A Case of Vagal Nerve Schwannoma: A Rare Case Report

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Abstract:

Schwannomas are rare, benign tumors that originate from Schwann cells, which are cells that provide support and insulation for nerve fibers. With the exception of the olfactory and the optic nerves, they may emanate from any of the body's peripheral, cranial, or autonomic nerves. This tumor often manifests as a solitary, slowly developing, asymptomatic neck lump with a slight probability of malignant transformation. The initial workup necessitates computed tomography with a contrast medium or magnetic resonance imaging. A precise pre-operative diagnosis may be challenging, as investigations such as Fine Needle Aspiration Cytology (FNAC) have a low specificity. Schwannomas arising from the carotid triangle and involving the Vagus nerve are very rare, but they can cause significant morbidity if left untreated. The treatment of choice is to surgically excise the tumor, with recurrences being rare. Here, we report a case of Schwannoma presented at our hospital which was treated by the surgical excision of the lesion.

Keywords: Schwannoma, Paraganglioma, Vagus nerve, Cervical Sympathetic Nerve, Extracranial

Introduction:

Cervical vagal schwannoma is a benign, slow-growing tumor that is frequently asymptomatic and has a very low lifetime risk of malignant transformation in the general population, although diagnosis remains challenging.[(1)] Verocay published the first description of schwannomas in 1908. The World Health Organisation (WHO) classification has since amended and added the terms schwannomas, solitary nerve sheath tumors, perineural fibroblast tumors, and neurilemmomas to describe this tumor.[(2,3)]

According to Calcaterra et al., the lateral side of the neck represents the area where solitary schwannomas most frequently occur, with the head and neck accounting for more than one-third of all cases. There may be antero-lateral displacement of the internal jugular vein and carotid artery. In the cases of extracranial schwannomas, the most frequent affected site is the parapharyngeal space. In this space, schwannomas can develop from either the autonomic nerves or the last four cranial nerves —the vagus being the most common site.[(4,5)] Vagal schwannomas account for about 3% of all intracranial schwannomas and are most commonly located in the jugular foramen. If the schwannomas originate from the vagus nerve, the patient may complain of hoarseness or occasionally cough when the mass in the neck is palpated. Additionally, Horner's syndrome, which is characterized by ptosis, mydriasis, pupillary constriction, or meiosis on the affected side, will be observed during the clinical examination of a patient with sympathetic nerve schwannoma.[(6)] Diagnosis is done on the basis of clinical examination and suspicion and a tissue biopsy is required to confirm the final diagnosis of schwannoma, however it is not typically recommended because it might result in complications such bleeding, hematomas, and hoarseness.[(1)] Here, we report a case of Vagal Schwannoma who was presented in our hospital which was treated with the surgical excision of the tumor.

Case Report:

A 38-year-old female patient presented with an asymptomatic swelling on the right upper neck since 2 years. Upon clinical examination, the right carotid triangle had a substantial 1.5 x 1 cm enlargement. It had no concomitant bruit and was movable, non-tender, and non-pulsatile. Upon oropharyngeal inspection, the right tonsil and posterior pillar were both clearly enlarged. Both vocal cords were movable during indirect laryngoscopy. The evaluation of the cranial nerves revealed no abnormalities.

A high resolution sonography was done that showed a large heterogeneous solid lesion measuring approximately 14 x 12 mm in the right side of neck. The lesion was located posterior to the right Internal Jugular vein and Carotid artery causing splaying and anterior displacement of the above mentioned vessels. Fine Needle Aspiration cytology showed mainly blood over which few fragments of spindle cells were evident. Computed Tomography (CT Scans) revealed a moderate sized soft tissue density in the right carotid space causing antero-medial displacement of carotid arteries and postero-lateral displacement and effacement of internal jugular vein. **(Fig 1)**

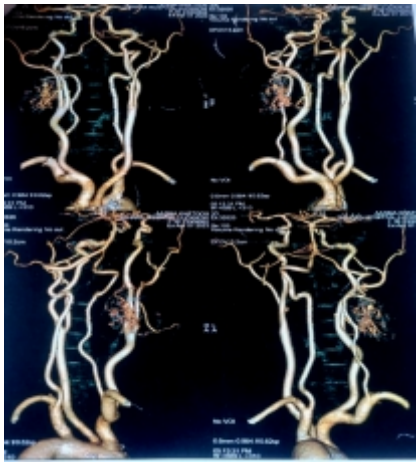

Fig 1: CT Angiogram showing a moderate sized soft tissue density in the right carotid space

Using a transcervical approach under general anesthesia (GA), a well-encapsulated tumour emerging from the vagus nerve was identified. **(Fig 2A, 2B)** The nerve was spared during the surgical excision of the tumour. **(Fig 3)** The post-operative period was uneventful, with no evidence of IX cranial nerve paresis. The Antoni A and B bodies, pallisading nucleus, and verruca bodies determined through histopathology were suggestive of schwanoma. No evidence of Horner's syndrome was observed post-operatively.

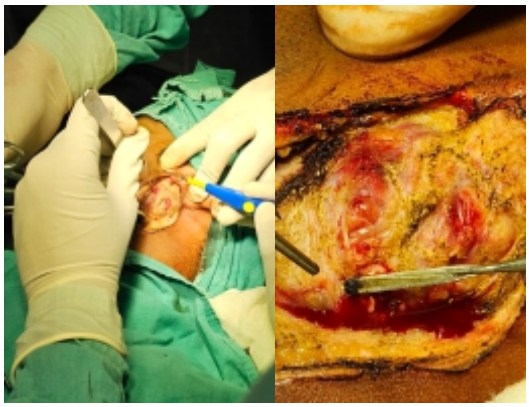

Fig 2A, 2B: Intraoperative photograph showing well encapsulated tumor arising from the vagus nerve.

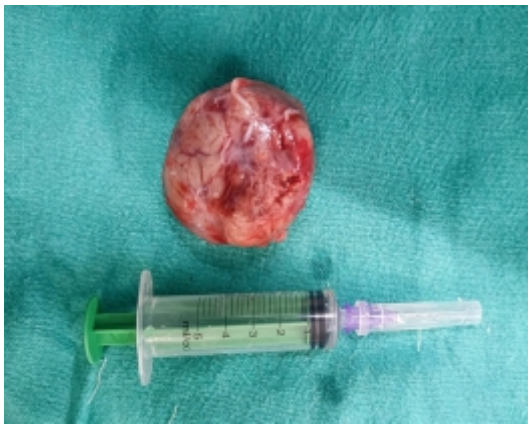

Fig 3: A pear-shaped 6 × 4 cm encapsulated tumor is excised

Discussion:

Paraganglioma is a benign tumour that occurs in the head and neck region, accounting for approximately 1% of all tumours. It usually manifests as a slow-growing tumour and rarely produces severe symptoms. Neurilemomas (also known as schwannomas or neuromas) account for half of these tumors. The schwannomas of vagus nerve are often benign tumours. The majority of schwannomas manifest as a slowly expanding, firm, painless mass in the lateral neck between the third and sixth decades of the patient's life. The frequency of the tumor in both the genders is 1:1.[7] The presenting symptoms might include hoarseness, pain, or cough. These swellings may move in transverse directions but not vertically. Pulsation is an unusual finding that, upon initial workup, suggests the possibility of a carotid body tumour. Angiography might be required in this case for the differential diagnosis.[2,8] Pulsation might be a reflection of the carotid artery system, however, it could also be a true pulsation emanating from the hypervascularity of the tumor (Schwannoma).

Schwannomas are often limited to the retrostyloid parapharyngeal region, although patients with schwannomas that extend into the posterior cerebral fossa through the jugular foramen have been documented.[9] Schwannomas are well encapsulated tumors with discernible cylindrical features (Antoni type A tissue), and are embedded in an ambiguous, loose stroma of fibres and cells (Antoni type B tissue). Necrosis, hemorrhage, and cystic degeneration are typical characteristics of this tumor.[1]

MRI or computed tomography with a contrast medium is essential for the initial workup. Margin lines are usually clearly delineated in schwannomas. On contrast-enhanced CT, they frequently exhibit higher attenuation than the adjacent muscle, although they can also be isodense or, less frequently, exhibit lower attenuation than the adjacent muscle. On MRI, the signal intensity of schwannoma appears to be high on T2-weighted images and intermediate on T1-weighted scans. On Ultrasonography, the tumor appears as a round or elliptical shaped in the cross section with well defined margins. Vagal nerve schwannoma can be observed on Computed Tomography (CT) as a well-covered, well-defined mass that is frequently more attenuated than muscle on contrast-enhanced images.[1,10] According to several authors, FNAC should be used routinely to determine the origin of all neck masses[11]. However, there is debate concerning the significance of utilising FNAC to diagnose Vagal Nerve Schwannoma prior to surgery.[12-14] The accuracy of the preoperative diagnosis obtained from FNAC relies on the specimen's quality and the cytopathologist's experience.[12] Additionally, FNAC can occasionally be hazardous, and needle or open biopsy is not recommended if

the cervical mass seems to be benign clinically and radiographically.[14]

As part of a neurovascular bundle, the cervical vagus nerve traverses via the carotid sheath between the internal carotid artery (ICA) or common carotid artery (CCA) and the internal jugular vein (IJV). The cervical sympathetic trunk passes longitudinally through the longus capitis and longus colli muscles, deep to the prevertebral fascia and posteromedial to the carotid arteries. On radiological imaging, replacement of vascular structures as a result of the mass effect of the schwannoma may provide insight into the origin of the tumor. A schwannoma that originates from the sympathetic chain or vagus nerve is suggestive of a tumor that is pressing the common carotid artery or internal carotid artery anteriorly. According to study done by Furukawa et al.[15], Schwannomas of the cervical sympathetic chain do not separate the common or internal carotid arteries from the jugular vein, whereas, vagal schwannoma separates the two structures. On enhanced T1-weighted MR images, both tumours exhibit strong enhancement, resulting in a distinctive "salt-and-pepper" appearance due to the frequent presence of flow voids within the tumor mass. The appearance of salt and pepper,however, is not a pathognomic of schwannomas.[1]

There is substantial debate over the disparity in recurrence of schwannomas between complete resection of the tumour including nerve fibres and intracapsular enucleation. Between these two procedures, Zbren et al discovered no significant difference in the prevalence of recurrence.[16] However, incomplete tumour removal has been shown to increase the likelihood of recurrence.

There are several surgical approaches for paraganglioma, and they primarily vary based on the location of tumor and whether adjacent structural involvement is evident. The transcervical skin incision for the neck paraganglioma is preferred with a suplatysma flap. Following careful dissection, this will sufficiently expose the tumour. Identification and protection of the vagal nerve, internal jugular vein, and carotid artery are of utmost importance. In this instance, a better surgical plane around the tumour was achieved by medially and laterally retracting the internal jugular vein and carotid artery, respectively.[17] Transtemporal, transoral, transmandibular, and combinations of these methods are a few alternative methods of accessing the tumour. For deep-seated paragangliomas, especially ones that are found in the parapharyngeal spaces, the transmandibular technique is suitable. In certain circumstances, a mandibulotomy must be done in order to locate, protect, and laterally mobilise the facial nerve.[17]

Pezzullo et al. reported a 12% prevalence of preoperative vocal cord paralysis, while hoarseness is usually always evident following surgery for schwannomas arising from the cervical vagus nerve.[18] Therefore, it is strongly recommended to evaluate the mobility of vocal cord postoperatively. Aggressive voice therapy for vocal cord compensation should be started as soon as possible following surgery to prevent postoperative vocal cord palsy.[5] Malignant transformation in nerve sheath tumours of the head and neck is extremely rare. Malignant transformation is exceptional in solitary schwannomas and evidence suggests that subtotal resection might provide adequate disease control of non-vestibular head and neck schwannomas, however regular follow-up is mandatory.[1]

Conclusion:

Vagal schwannoma of the cervical region is a significant clinical entity. Schwannomas of the carotid triangle and vagus nerve are rare but potentially debilitating tumors that can cause a variety of symptoms related to nerve compression. Diagnosis involves a combination of physical examination, imaging studies, and biopsy, and treatment typically involves surgical resection. While surgery can be effective in removing the tumor and preserving nerve function, it carries a risk of complications, and observation may be appropriate for small, asymptomatic tumors. Close monitoring is essential for all patients with schwannomas of the carotid triangle and vagus nerve to detect any changes in the tumor and to guide appropriate management.

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