A Huge Carotid Space Schwannoma Arising from The Cervical Sympathetic Chain - A Case Report

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Abstract

The Rationale: Although 25%–45% of schwannomas are originating from the neck, carotid space schwannomas are extremely rare. **Patient Concerns:** We report a rare case of huge-sized schwannoma in a 20-year-old student who presented with a symptomless large carotid space mass. **Diagnosis:** Cervical magnetic resonance imaging (MRI) with contrast revealed a huge, well-defined mass measuring 13.7 cm × 6.4 cm × 4.1 cm. Cervical MRI along with brain MRI were consistent with neurofibromatosis type II. **Treatment and Outcomes:** Preoperative tracheostomy and wide local excision of schwannoma via a transcervical approach were performed with nerve preservation. **Takeaway Lessons:** The scarceness of cervical sympathetic chain schwannoma made this case of our patient very interesting to report. Moreover, our patient's huge tumour size is extremely rare, and we could not find any similar cases in the literature.

Keywords: Carotid space, schwannoma, sympathetic chain

Introduction

Cervical region schwannomas contribute 25%–45% of the cases, most commonly from the vagus nerve. On the other hand, schwannomas originating from the cervical sympathetic chain are extremely rare.[1]

Although clinical presentation is largely dependent on the nerve of origin, vagal schwannomas can be confused with paragangliomas, metastatic lymph nodes, and schwannomas of the cervical sympathetic chain.[2] Preoperative diagnosis of cervical schwannomas is a challenge; however, computed tomography (CT) and magnetic resonance imaging (MRI) have solved this problem regarding the nerve of origin, detailed location, as well as surgical planning.[3]

We report a rare case of huge carotid space schwannoma arising from the cervical sympathetic chain in a 20-year-old patient which was surgically removed through a transcervical approach. This case has been reported with respect to the SCARE criteria.[4]

Case Report

Patient concerns

A 20-year-old male student presented to our surgery at Suez Canal University Hospital, referred by his neurosurgeon, with a 7-year history of slowly enlarging lump of the left side of his neck. This swelling was associated with multiple swellings of the scalp, trunk, and left forearm. The patient had a recent history of dysphagia and obstructive sleep apnea along with hoarseness of his voice. However, he had no hearing or neurological problems. There was no significant past or family history.

Diagnosis

On physical examination, the patient looked comfortable at rest with no speech abnormality. There were no obvious abnormal facial appearance and no evidence of any kind of dyskinesia. The patient had multiple swellings of his scalp, trunk, and left arm [Figure 1].

Neck examination revealed a solitary symmetrical oval swelling of the left carotid triangle measuring approximately

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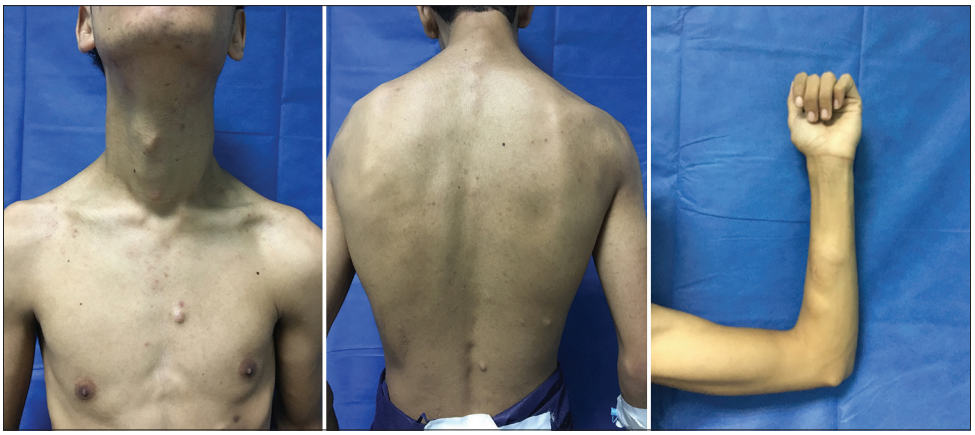

Figure 1: Multiple swellings of the anterior/posterior trunk and the left forearm of the patient

15 cm × 8 cm with normal skin overlying and no visible scars [Figure 2]. The swelling was nontender, firm, well defined, with limited cephalocaudal mobility. There was no pulsation, no fluctuation, and negative transillumination. Trachea was shifted to the right side. No cervical lymphadenopathy was detected, and no abnormal sounds were heard on auscultation.

Cranial nerve examination showed no abnormalities other than tongue deviation to the left side, which is consistent with left hypoglossal nerve involvement [Figure 3]. Indirect laryngoscopy failed to reveal the mobility of both vocal cords, due to the mass effect of the swelling obscuring the view.

Cervical MRI with contrast [Figure 4] highlighted a large prevertebral retroesophageal mass lesion measuring 13.7 cm × 6.4 cm × 4.1 cm in maximum craniocaudal, transverse, and anteroposterior dimensions, respectively, and displaying isointense signal on T1 and bright signal on T2 WIs with heterogeneous contrast uptake. It was seen indenting the esophagus and the posterior airway column. In addition, MRI of the brain [Figure 5] showed multiple meningiomas with bilateral acoustic schwannomas; findings were consistent with neurofibromatosis type II.

Treatment

The clinical findings and imaging observations were discussed at a multidisciplinary team meeting in the same week. The plan was to perform a preoperative tracheostomy and wide local excision of the schwannoma via a transcervical approach. Under general anaesthesia, supine position, and 30° head up, a subplatysmal flap was raised. Marginal mandibular, hypoglossal, and lingual nerves were identified and preserved. An ovoid well-circumscribed mass, with smooth yellow/tan outer surface showing prominent vascular arborization, was identified medial to the carotid sheath [Figure 6]. The mass was seen compressing the left hypoglossal nerve. Successful nerve preserving surgical excision was performed.

Outcomes

The specimen was sent for histopathological examination. Microscopic pictures [Figure 7] revealed a biphasic tumour formed of compact hypercellular (Antoni A) areas, with nuclear palisading around the fibrillary process (Verocay bodies), and myxoid hypocellular (Antoni B) areas. These findings confirmed the diagnosis of schwannoma. No immunohistochemical stains were used.

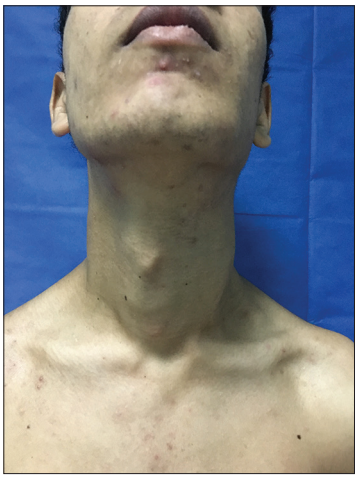

Figure 2: Left-sided huge swelling of the neck (note the tracheal shift to the right side due to mass effect)

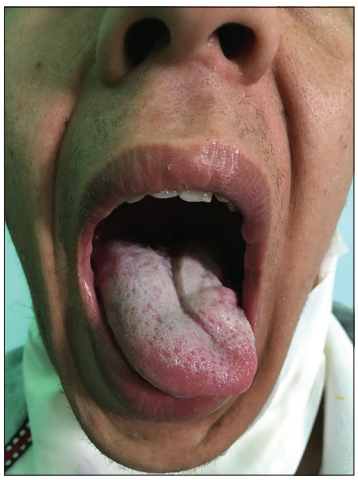

Figure 3: Tongue deviation to the left (affected) side

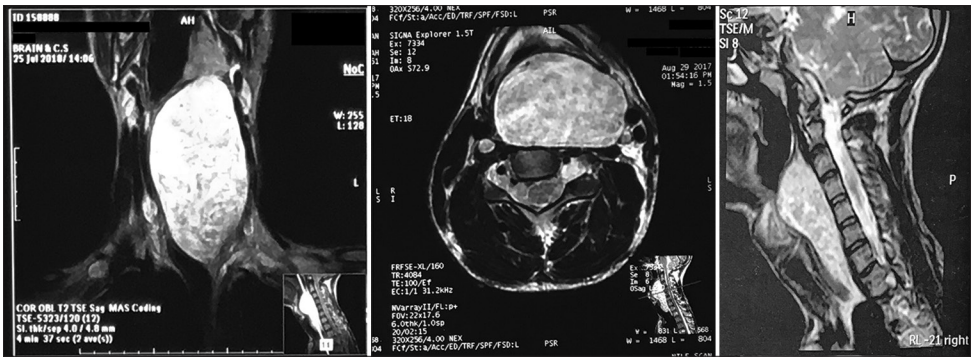

Figure 4: Coronal, axial, and sagittal magnetic resonance imaging demonstrating a large homogenous encapsulated mass in the left carotid space

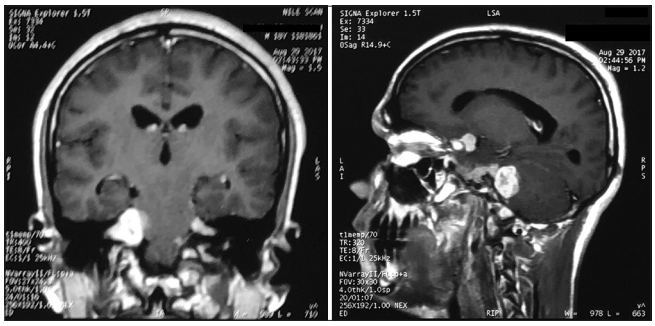

Figure 5: Coronal and sagittal magnetic resonance imaging of the brain showing multiple meningiomas and bilateral acoustic schwannomas

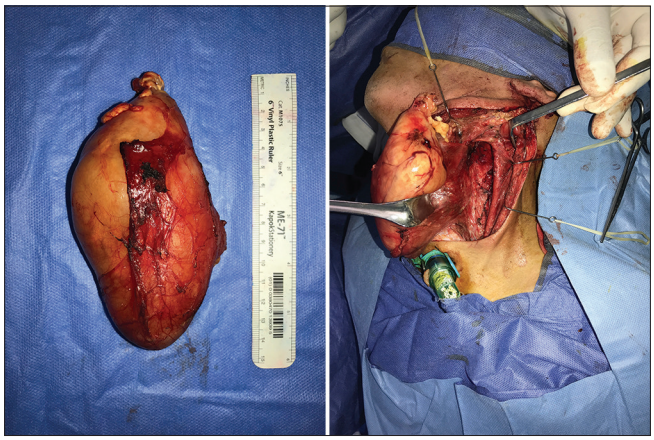

Figure 6: Intraoperative pictures of the mass and the excised patient's specimen

Follow-up

On subsequent follow-up appointments, the patient was recovering very well, with no evidence of neurological affection including Horner's syndrome. The tracheostomy tube was removed 1 week later after assuring the patency of the patient's airway. The patient has been regularly reviewed for 1 year with no signs of recurrence.

Discussion

In our case report, a huge mass measuring 137 mm × 64 mm × 41 mm in maximum craniocaudal, transverse, and anteroposterior dimensions, respectively, was highlighted. This massive tumour size is extremely rare, and we could not find any similar case reports in the literature.

Schwannomas of the head and neck can originate from any cranial, peripheral, or autonomic nerve, with the exception of olfactory and optic nerves, as they are devoid of Schwann cell sheath.[5]

The presentation can vary and are mostly nonspecific. However, a single, slow-growing lump in the neck is the most common symptom.[6]

The carotid space is a cylindrical space extending from the skull base through to the aortic arch. Most of schwannomas in the carotid space arise from the vagus nerve and the cervical sympathetic chain and are extremely difficult to differentiate, especially when their symptoms are not specific.[7]

CT and MRI have, to a certain degree, facilitated the diagnosis. Preoperative imaging supports information on the size, extent, and location of the tumour within the surrounding anatomy, thus aiding surgical planning.[8] However, MRI is considered superior to CT because of higher soft-tissue delineation, particularly the fat signs, which determined the relationship between the tumour and the great vessels of the carotid space.[7]

Schwannomas arising from the vagus nerve, tend to separate the common carotid artery (CCA) anteriorly and internal jugular vein (IJV) posteriorly. On the other hand, In schwannomas originating from the sympathetic chain, the tumour displaces both the CCA and IJV anteriorly without separating them.[9]

The usefulness of fine-needle aspiration (FNA) is equivocal. Most authors do not recommend FNA for these masses.[8] Histologically, schwannomas typically show a biphasic histological pattern named Antoni A and Antoni B.[10]

Tumour enucleation is the surgical method of choice to preserve the function of the affected nerve; however, enucleation does not assure completely intact nerve function postoperatively. Nerve functionality can be preserved in most vagus schwannoma cases. On the other hand, preservation of nerve function in sympathetic nerve schwannoma remains a challenge and necessity for further investigation.[11] In our case, we successfully excised the mass without sacrificing the

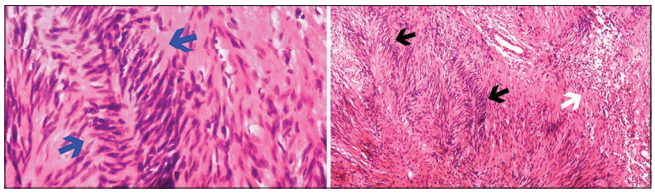

Figure 7: H and E-stained sections from the mass. Sections showing biphasic tumour formed of Antoni A areas (black arrows), with Verocay bodies (blue arrows), and Antoni B areas (white arrow)

nerve trunk. A common postoperative complication is Horner's syndrome. Our patient did not suffer from Horner's syndrome or dysphagia postoperatively.

Conclusion

Carotid space schwannomas are extremely rare and mostly present with a slow-growing, painless swelling in the neck. The rarity of this huge schwannoma makes our patient very interesting to report. Preoperative diagnosis can be challenging, and investigations such as CT and MRI can be very beneficial for surgical planning and the most important, identifying the nerve of origin. Nerve-preserving surgery is the treatment of choice and can be achieved through a number of different surgical approaches. Recurrence is unlikely as in this case.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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