

Case Report

Schwannoma of the cervical vagus nerve: a rare case report

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ABSTRACT

Schwannomas, also called as neurilemmomas, are well-encapsulated, slowly growing tumors that arise from schwann cells of peripheral nerves. Clinically, schwannomas present as asymptomatic, slowly enlarging lateral neck masses and so they tend to present late. The nerve of origin is not often made until the time of surgery. Preoperative diagnosis is aided by imaging studies such as magnetic resonance imaging and computed tomography. The treatment of choice of schwannomas of the neck typically involves complete surgical resection. Rapidly growing tumors with evidence of invasion, or complete loss of nerve function should trigger complete surgical excision of the tumor. Here, we report a rare case of cervical vagal schwannoma in a 45 year old female who got admitted with the complaint of a firm, painless mass on the left side of the neck. The management of the case is discussed along with the relevant literature.

Keywords: Schwannoma, Neurilemoma, Vagus

INTRODUCTION

In the head and neck region various types of swellings appear, but neurogenic tumours arising from head and neck are quite rare and found more commonly in adult population. Schwannomas usually involve the cranial nerves 9 to 12 as well as brachial plexus. Because they grow at a rate of about 2-3 mm per year they present late in age groups 30 to 70 years.^{1,2} The term 'Schwannoma' first introduced in 1935 by Stout, who identified it as an benign tumour with sporadic malignant degeneration arising from cranial, peripheral and autonomic nerve sheath cells.³ Schwannomas are rare benign nerve sheath tumour composed of Schwann cells, that produce the insulating myelin sheath covering the peripheral nerves. These are also commonly referred as neuromas or neurilemmomas. About 25%-33% of of all the extracranial schwannomas occur in head and neck region.^{4,5} Vagal schwannomas are extremely rare homogeneous tumours lying over the vagus nerve, but the tumour itself can push the nerve aside and/or up against a bony structure thereby possibly causing damage to the

nerve. Vagal nerve schwannoma usually occurs between the third and fifth decades of life, and it has no sexual preference.⁶ It most often presents as a painless, slow-growing, lateral neck mass and are mostly benign with less than 1% degenerating into a form of cancer known as neurofibrosarcoma. The treatments of vagal schwannoma are wide with differing opinions because the close adherence of schwannoma capsule to nerve rendering it difficult to preserve the nerve. Hence, some recommend observing the tumour in case it is asymptomatic.⁷ The treatment of choice is, however, complete surgical excision with the preservation of the nerve. We report the case of a patient diagnosed with cervical schwannoma of vagal nerve origin.

CASE REPORT

A 40-year-old female patient presented to the Department of Otolaryngology at Mahatma Gandhi Memorial Hospital, Warangal with complaints of left sided neck swelling over the past six years. She did not give any history of hoarseness of voice, recent cough, or light

headedness, but complained of difficulty in neck movements. On examination, a firm, smooth, horizontally immobile, nontender swelling in the upper one third of sternocleidomastoid (SCM) measuring about 7×5 cms, located in the left carotid triangle. It extended from behind the SCM and vertically extended from angle of mandible to the level of thyroid notch while horizontally within the left carotid triangle (Figure 1). The skin over swelling appeared normal with no visible pulsations or engorged veins, sinuses or scars. Systemic examination was normal. Indirect laryngoscopy revealed obliteration of left pyriform sinus when compared to opposite side confirmed with Hopkins 70° endoscope (Figure 2). USG revealed 60 mm × 50 mm long well circumscribed oval mass with heterogeneous echo texture and hypoechoic mass occupying the left carotid space. Colour Doppler did not reveal any internal vascularity. FNAC of swelling revealed neurogenic origin of tissue. A contrast enhanced computer tomography showed a well-defined sharply margined low attenuated lesion in the left carotid space, displacing the carotid arteries anteromedially and compressing the internal jugular vein (Figure 3). It was associated with loss of intervening fat plane suggestive of a neurogenic tumor. The lesions also displaced the left SCM and left submandibular gland medially while the upper part of the mass abutting the ramus of mandible on left side and the lower level of the lesion at level of fifth cervical vertebrae (Figure 3 and 4). The larynx, thyroid gland and hypopharynx were normal.



Figure 1: Swelling located on left side of neck (arrow).



Figure 2: Endoscopic picture showing obliteration of left pyriform sinus (A) compared to opposite pyriform sinus (B).

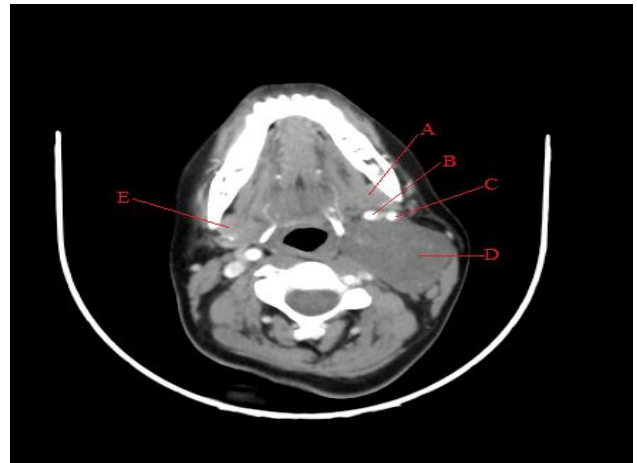


Figure 3: Axial CT showing Schwannoma on left side (D), pushing the ipsilateral submandibular salivary gland forward (A) compared to contralateral gland (E) and pushing the ICA (C) and ECA (B) anteromedially.



Figure 4: Coronal CT showing the schwannoma (D) compressing the jugular vein (F).

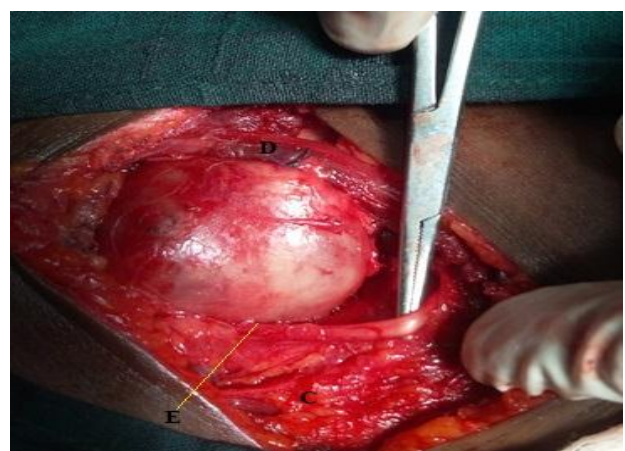


Figure 5: Schwannoma mass in relation to the vagus nerve (E) and strap muscles medially (D) and SCM laterally (C).

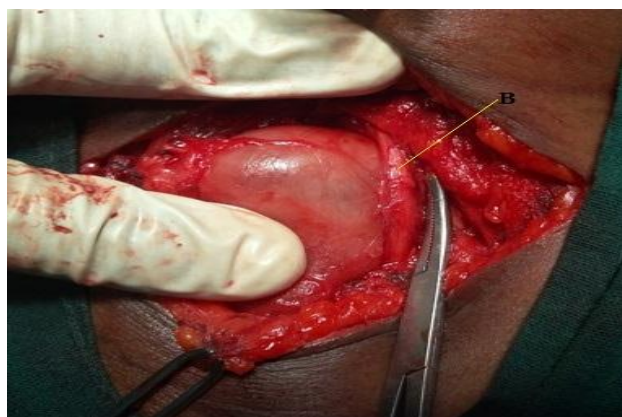


Figure 6: Posterior surface of schwannoma turned to reveal its relation to the vagus nerve (B) showing the nerve fibres intricately related to the tumour.



Figure 7: Cut section of the schwannoma measuring 6×5 cms revealing the characteristic yellow white tan.

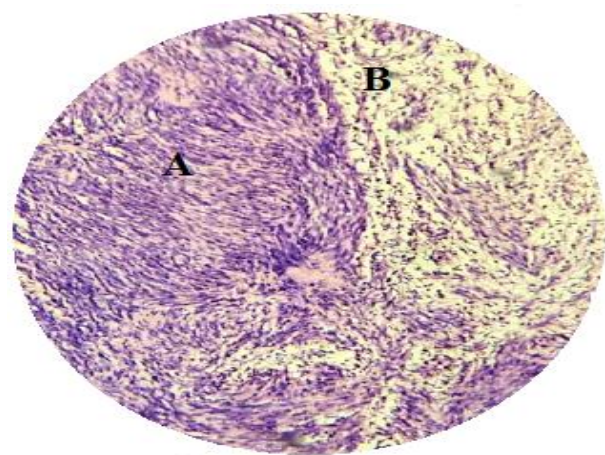


Figure 8: Histopathology of the specimen showing the hypercellular areas (A) called as Antoni A and adjacent to it, hypocellular areas, Antoni B (B).

Under general anesthesia, a transverse incision was made about 2 cms below the angle of the mandible extending

posteriorly up to anterior border of left SCM and anterior extent upto the midline. Following raising the flaps in subplatysmal plane, the straps were separated and lesion visualized. A well-defined smooth encapsulated yellowish mass was visualized in the carotid triangle (Figure 5). When it was dissected from the surrounding tissue, it was found to have posterior attachment to the vagus nerve of that side (Figure 6). The vagus nerve was splayed over the surface of the mass. The tumor was excised by intracapsular dissection technique thus sparing the left vagus nerve (Figure 7). Haemostasis was acquired and wound closed in layers after placing a suction drain. The patient had an uneventful postoperative period and was discharged on the 7th post op day. Post-operative endoscopy showed no evidence of vagal nerve injury and patient had no complains of hoarseness, or cough. Final histopathology report showed the presence of schwannoma with degeneration, composed of focal hypercellular (Antoni A) areas and hypocellular areas (Antoni B) (Figure 8). Immunohistochemical examination of tumor cells showed strong S-100 positivity (Figure 9).

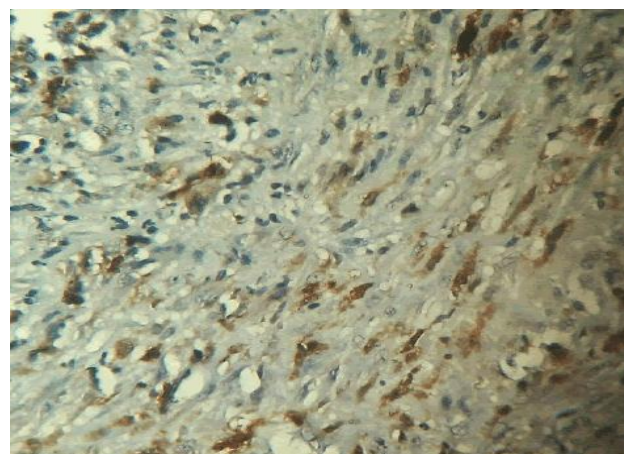


Figure 9: Showing Immunostaining with S-100 antibody which shows dark brown immunostained characteristic of schwannoma cells.

DISCUSSION

Vagal schwannomas are slow growing lateral masses that can rarely cause symptoms such as hoarseness of voice and cough. They present as a mass along the medial border of SCM. Because of their asymptomatic nature preoperative diagnosis is difficult with respect to nerve of origin. Therefore a possible differential diagnosis includes metastatic cervical lymphadenopathy, malignant lymphoma, carotid body tumor, branchial cyst, as well as aneurysm. Vagus nerve is reported to be the origin for 50% of parapharyngeal schwannomas and cervical sympathetic chain being the next common source.⁸ Schwannomas of cranial nerve origin are mostly from vestibulocochlear nerve and affect other cranial nerves such as trigeminal, facial, glossopharyngeal, vagus and spinal accessory in the descending order.⁹ Among cervical

schwannomas, vagus nerve is involved in approximately 10% of cases.¹⁰ Anatomic evaluation by direct vision intraoperatively is an acceptable way. According to Lin et al, the site of the schwannoma in the neck and the way it displaces the major neck vessels give clues on origin of the schwannoma.¹¹ Vagal schwannomas typically separate the internal jugular vein (posterior) and carotid arteries (anteromedial). On the contrary, sympathetic chain schwannomas mildly splay the carotid bifurcation, but do not separate the great vessels. The splaying of the carotid bifurcation is seen in carotid body tumors called as the “lyre sign”.¹²

Excision of vagal schwannoma is challenging in sense that the tumor must be excised while sparing the cranial nerve. Unilateral vagal nerve transection leads to symptoms ranging from mild hoarseness, impaired cough mechanism to mild pharyngeal dysphagia with chronic aspiration. The vagal nerve fibers are intimately related to the tumor capsule requiring meticulous dissection to spare the nerve and thus increasing its chances of survival. Despite the care taken there is every chance of injury to the nerve unless nerve sparing techniques are employed. Intracapsular dissection which employs subcapsular plane of dissection of the tumor was introduced in 1920, by Cushing et al, for dissection of vestibular schwannomas.¹³ Valentino et al employed intracapsular enucleation preserved vagal function in more than 30% cases.¹⁴ Torossian et al reviewed postoperative neurological outcomes in 15 head and neck schwannomas excised by intra capsular enucleation and showed 86% successful preservation of neurological function by the technique.¹⁵ However, a comparison study by Giulio Illuminati between en block excision and intracapsular excision favours enblock with a recurrence of 33% with intracapsular excision.¹⁶

CONCLUSION

Further studies involving larger sample sizes and long term follow ups have to be undertaken to establish the success and drawback of this technique, but until then, Intracapsular technique should be resorted to in managing head and schwannomas.

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