CASE REPORT

Fron Neck to Skull Base—A Rare Case of Cervical Sympathetic Chain Causing Diagnostic Dilemma—Case Report

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ABSTRACT

Cervical sympathetic chain schwanomma is a rare tumor of benign nature commonly presenting as an asymptomatic neck mass. It shows rare malignant transformation potential and is usually slow growing. Pre-operative imaging studies are sometimes useful to delineate the origin of tumors but not always accurate but can help in surgical planning by analyzing extent of lesion and the encasement of major neurovascular structures in the neck. The condition is usually treated by surgical excision, and the most common complication following excision is Horner's syndrome. The case report is of a young lady presented with an asymptomatic left neck mass. Multiple imaging studies showed various differentials, but surgical excision followed by histopathological evaluation concluded it to be a cervical sympathetic chain schwanomma. The workup and management of the case are discussed

Keywords: Schwanomma, Cervical area, sympathetic chain

INTRODUCTION

Schwanomma are benign neoplasms originating from Schwann cell which is a nerve sheath tumor.[1,2] They are rare and typically are solitary, well encapsulated, running along the course of the involved nerve and arise in any nerve cell ensheathed with Schwann cell. In the neck, commonly arising from the vagus nerve with other possibilities remaining lower cranial nerves, autonomic nerves or the cervical or brachial plexus.[2] Cervical sympathetic chain schwannomas are quite uncommon and mostly are asymptomatic but rarely presenting with Horner's syndrome. Pre-operative imaging is beneficial with CT or MRI but often fails to identify the exact pathology or nerve of origin because of close origin of nerves and shared imaging characteristics between other differentials. But radiological imaging is useful for surgical planning to tackle the anticipated intimate relation of tumor with the surrounding neuro-vascular structures, to understand its vascularity and blood supply as well as its extent into skull base or intracranial regions. Here, we describe a patient with cervical sympathetic chain schwanomma and shed some light into how the patient was worked up and the steps in surgical planning, intraoperative precautions as well as post-operative complications and their management.

CASE REPORT

The patient was a 25-year female who had presented to the OPD with a left side neck swelling in the region below angle of mandible (Fig 1), of approximate size 4 × 4 cm, from last 4 months which was insidious in onset, gradually progressive in size and not associated with any pain/breathing or swallowing difficulties/voice change/shoulder or neck weakness/tongue movement abnormalities/loss of taste/nasal regurgitation/choking or coughing while drinking fluids/speech abnormalities/drooping of eyelids/decreased

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sweating on ipsilateral side of face/pain in the arm/tingling or numbness in neck/shoulder/arm or any weakness in the left limb./epistaxis/palpitations/tremors/anxiety. The patient had no similar swelling in the past or anywhere else in the body and had no known family history of such disease. She had no history of addiction and had no other comorbidities. It was mobile in all directions, non-tender, non-pulsatile, and was not associated with any audible bruit.

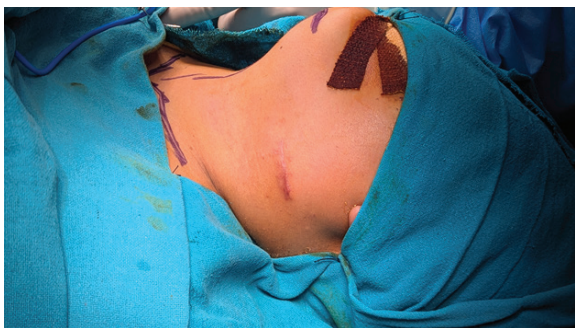

Fig. 1: Left sided neck swelling: scar over the swelling resulted from multiple prior FNACs which all were inconclusive

24-hour urinary metanephrine levels were found to be normal, however the contrast enhanced MRI showed a well-defined lobulated soft tissue lesion along the expected course of left vagus nerve, of size 4.2 × 4.26 × 3.6 cm. The lesion was described to be mildly hypointense on T1-weighted images and heterogeneously hyperintense on T2-weighted images, with a salt and pepper appearance, with differentials being Schwannoma and vagal paraganglioma. The lesion showed avid post-contrast enhancement, and it was situated lateral to left ICA and left IJV (Fig. 2). FNAC of the lesion showed spindle cells, suggesting more towards schwanomma, but the nerve of origin could not be concluded based on image findings alone.

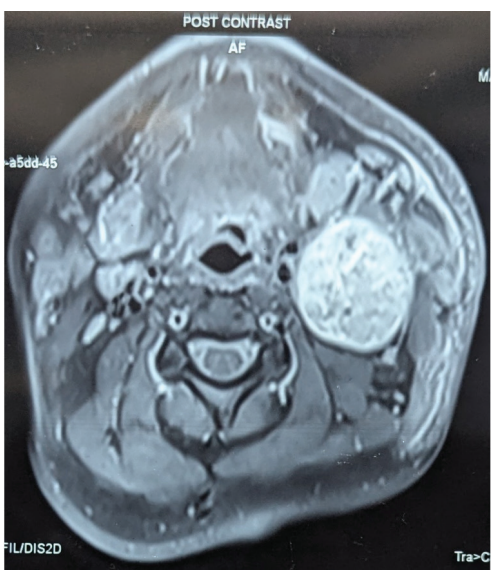

Fig. 2: CE MRI of the lesion showing avid heterogenous enhancement post-contrast with flow voids inside giving a "salt and pepper' appearance

The tumor was excised through an incision along the anterior border of sternocleidomastoid. After inducing the patient by general anesthesia, the patient was positioned in neck extension. Placing the incision and raising the subplatysmal flaps followed, and meticulous dissection was done for placing vessel loops around left IJV and left common carotid artery for vascular control. The tumor mass was delineated from the surrounding soft tissue attachments, and utmost care was taken around the major vessels as well as vagus nerve. Tumor was globose, firm in consistency and was finally found to be arising from the cervical sympathetic chain and was free from other lower cranial nerves as well as the cervical plexus. (Fig. 3, 4) The lesion was extending to the skull base and was completely removed with no residual disease (Fig. 5, 6)

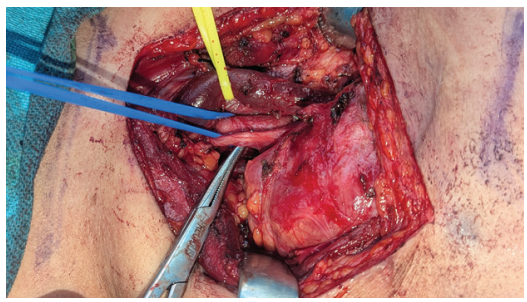

Fig. 3: Neck tumor found arising from sympathetic chain. Tumor was found free from the left vagus nerve (arterial clamp tip point). Vascular control over left ICA (blue) and left IJV (yellow) also shown

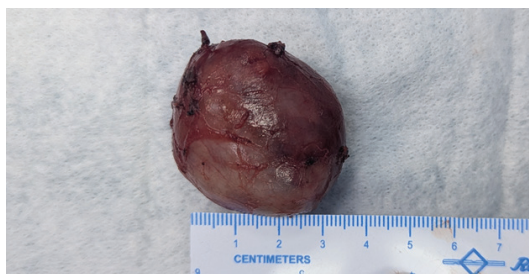

Fig. 4: Excised tumor

Post-operatively patient developed left Horner syndrome and managed conservatively. (Fig. 7) Patient developed no other complications and was discharged with stable parameters. Post-operative voice as well as shoulder and neck muscle function remained normal. The final histopathology report revealed diagnosis as ancient schwanomma of the cervical sympathetic chain.

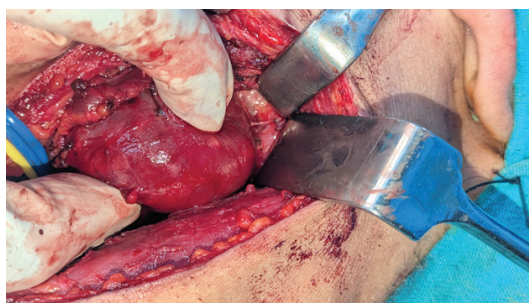

Fig. 5: Tumor origin tracing to skull base foramen

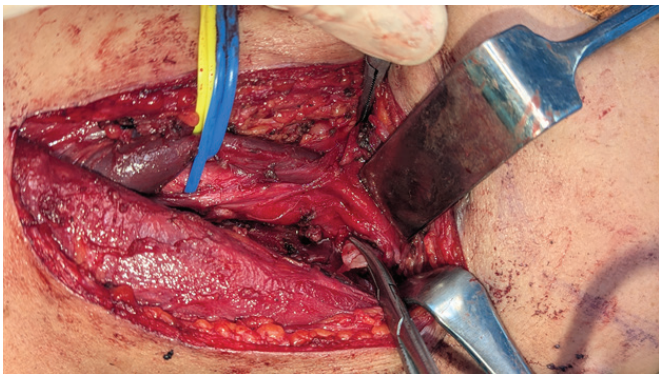

Fig. 6: The remaining stalk was removed after fine dissection

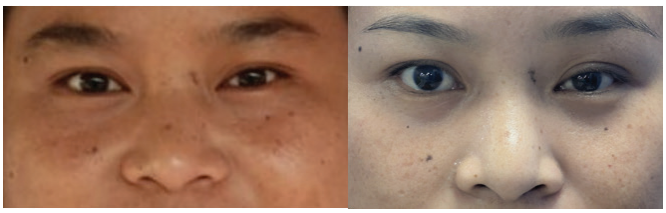

Fig. 7: Pre-operative and post- operative pictures showing post-operative left Horner's syndome

DISCUSSION

According to literature, cervical sympathetic chain schwannoma is an uncommon benign tumor among the other schwanommas or neurilemmommas, reported more common from Asia and presented as an asymptomatic solitary neck mass in patients between 20 and 50 years of age.[1,2] They arise from schwann cells and grow slowly along the course of the nerve with rare malignant transformation.

On microscopy, Antoni A areas comprising densely arranged cells with areas of pallisading nuclei in rows and hypocellular Antoni B regions are found Fig. 8 3.

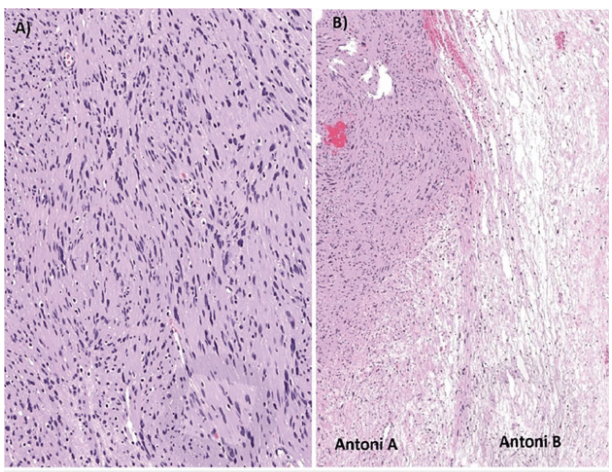

Fig. 8: Antoni A and Antoni B areas in histopathology

Pre-operative Horner's syndrome is rare.[4] Imaging prior to surgical removal usually include contrast-enhancing computed tomography (CT) or magnetic resonance imaging (MRI) and ultrasound (US) and occasionally angiography when having confusion with vascular paragangliomas which also arise from the same location. In cervical sympathetic chain schwanommas no separation is observed between the internal jugular vein and the common carotid artery.[5]

Presurgical diagnosis confirmation showed low values (11 percent) and definitive diagnosis was achieved on post-operative histopathology.[1] The tumor was excised in most cases via an external approach by an extra-capsular dissection technique and post-operatively resulted in Horner's syndrome in majority and first bite syndrome in few. Horners syndrome although didn't leave the patient in distress and was correctable by advancement of the levator aponeurosis, or resection of the conjunctiva and Müller's muscle.[6,7]

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