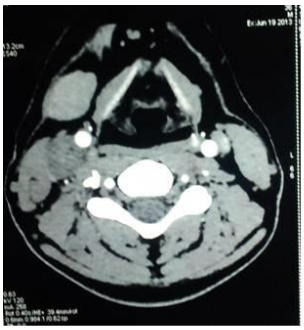


Fig 1: Preoperative photograph showing 2x2cm mass in the upper part of neck



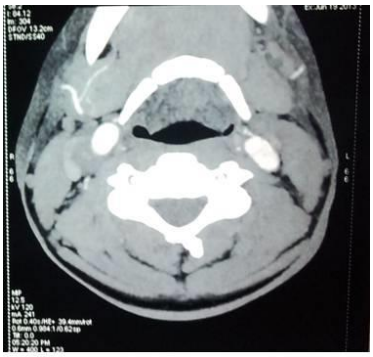
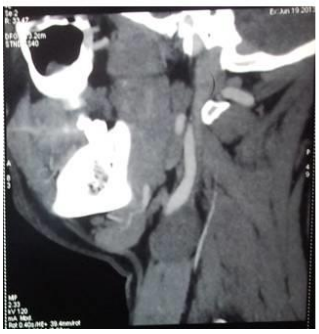


Fig 2a



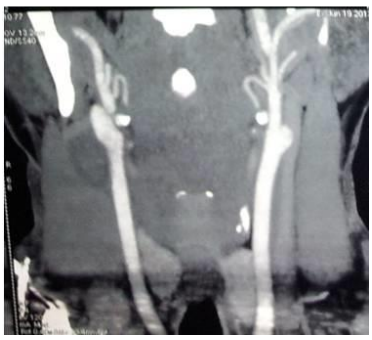


Fig 2b

Fig 2: Contrast enhanced CT Neck showing heterogeneous soft tissue tumor in the carotid space. 4b) CT angiography showing anterior and medial displacement of common carotid artery and lateral displacement of Jugular vein

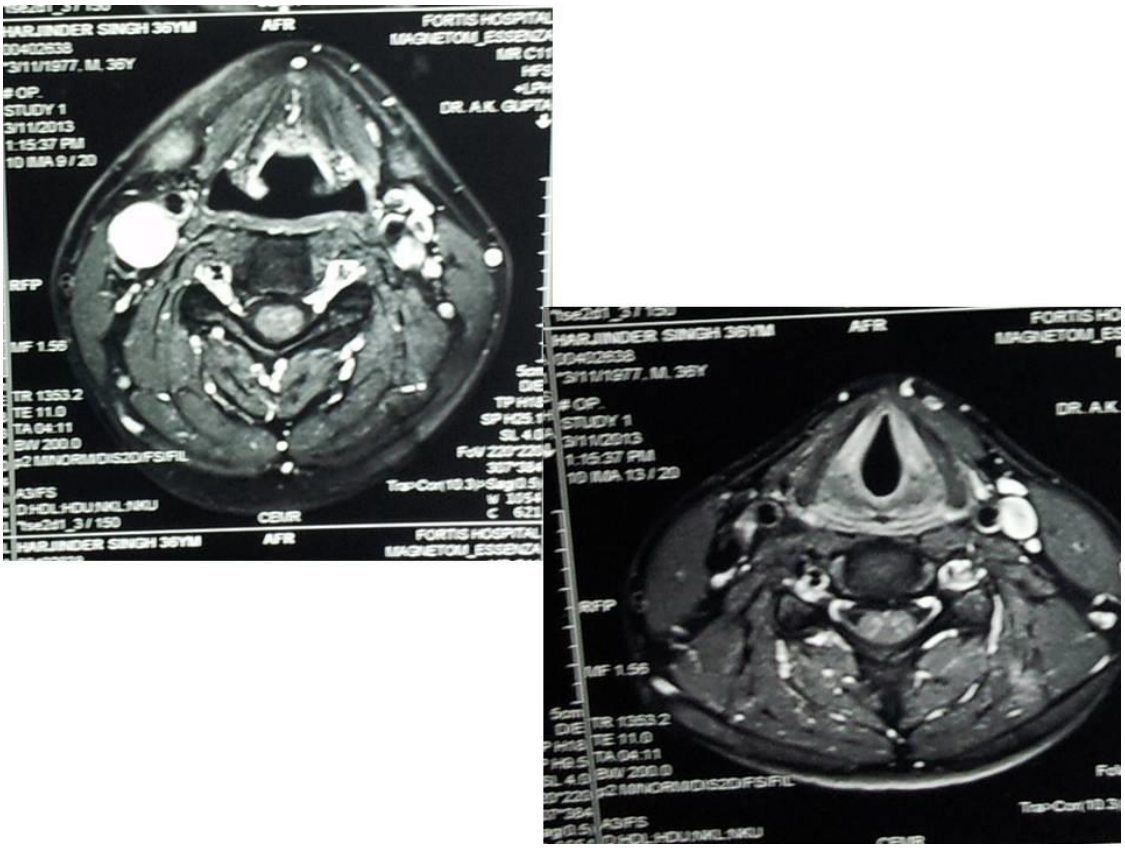


Fig 3 a and b) MRI showing T1 and T2 weighted showing ovoid mass in the carotid sheath arising from the vagus nerve.

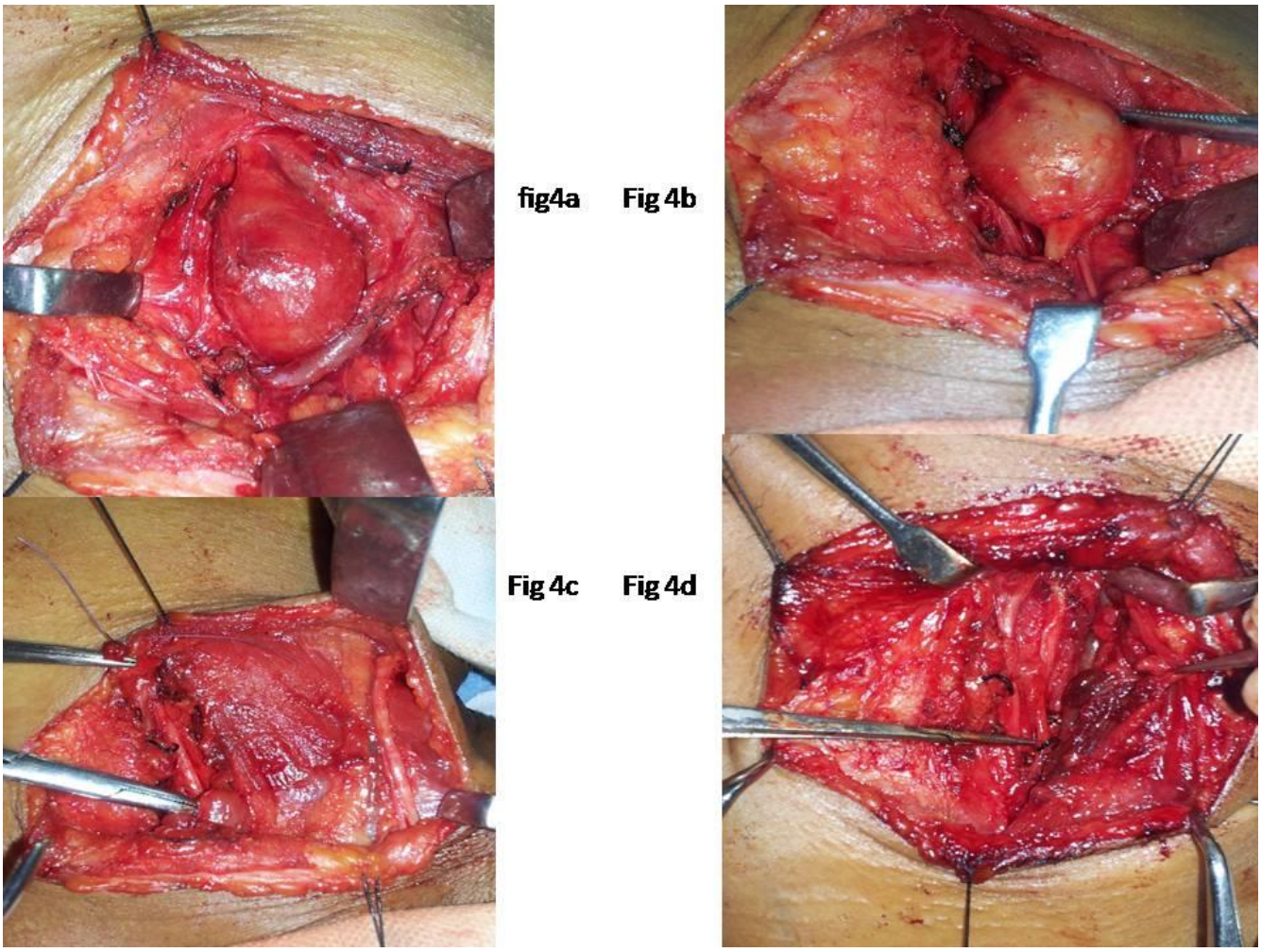


Fig 4: 4 a and b) Intra operative photograph showing well encapsulated vagal schwannoma with displaced common carotid artery and internal jugular vein.4c) intraoperative photograph showing cut ends of vagus nerve. 4d) shows great auricular nerve anastomosis with cut ends of vagal nerve.

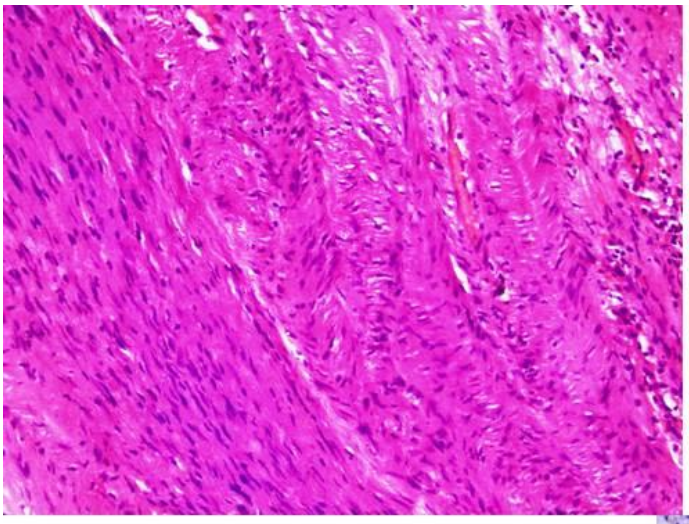


Fig 5a

Fig 5b

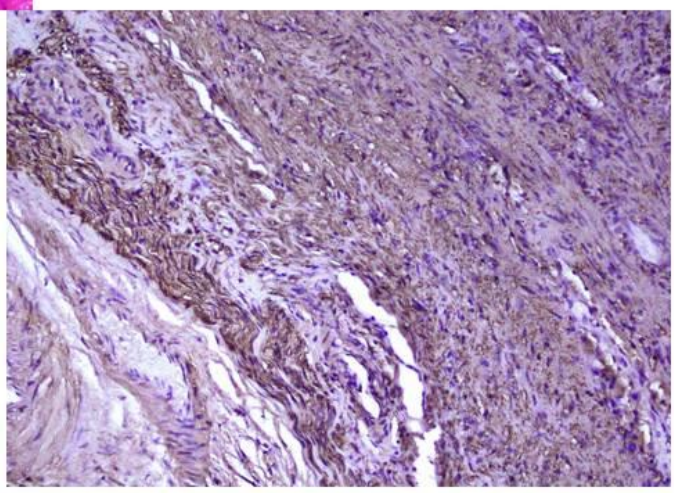


Fig 5: Histopathology 5a): H$E stain: hypocellular and hypercellular areas showing typically spindle shaped cells with occasional verocay body formation. 5b) S100 protein immune histochemical positivity of the schwannoma.