



Cervical Schwannoma of the Vagus Nerve: Diagnostic and Therapeutic Challenge

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ABSTRACT

Schwannomas arising from the cervical vagus nerve are extremely rare benign neurogenic tumours. They usually occur in ages between third and sixth decades, affect both sexes equally and most often present as painless, slow-growing, lateral neck masses. The treatment of choice is the complete surgical excision. We report our experience with the clinical presentation, diagnosis and management of a case of cervical vagal schwannoma.

Introduction:

Schwannomas are rare, benign tumours of nerve sheath origin that may originate from any of the cranial, peripheral or autonomic nerves (1). Cervical schwannomas arising from the vagus nerve are extremely rare. They typically present as painless, slow-growing, lateral neck masses in between 3rd and 6th decades, do not show sex predisposition (2). Malignant transformation is unusual for these tumours. Complete surgical resection is the treatment of choice (3). Imaging plays a pivotal role in pre-operative diagnosis of these tumours and in particular, magnetic resonance imaging provides important information for planning surgical excision (4). Here, we report the management and outcome of a case of cervical vagal schwannoma in a 30-year-old male who presented with a slow-growing upper lateral neck mass without any other clinical presentation.

Case Report:

A 30-year-old male presented with a painless, gradually increasing right sided neck mass of 1 year. There was no history of fever, hoarseness, nasal regurgitation, dysphagia, dyspnoea or compressive symptoms. There was no significant past medical or surgical history. Clinical examination revealed a mass in the right upper cervical region between the right mandibular angle and mastoid process; this was approximately 5 cm x 4 cm in diameter, non-tender, non-pulsatile, firm to hard, smooth surfaced, mobile only in the horizontal plane and lying deep to the sternocleidomastoid muscle. There was no paroxysmal cough on palpating the mass. No murmur was heard on auscultation. Oropharyngeal examinations did not reveal medial displacement of peritonsillar structures. 70 degree laryngoscopy did not reveal any vocal cord abnormality. Rests of the otorhinolaryngological examinations were normal. No cranial nerve deficits or Horner's syndrome were present. All routine investigations were within normal limits. Fine needle aspiration cytology (FNAC) of the mass was inconclusive. Contrast enhanced computed tomography (CECT) scan neck showed a well circumscribed partially enhancing mass lesion measuring 5 cm x 4 cm over the right side of the neck.

It extended from skull base above to the level of hyoid bone below. The mass splayed the right internal jugular vein (IJV) laterally and the right carotid arteries medially. The right IJV was compressed by the tumour (Fig. 1a, 1b). Patient underwent excision of the mass through a transcervical incision at the level of the hyoid bone under general anaesthesia. Per-operatively, a yellowish-white, ovoid-shaped mass, measuring approximately 5 cm x 4 cm was seen originating from the vagus nerve.

It was extending laterally to the internal jugular vein and sternocleidomastoid muscle, medially adjacent to the internal carotid artery, and superiorly to the skull base (Fig.2a, 2b). Both the superior and inferior ends of the mass were found in continuity with the vagus nerve. Since an adequate dissecting plane could not be reached, it was impossible to dissect the nerve trunk off the tumour. The tumour was resected "en bloc" along with the involved segment of the vagal nerve trunk, after mobilization from the internal jugular vein and the carotid artery (Fig. 2c, 2d). Microscopic examination of the mass showed focal hypercellular (Anthoni A) and hypocellular (Anthoni B) areas along with areas of haemorrhage and myxoid degeneration. Immunohistochemical examination of tumour cells showed strong S-100 positivity (Fig 3a, 3b). Therefore, a final diagnosis of vagal nerve schwannoma was obtained. Postoperatively, patient had hoarseness and 70 degree laryngoscopy revealed right vocal cord palsy. and prompt speech therapy was started. A repeat laryngoscopy on fifteenth post-operative day showed compensated right vocal cord palsy.

Discussion:

Vagal schwannoma mostly present as asymptomatic, slow growing, painless, neck mass without neurological deficit. When symptoms are present, hoarseness is the most common presentation.

Occasionally a paroxysmal cough may be produced on palpating the mass. Presence of this sign in association with a mass located along the medial border of sternocleidomastoid muscle should alert the clinicians towards vagal nerve sheath tumours (4). Horner's syndrome may be seen in a schwannoma of the cervical sympathetic chain. In our case, the schwannoma was seen arising from the vagus nerve and patient presented with neck mass without any other clinical features.

A definite pre-operative diagnosis of vagal schwannoma is difficult, as patients have few symptoms and most do not have neurological deficits. The role of FNAC in schwannoma is still controversial and most authors do not recommend it (5). Kang et al. reported that the cytological diagnosis of schwannoma was only definitive in 20% of cases, based on the observation of characteristic Verocay bodies and spindle cells (3). In our case, FNAC was inconclusive.

Imaging has an important role in the diagnosis of head and neck schwannomas. MRI findings of schwannomas include isotense or hypointense signal relative to the skeletal muscle on T1-weighted images and hyperintense signal on T2-weighted images. CECT of schwannomas typically reveal well delineated margins with a high attenuation than adjacent muscle but may be isodense, or, less commonly of lower attenuation than adjacent muscle. Approximately one third of the schwannomas enhance significantly on CT (6).

Furukama et al. found that vagal schwannomas displace the internal jugular vein laterally and the carotid medially, whereas schwannomas from the cervical sympathetic chain displace both the carotid and the jugular vein without separating them (7). In our case, the mass separated the right IJV laterally and carotid arteries medially.

Cervical vagal schwannomas must also be differentiated from carotid body and glomus vagale tumours because the distinction may influence treatment planning. Carotid body tumours arise at the carotid bifurcation, splaying the external and internal carotid arteries, whereas glomus vagale tumours usually displace the internal carotid artery anteriorly or medially or both. Both tumours enhance intensely on both CT and MR images and reveal a characteristic "salt-and-pepper" appearance on enhanced T1-weighted MR images. This salt-and-pepper appearance is not a feature of schwannomas.

Histologically, schwannomas exhibit two main patterns: Antoni A areas composed of compact spindle cells with indistinct cytoplasmic borders and Antoni B areas having looser Schwann cell proliferation. A mixed picture of both the type can exist. Other typical features include cystic degeneration, necrosis and haemorrhages. Immunohistochemical examination of the tumour cells show S-100 and neuron specific enolase (NSE) positivity. Similar histological finding was seen in our case.

Treatment of vagal schwannomas is complete surgical resection with preservation of the neural pathway whenever possible. Most lesions can be removed surgically without damage to involved nerve. For tumours arising from the major cranial nerves, complete tumour resection renders lifelong morbidity to the patients. On the other hand, the nerve-preserving excision method, such as intracapsular enucleation, does not guarantee intact nerve function after surgery (8).

Our patient underwent excision of the mass along with the involved segment of the nerve as the tumour was inseparable from the nerve trunk and preservation was technically difficult. The reported incidence of preoperative vocal cord palsy is about 12% but hoarseness is almost always present following surgery. Postoperative vocal cord paralysis has been reported in 85% (9, 10). Postoperatively, our patient developed hoarseness and ipsilateral vocal cord palsy, which got compensated after prompt vigorous speech therapy.

Conclusion:

In conclusion, vagal schwannomas presenting as asymptomatic unilateral neck masses, are rare benign tumours. Preoperative diagnosis relies on clinical suspicion and imaging. Final diagnosis depends on histopathological confirmation. Surgical excision is associated with high rate of postoperative vocal cord paralysis. In the presence of postoperative vocal cord palsy, voice therapy for vocal cord compensation should be started soon after the surgery.

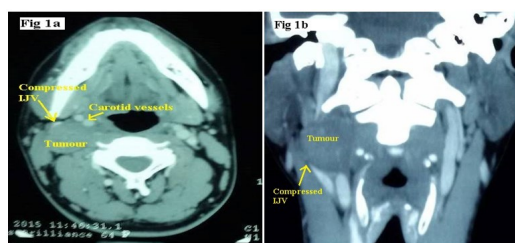


Figure: CT neck axial and coronal views showing tumour mass displacing right internal jugular vein (IJV) laterally and the right carotid arteries medially. IJV is seen

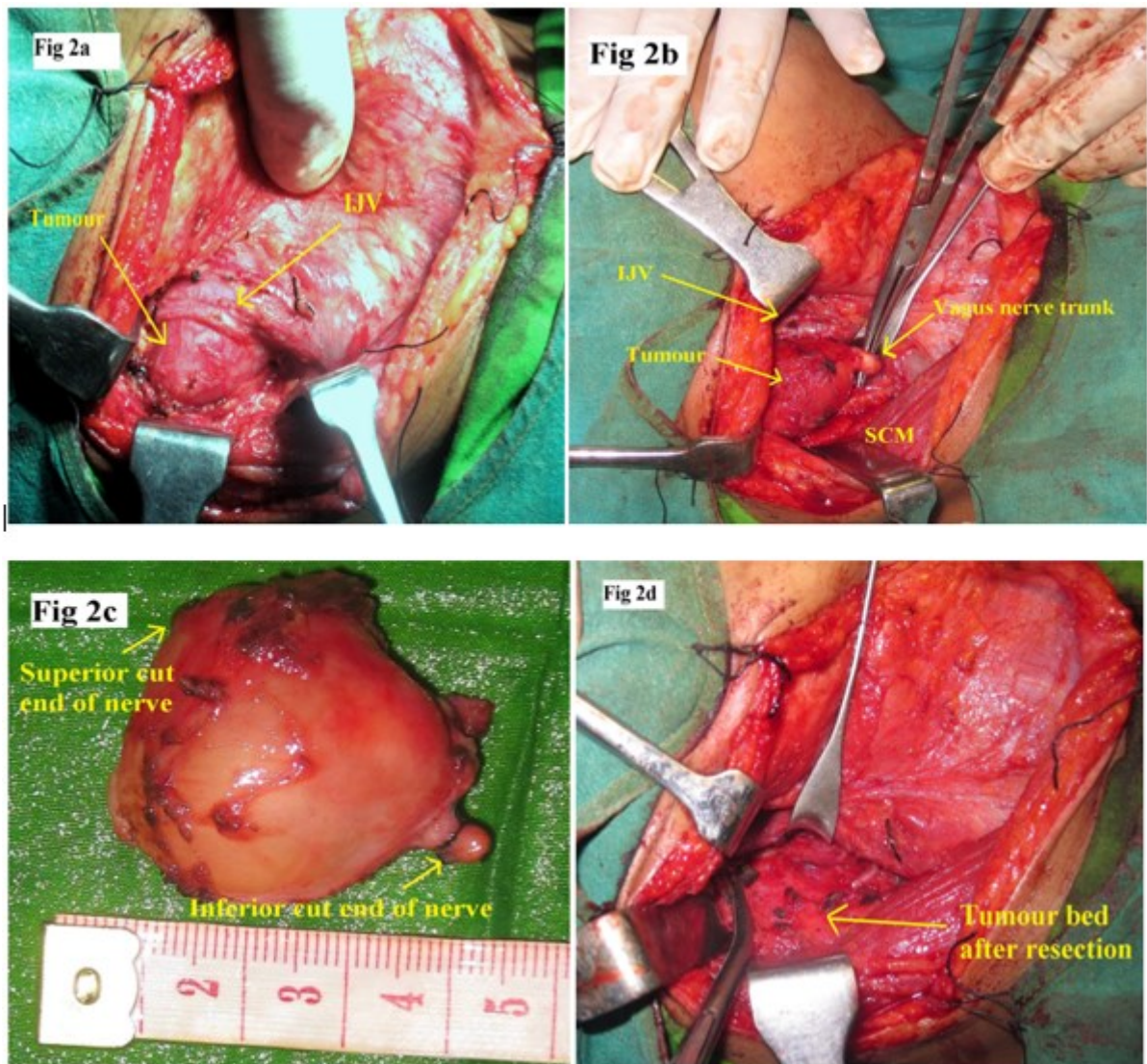


Figure: 2a) Showing tumour mass with IJV lateral to the mass. 2b) Tumour mass seen in direct continuity with right vagus nerve. 2c) Resected tumour mass with part of the vagus nerve trunk seen tied. 2d) Tumour bed after resection of the mass.

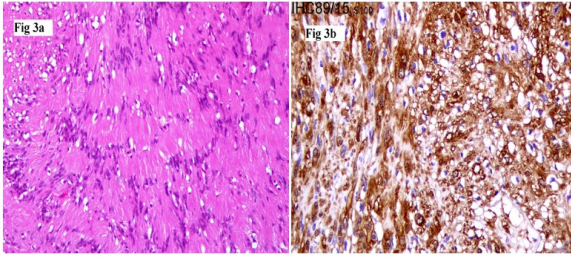


Figure: 3a) H & E staining showing hypercellular (Antoni A) and hypocellular (Antoni B) areas. 3b) Immunostaining showing S-100 positivity.

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