

Case Report

Cervical root schwannoma: a case series

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ABSTRACT

Cervical root schwannoma is an infrequent benign peripheral nerve tumor though those arising from high cervical spinal root are common amongst the spinal schwannomas. This mass commonly presents as a slow growing, asymptomatic, solitary neck mass with rare potential of malignant degeneration. Displacement of Internal jugular vein and carotids anteriorly is frequently seen though abutment of esophagus is uncommon. Pre operative diagnosis can be established with imaging and Fine needle aspiration cytology (FNAC). Complete Surgical excision is the treatment of choice. Horner's syndrome is the most common post operative neurological manifestation. Here we have recorded clinical features intra-operative findings and surgical histopathology. We have described a case of 19 year old male with cellular schwannoma of cervical sympathetic chain with intraspinal extension and arising from C5-C8 level. Another case of cervical root schwannoma is described in a 5 year old girl arising at C4-C5 level. Complete surgical excision was done for both patients with no postoperative neurological affection. The clinico-pathological evaluation and management are described.

Keywords: Cervical root schwannoma, Surgical treatment

INTRODUCTION

Schwannomas, also known as neurilemmomas, are benign tumors of Schwann cells.

Schwann cells originate from neural crest cells and surround peripheral nerve tissues.¹ They are morphologically solitary, well encapsulated benign nerve tumors notably running in the line of the nerve or attached to peripheral, cranial, sympathetic nerves. Malignant transformation is rarely seen. Schwannomas usually arise from lower cranial nerves, more commonly from vagus nerve in the parapharyngeal space. Cervical sympathetic chain schwannomas are rare^{2,4,5} especially appear as slow growing solitary neck mass. Cervical spinal schwannoma is benign and outcomes after surgical resection are generally excellent.³ Horner's syndrome is rarely apparent on physical examination.⁶ Pre-operative diagnosis is made using contrast enhanced MRI, MR Angiography and FNAC. Pre and Post-operative

neurological assessment is essential. We describe a case of 19 year old male with cervical sympathetic chain schwannoma with intraspinal extension arising at the level of C5-C8. Another case of cervical root schwannoma is described in a 5 year old girl arising at C4-C5 level.

CASE REPORT

Case 1

A 19-year old young male presented with left sided gradually progressive, painless neck mass, which measured approximately 8X6X7 cm, located in posterior triangle extending from mandible superiorly to clavicle inferiorly along the anterior border of trapezius (Figure1). It was mobile, non-tender, non-pulsatile with cystic consistency and no associated bruit. The mass was not associated with dysphagia, dyspnea, dysphonia, aspiration, visual complaints or any abnormality of arm

and hand movements. On further enquiry, he complained of occasional numbness over left forearm and arm. He denied any addictions. Diagnostic MRI revealed a 5.5x4.4x8.5 cm well defined, heterogeneously enhancing, dumbbell shaped soft tissue mass in the left paravertebral region with widening of the neural foramina of C5-C8 nerves and intra-spinal extra-dural extension at C6-C7 levels and intra- dural and extra-medullary extension causing indentation of cord at C6 level. (Figure 2 a & b) The left vertebral artery is encased and compressed by the lesion with further anterior extension to abut the esophagus and displacing carotid space causing compression of internal jugular vein and common carotid artery. The lesion extends inferiorly into the thoracic inlet. MR angiography was normal. The other differentials are vagal schwannoma, metastatic or reactive node, paraganglioma. Fine needle aspiration revealed tumor cells arranged in interlacing fascicles with hypocellular areas. Verocay bodies were identified.

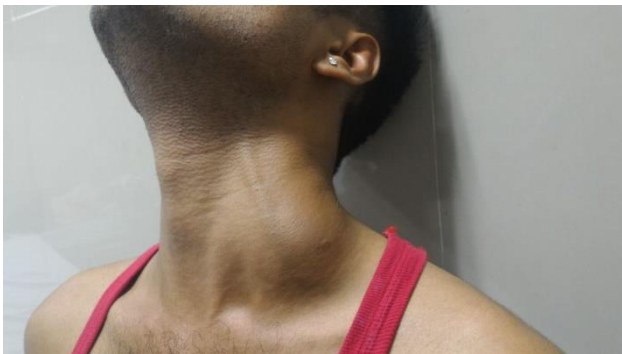


Figure 1: Neck swelling in the left posterior triangle.

The mass was excised through trans-cervical approach. The mass was seen to be arising from the cervical body and dissected carefully preserving the nerve roots. Tumor was dissected from hypoglossal, lingual, spinal accessory nerve, common carotid artery and internal jugular vein preserving them. Post operatively patient persisted with occasional numbness of forearm and had normal arm movements and normal ocular examination (Figure 3). Post-operative course was normal and patient was discharged on 7th postoperative day.

Histopatholgy

Histopathological examination of the resected specimen revealed a partly encapsulated tumor composed of spindle cells arranged in sheets, intersecting fascicles and focally palisading pattern. Majority of tumor was hypocellular with very few hypocellular areas focally. Tumor cells had oblong to spindle nuclei, fine nuclear chromatin, mild nuclear pleomorphism and few prominent nucleoli. Mitotic count was 1-2/hpf. The findings were suggestive of cellular schwannoma.

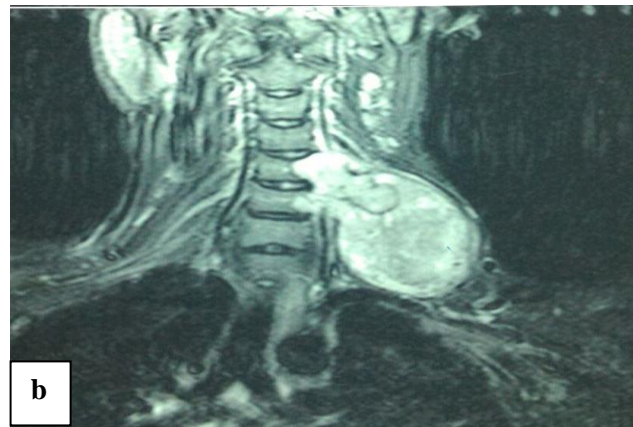
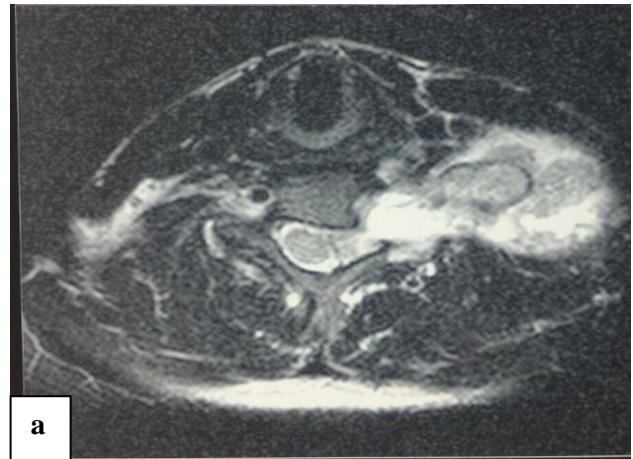


Figure 2 (a and b): MRI showing the dumbbell shaped soft tissue mass in the left paravertebral region with widening of the neural foramina and intra-spinal extra-dural extension at C6-C7 levels and intra-dural and extra-medullary extension causing indentation of cord at C6 level.



Figure 3: Patient at 2 weeks postoperatively.

Case 2

A 5 year old girl presented with complaints of a painless, gradually enlarging anterior neck swelling since 1.5 years. Physical examination revealed a 4X3 cm mobile, non-tender, firm swelling extending from anterior border of right sternocleidomastoid to 2 cm from midline. Superiorly, it extended upto 2 cm from the hyoid and inferiorly upto 1 cm from suprasternal notch (Figure 4). MRI revealed a 5X2.2X2.1 cm T1 isointense T2 iso to hyperintense mass anteromedial to right carotid sheath and posterolateral to right lobe of thyroid. Superiorly, the mass extends to inferior margin of submandibular gland and inferiorly to just above the junction of subclavian and internal jugular vein. It abuts and displaces the carotid sheath posterolaterally and the right thyroid lobe anteromedially. Posteromedially, it abuts the spinal column at C4-C5 level. It shows intense heterogeneous post-contrast enhancement with few tiny flow voids within (Figure 5 a, b). The patient was taken up for transcervical excision of tumor under general anesthesia. The mass was dissected free from strap muscles, right thyroid lobe, sternocleidomastoid and carotid sheath. It seemed to arise from one of the cervical roots. Post-operatively, the patient did not develop any motor weakness or sensory abnormality/ Histopathological examination of the resected specimen was suggestive of schwannoma.



Figure 4: Neck swelling in the right anterior triangle.

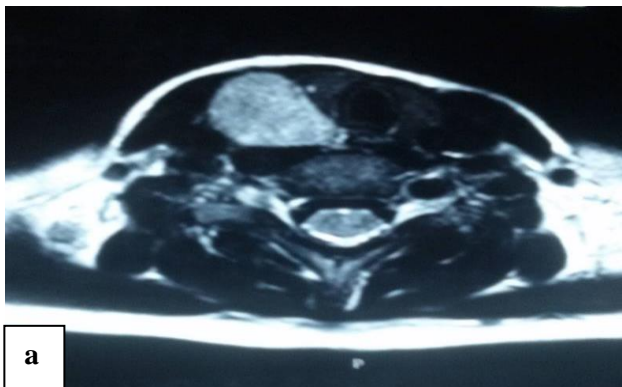


Figure 5: a and b: MRI showing T2 iso to hyperintense mass displacing the carotid sheath posterolaterally and the right thyroid lobe anteromedially. Posteromedially, it abuts the spinal column at C4-C5 level.

DISCUSSION

Cervical spinal schwannoma is benign and outcomes after surgical resection are generally excellent. The surgical dilemma sometimes arises whether to perform total tumor removal, which carries the risk of sacrificing the nerve root, or subtotal removal, where the risk of recurrence is high.³ A review of literature revealed fewer than 45 cases of schwannoma^{4,7,8} of sympathetic chain in patients aged between 20 to 50 years. Spinal schwannomas account for around 25% of primary spinal cord tumors with no sex prevalence. In the literature, 70 to 80%^{9,10} of spinal schwannomas are reported to be located intradurally and those presenting as dumbbell with both intra and extra-dural extension account for another 15%. Intra medullary schwannomas are extremely rare.¹¹

Pre-operative diagnosis is usually established with CT, MRI, MR Angiography. Our patient was examined by FNAC, MRI and Angiography. In order to distinguish between sympathetic ganglioma, vagal schwannoma, paraganglioma and metastatic or reactive lymphadenopathy imaging with CT or MR with contrast is always done.

Schwannoma on plain CT examination, is generally hypo-dense compared to the muscle, with contrast, this lesion shows mild enhancement. MRI reveals low signal intensity on T1 and high signal intensity on T2-weighted images.

Paraganglioma, on the other hand, is classically iso-dense on plain CT when compared to muscle, with more reliable homogeneous enhancement post-contrast.¹² The post-gadolinium MRI sequences of paraganglioma, show contrast enhancement in a characteristic “salt-and-pepper” pattern.^{13,14} Usually, the origin of the paraganglioma is more cranial in the supero-medio-latero-cervical neck region with respect to the schwannoma. The mass, in our adult patient, was located in the paravertebral region from mandible to clavicle in

close proximity to the cervical vertebrae. The diagnostic challenge was to differentiate between vagal schwannoma and schwannoma of the cervical sympathetic chain. Usually the vagal schwannoma grows between common carotids and internal jugular displacing the vessels in contrast the carotid space in maintained in cervical sympathetic chain glioma.¹⁴

In a study done by Yamane K et al,³ it was observed that all permanent motor deficits were the result of resecting functioning relevant nerve roots that is C5-C8. The rate of permanent sensory deficit was 67% after C5-C8 nerve root resection. Dumbbell tumours were associated with the need for total or ventral nerve root transection as well as with a high incidence of tumour recurrence. The incidence of permanent neurological deficit was significantly higher in patients undergoing C5-8 nerve root resection.

Therapeutic modality of choice is Trans-cervical approach as it gives access to major structures in the neck and minimizes neuro-vascular complications. In the adult patient, we were able to dissect the tumour from vertebral body preserving the spinal roots around the foramina causing no neuro-vascular damage intra-operatively, however outcome of surgery correlates to preoperative neurological condition of the patient.¹⁶

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