CASE REPORT

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A Rare Case of Hypoglossal Nerve Schwannoma Arising at the Level of Carotid Bifurcation: A Case Report

ABSTRACT

Schwannoma's are solitary benign tumours, most commonly intracranial - arising from the vestibular nerve. Among extracranial schwannomas, only around 5% arise from the hypoglossal nerve with purely extracranial origin being least common. To the best of our knowledge, there have been only 3 reported cases in literature of extracranial hypoglossal schwannoma arising at the level of carotid bifurcation. We hereby present a case of a 51 year old female who presented with a symptomless swelling on the right side of her neck. Pre-operative diagnosis of extracranial primary schwannoma is difficult, and definitive diagnosis is by postoperative histopathologic examination. This tumor shows a low recurrence rate post complete surgical excision; which is the accepted modality of treatment at present.

Key words: Hypoglossal, Neck swelling, Schwannoma

INTRODUCTION

Schwannomas are benign slow growing tumors that originate from the myelin sheath of the nerves; predominantly arising from the vestibular nerve in 90% of the cases.[1] Most schwannomas are sporadic and few are associated with NF1, Schwannomatosis, Carney's complex, etc. Among the non-vestibular schwannomas, only about 5% arise from the hypoglossal nerve; tumor composed purely of motor nerves. Most hypoglossal schwannomas are intracranial in location, followed by intracranial lesion with a secondary extracranial extension – most frequently in the parapharyngeal space.[2] Primary extracranial hypoglossal schwannoma is very rare. Among them, to this date, there are only three reported cases of hypoglossal nerve schwannoma located at the carotid bifurcation.[3-5] These tumors are often diagnosed late due to non-specific clinical history and examination findings. They are asymptomatic for many years, and hence, histopathology is the gold standard of diagnosis.[6] We hereby present a case of a 52 year old female who came with a slow growing painless mass in the right side of the neck.

CASE REPORT

A 51-year-old female presented to the outpatient department (OPD) with complaints of a slow growing lump in the right lateral side of the neck for 10 years. The patient was apparently alright 10 years ago when she first noticed a lump in the right side of the neck, insidious in onset, initially the size of a pea, gradually progressive to its current size of an orange. It was not associated with pain, redness, tenderness, fever, cold, dyspnea, swallowing difficulties, paresthesias, or similar swellings anywhere else on the body. Patient did not have any comorbidities and did not give any history of past tuberculosis (TB) or TB contact. Her family history is unremarkable.

On examination, the patient was averagely built and well nourished. Patient had good oro- dental hygiene and

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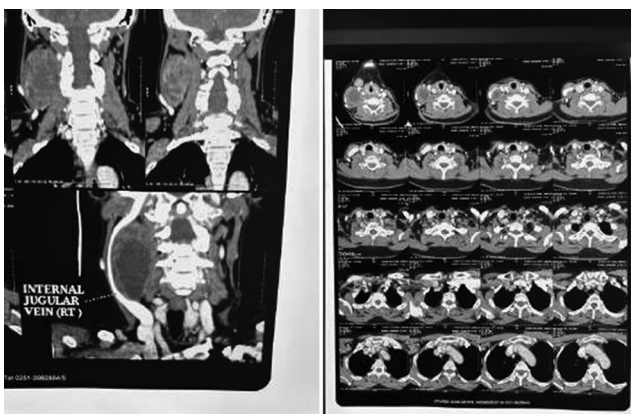
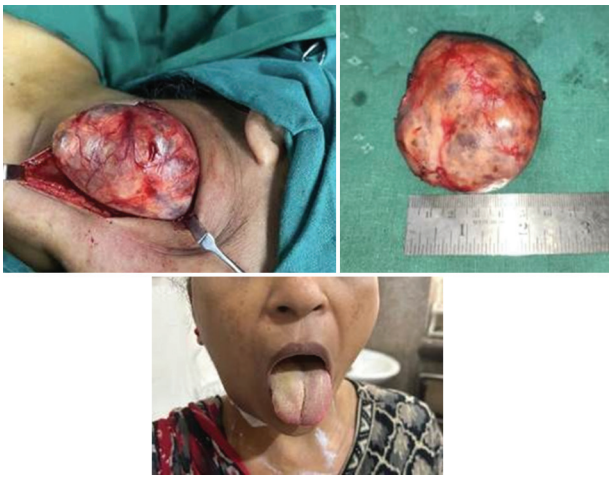
no lymphadenopathy noted on general examination. On inspection, a 7 × 10 × 5 cm smooth swelling noted in the right lateral side of the neck extending into the carotid and posterior triangles of the neck without any skin changes. On palpation, inspectory findings were confirmed. The swelling was tense, cystic, smooth surface, had no nodularities, no local rise of temperature, no tenderness, no skin changes, and not adherent to the overlying skin. It was mobile in mediolateral direction with restricted mobility in anteroposterior direction, no movement noted with deglutition and tongue protrusion. No pulsations felt and no bruit was heard on auscultation.

The patient presented 2 years ago with an outside ultrasound-guided fine-needle aspiration cytology (FNAC) report done in 2014 suggestive of a benign cystic lesion. Computed tomography (CT) head and neck was done suggestive of a 6.4 × 4.5 × 7.6 cm inhomogeneously enhancing solid cystic soft-tissue lesion extending into the upper and lower deep cervical region and posterior triangle region, laterally indented by the sternocleidomastoid muscle. The right internal jugular vein was compressed and displaced laterally; common carotid, internal, and external carotids were displaced anteromedially. Provisionally, differential diagnosis of simple cyst or carotid body paraganglioma was considered and the patient was advised excision of the lesion. The patient decided to not undergo surgery then and presented in OPD 2 years later with complaints as described above for surgery.

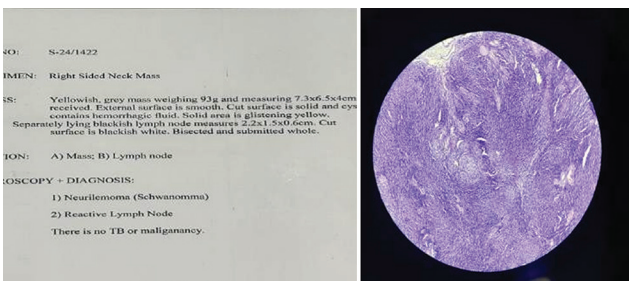


Intraoperatively, the solid thick walled lesion was seen to arise from the carotid sheath at the level of carotid artery bifurcation. It was separated from the hypoglossal nerve from its posteromedial aspect and removed *in toto*. A drain was placed and the wound closed in layers. Postoperatively, the patient showed paresis of the right hypoglossal nerve with deviation of the tongue to the right side. With no further postoperative complications, the patient was discharged with the drain 2 days after surgery.

Histopathological examination showed spindle shaped cells arranged in interlacing fascicles with hypercellular Antoni A areas and hypocellular Antoni B areas suggestive of neurilemmoma (schwannoma). On follow-up after a week,



the drain was removed and only mild hypoglossal nerve paresis noted. On follow-up after 4-week post-surgery, the hypoglossal neuropraxia resolved.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Right Sided Neck Mass

Yellowish, grey mass weighing 93g and measuring 7.3x6.5x4cm received. External surface is smooth. Cut surface is solid and cys contains hemorrhagic fluid. Solid area is glistening yellow. Separately lying blackish lymph node measures 2.2x1.5x0.6cm. Cut surface is blackish white. Bisected and submitted whole.

A) Mass; B) Lymph node

OSCOPY + DIAGNOSIS:

1) Neurilemoma (Schwanomma)

2) Reactive Lymph Node

There is no TB or maliganancy.

DISCUSSION

Schwannomas are very slow-growing tumors of the myelin-producing Schwann cells; they can occur along both sympathetic and somatic nerves in the body, with the exception of the olfactory and optic nerves, as these lack Schwann cells. Hypoglossal nerve schwannoma is very rare; as most cranial nerve schwannomas arise from sensory nerves. According to the Hoshi *et al*.,[7] the hypoglossal schwannomas recorded in the literature can be divided into three types: Type A, intracranial in 31.5%; Type B, dumbbell-shaped or extra- and intracranial in 50%; and Type C, extracranial in 18.5%. They are twice as common in females compared to males (M: F=1:2) and occur slightly more on the left side.

These tumors are benign, well encapsulated, and 52% cases present as asymptomatic palpable solitary masses.[8] Over long periods as they enlarge, the compressive symptoms could indicate towards the nerve of origin. A pre-operative diagnosis of schwannoma is difficult with varied differential diagnoses, including solitary thyroid nodule, enlarged lymph nodes, carotid body paragangliomas, thyroglossal cyst, brachial cyst, or tumor metastasis.

CT and magnetic resonance imaging are helpful in diagnosis; however, the role of FNAC is controversial – it

may be used to rule out other malignancies. Intraoperative attempt could be made to search for the originating nerve. However, post-operative histopathologic examination is still the gold standard for diagnosis.[9] The accepted treatment for these tumors is surgical resection with preservation of the neural pathway.[10] In case of extensive spread, subtotal resection may provide adequate disease control along with continued follow up; and if nerve of origin is sacrificed – immediate reconstruction and post-operative rehabilitation is advised.[11] Postoperatively, mild temporary neurological deficits are expected. After microsurgical resection, studies report 50–100% resection rate, no disability to new cranial nerve deficits in a third cases, and 8.3% tumor recurrence (1 of 12; adjunctive radiosurgery advised) to 100% tumor control.[12-14]

CONCLUSION

Hypoglossal schwannoma arising at the level of carotid bifurcation presenting as a lateral neck swelling is extremely rare with only three other cases reported till date; however should be considered as a differential. Definitive diagnosis is made by postoperative histopathology. It is recommended to excise the entire tumor and preserve the underlying nerve to prevent any neurological defects.

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