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Case Report

Facial pain: a rare presenting symptom of sympathetic chain schwannoma

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ABSTRACT

Schwannoma is a nerve sheath tumour that usually presents as an asymptomatic lump. We report a case where facial pain was the only symptom of the tumour. Surgical excision was done. Intraoperative findings and Horner syndrome confirmed the diagnosis of cervical sympathetic chain schwannoma. Histopathology was also consistent with the diagnosis. The patient did well postoperatively.

Keywords: Cervical schwannoma, Facial neuralgia, Sympathetic chain schwannoma

INTRODUCTION

Schwannomas are benign, slow-growing tumours of neural origin. They are also known as neurinomas and neurilemmomas. They may arise from cranial nerves including glossopharyngeal, accessory, vagus, hypoglossal nerve or the cervical sympathetic chain in parapharyngeal space. The vagal schwannomas are the commonest parapharyngeal schwannomas. Cervical sympathetic chain schwannomas are rare. Parapharyngeal schwannomas, can remain asymptomatic for months when they are small. The presenting complaints are an asymptomatic lump, dysphagia, hoarseness, dysarthria, Horner syndrome and rarely neuralgic pain. Radiological evaluation is mandatory for detailed evaluation and surgical planning of these tumours. The diagnosis is confirmed with histopathology results of tumour specimen as preoperative fine needle aspiration cytology is usually inconclusive. Treatment is surgical excision via external approach. The long term sequala in terms of recurrence or malignant transformation is rare. The prognosis after surgical excision is excellent.[1,2,5]

We report an unusual case of cervical sympathetic chain schwannoma in a 47-year-old female, presenting as chronic facial neuralgia and discuss its management.

CASE REPORT

A 47-year-old female presented to outpatient clinic with right-sided facial pain for two years. The aching pain was spontaneous in onset, gradually progressive, continuous, and partly relieved with analgesia. There were no other symptoms. Past medical history was not significant. Physical examination of the head and neck did not reveal any abnormality. The examination of larynx was unremarkable. The neurology team had ruled out other aetiologies for hemifacial pain. She was taking gabapentin, amitriptyline, and tramadol for one year but without complete relief.

MRI of head and neck with contrast revealed a well-defined lobulated soft tissue lesion of size 5.9×3.6×4.5 cm in the right paravertebral region at C7-D2 level appearing hypointense on T1W and heterogeneously hyperintense on T2W/STIR images. The lesion showed homogenous enhancement on the post-contrast study.

Ultrasound-guided fine needle aspiration cytology was done but was inconclusive and did not contribute to the diagnosis.

Surgical excision was planned by the transcervical approach. Surgical exploration revealed a firm mass that was displacing the carotid anteromedially and internal jugular vein and vagus anterolaterally going up to the vertebral column posteriorly. The lower end of the tumour was going into the thoracic inlet which was pulled into the neck by insinuation and dissection in the sub-adventitial plane. The tumour was not found adherent to any of the surrounding structures and was removed in toto via extracapsular dissection. The patient developed Horner syndrome in the immediate post-operative period. Histopathology showed encapsulated lesion composed of Antoni A and Antoni B cellular areas consistent with Schwannoma. In a follow up of 1 year, the hemifacial pain was relieved, and symptoms of Horner syndrome also subsided.

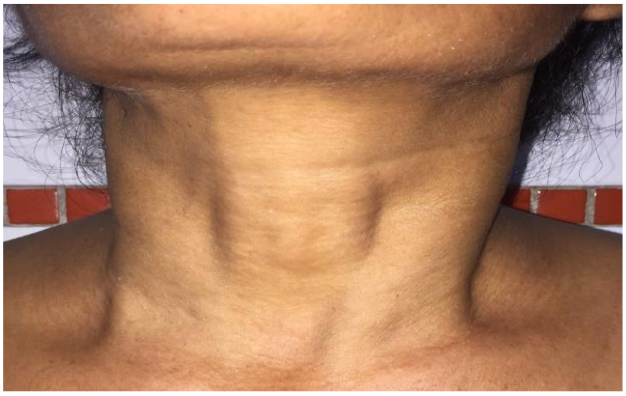

Figure 1: Neck examination-no visible or palpable swelling in the neck.

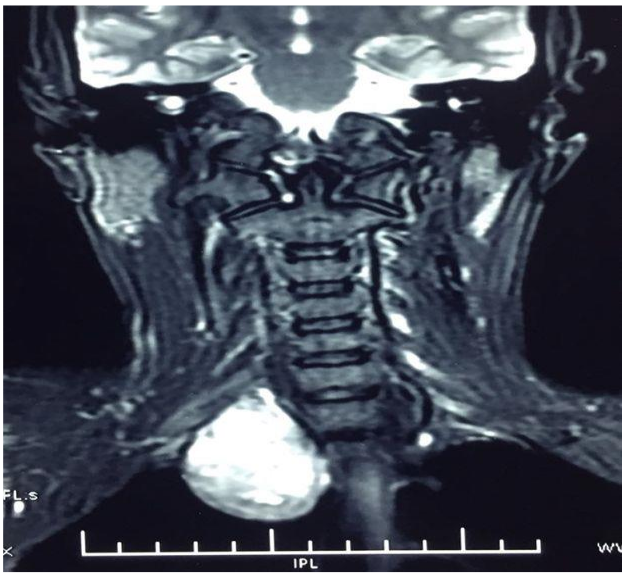

Figure 2: MRI T2W image: hyperintense soft tissue lesion seen at the level C7-D2 in the right paravertebral region.

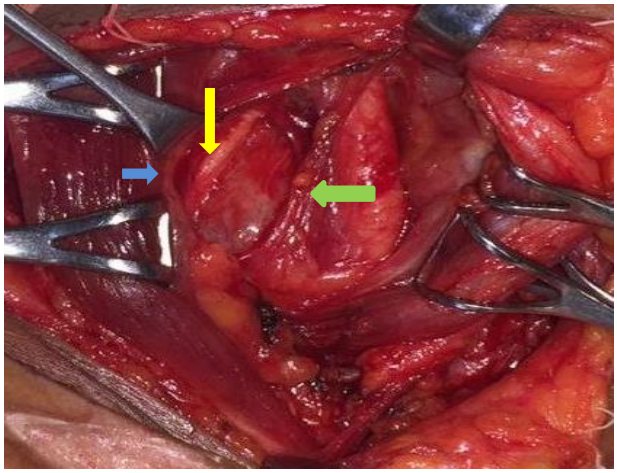

Figure 3: Surgical exploration-the tumour (yellow arrow) is located medial to vagus (blue arrow) and lateral to carotid (green arrow).

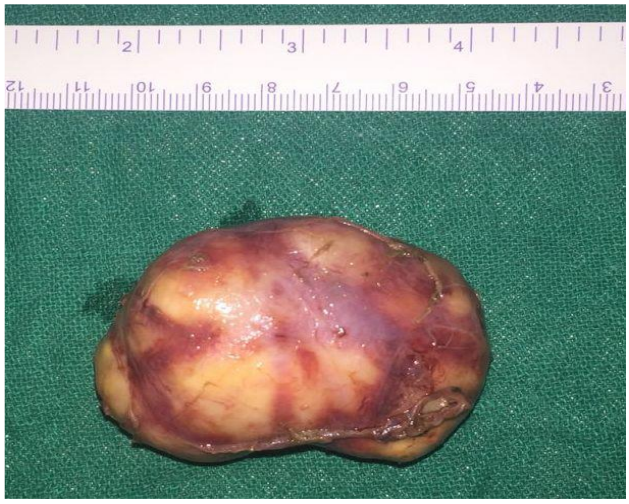

Figure 4: Gross specimen: ovoid mass of size-6×3.5×4.5 cm.

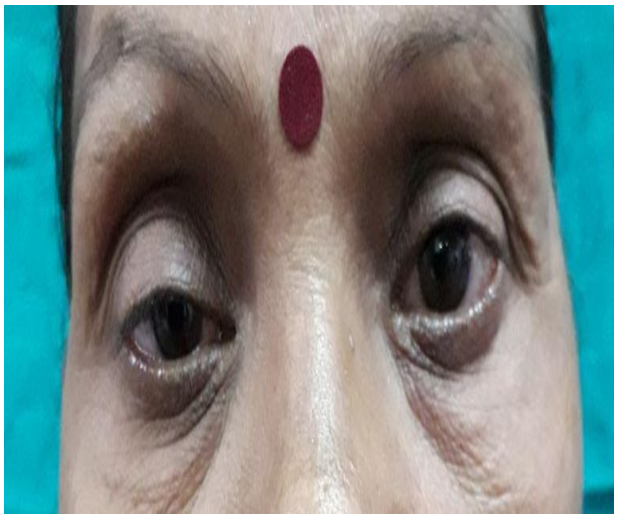

Figure 5: Horner syndrome: post-operative day 1-miosis and ptosis on right side.

DISCUSSION

Schwannomas are benign, slow-growing tumours originating from Schwann cells of the nerve sheath. Schwannomas can arise anywhere in the body. Approximately 25-45% of them are found in the head and neck region. In parapharyngeal space they are the second most common benign tumours after parotid tumours. The site of origin could be lower cranial nerves, most commonly vagus nerve, or sympathetic chain. Cervical sympathetic chain schwannomas are rare and affect middle-aged individuals with an equal predilection for both sexes.[3] In our case, the tumour was found to be located at the thoracic inlet which is an unusual location.

They remain asymptomatic for a long time or can present as a lump in the neck. The other associated symptoms are dysphagia, voice change, very rarely Horner syndrome.[3] Neuropathic pain is a rarely reported symptom.[6] In this case, the only presenting complaint was right-sided hemifacial pain which was corresponding to the trigeminal dermatome. This shows that along with sensory nerves, autonomic system also determines the neuralgic pain. The resolution of pain in the postoperative period also confirmed that it was sympathetic neuralgia.

The provisional diagnosis is made with imaging modalities. CT without contrast shows hypodense lesion and with contrast show rim enhancement at the periphery. MRI reveal low signal intensity on T1-weighted images and high signal intensity on T2-weighted images.[4] The MRI findings in our case were suggestive of the diagnosis of schwannoma.

Fine needle aspiration cytology is usually inconclusive for preoperative diagnosis. Histopathology of tumour specimen is confirmatory for the diagnosis which shows well defined encapsulated lesion and Antoni A and Antoni B areas on microscopy.

Surgical excision is the treatment of choice via external transcervical approach. Extracapsular excision is recommended while preserving surrounding neurovascular bundle.[5,6]

We managed to excise the tumour completely by the transcervical incision. Horner syndrome (miosis, ptosis, anhidrosis and enophthalmos) is a common complication following surgery.[5,6] Our patient had miosis and ptosis in the immediate postoperative period that resolved in one year follow up.

Neuralgic pain is a challenging entity to manage. In this case report, we have discussed how a sympathetic chain schwannoma can present with atypical symptom of facial neuralgia. We have highlighted the fact that the this was an unusual case and correlated the clinical, radiological, histopathology findings with the surgical outcome.

CONCLUSION

Cervical sympathetic chain schwannoma is a rare entity. It can present as lump in neck, dysphagia, hoarseness, Horner's syndrome and rarely with neuropathic pain. We have discussed a case with unique presentation and location with diagnostic and management challenges. The sympathetic facial neuralgia is a rare symptomatology of this tumour and should be kept in mind while making a differential diagnosis. The radiological investigation is a must for diagnosis. The treatment is via surgical excision. The prognosis is excellent with rare chance of recurrence and malignant transformation.

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