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Case Report

Cervical sympathetic chain schwannoma presenting as a carotid body tumor

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ABSTRACT

Nerve sheath tumours arising from the sympathetic chain are extremely rare and are a diagnostic challenge. Schwannomas are benign nerve sheath tumours deriving from Schwann cells that occur in the head and neck region in 25-45% of cases. Cervical lesions originate from spinal nerves, the last four cranial nerve roots, or occasionally from the sympathetic chain. Carotid body tumour (CBT), the most common tumour of the carotid bifurcation, presents as a pulsatile mass. Cervical sympathetic chain (CSC) schwannomas are slow growing lesions and may appear pulsatile due to the displacement of vascular structures by the non-vascular mass and thus may mimic a CBT. We here in discuss a case of 22year old female, who underwent surgical resection of a mass at carotid bifurcation, presuming a CBT, but postoperative histopathological examination revealed it a schwannoma.

Keywords: Carotid body tumour, Shwannoma, Cervical sympathetic chain

INTRODUCTION

Schwannoma arising from cervical sympathetic chain (CSC) is an uncommon, slow growing benign nerve sheath tumour originating from myelin-producing schwann cells, may appear as a pulsatile neck swelling due to the displacement of vascular structures when grow beneath the carotid sheath, occasionally misdiagnosed as a carotid body tumor (CBT).[1,2] We here in discuss a case of 22 year old female, who underwent surgical resection of a mass at carotid bifurcation, presuming a CBT, but postoperative histopathological examination revealed it a schwannoma.

CASE REPORT

A 22 year old female presented with a slowly progressive, painless, pulsatile swelling over the left side of neck for 3 months. There was no previous history of tuberculosis, trauma or parotitis.

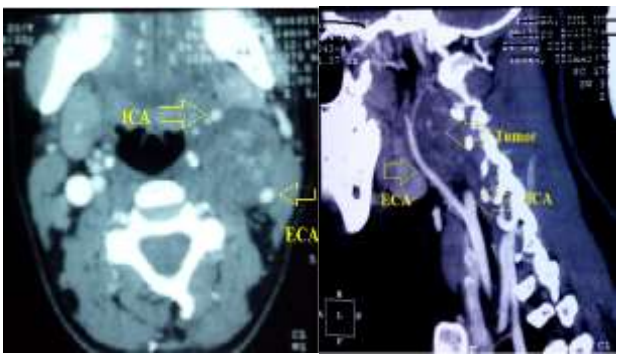

Figure 1: CECT neck reveal 44mmx36mmx35mm soft tissue mass lesion at the carotid bifurcation causing splaying of external carotid artery (ECA) and internal carotid artery (ICA) vessels.

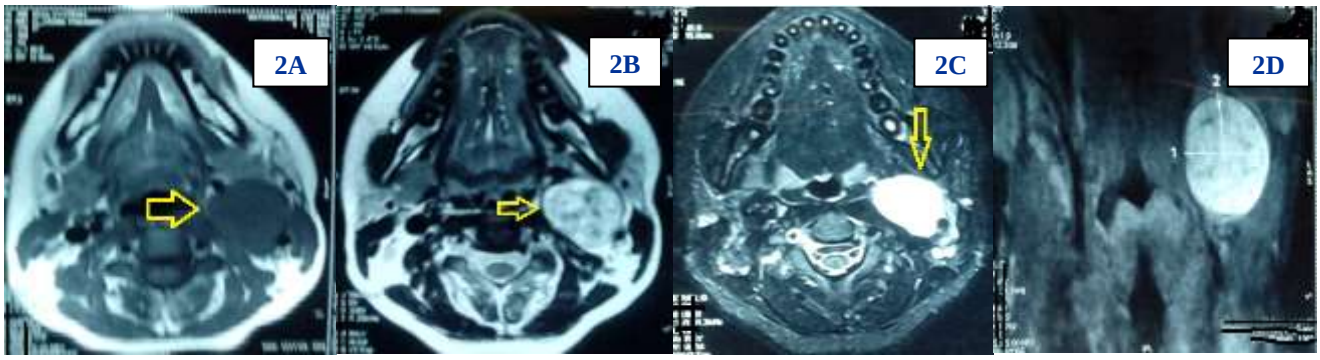

Figure 2: MRI neck: (2A) well marginated 3.4 x 4cm soft tissue mass in the left carotid space causing splaying of carotid vesels which appears hypointense on T1 W images, (2 B) hyperintense on T2W images with salt and pepper appearance (2C, 2D) post contrast enhancement.

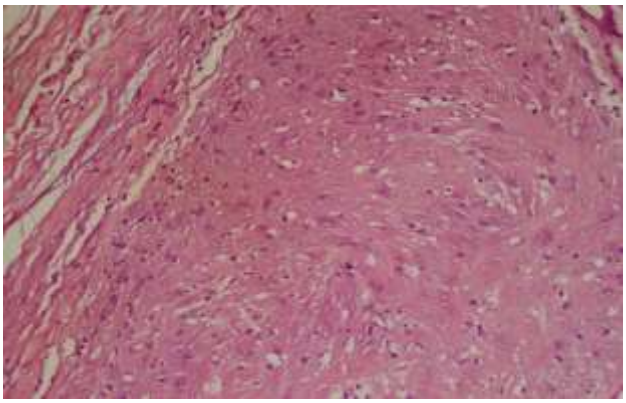

Figure 3: Histopathology of tumor showing benign oval to spindle cells arranged haphazardly and at places forming vesicles in loose myxoid matrix and few ganglion cells seen in capsule suggestive of schwannoma.

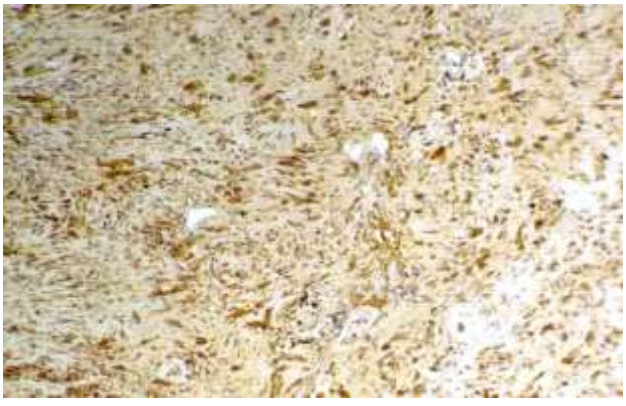

Figure 4: IHC magnification 200X, S-100 positive tumour cells, s/o Shwannoma.

Local examination revealed a 4x3cm size, firm, nontender, pulsatile, mass deep to investing layer of deep cervical fascia located above the level of hyoid and anterior to anterior border of left sternocleido mastoid muscle. Local ultrasonography, carotid Doppler, Contrast CT and MRI of neck affirmed a CBT (Figure 1, 2). Serum and urine catecholamine and vanillylmandelic acid levels were within normal range. Patient was taken for surgical excision with a radial arterial line for continuous blood pressure monitoring and a central venous access line through right internal jugular vein to transfuse fluid and ionotrope in emergency. Patient was operated for CBT, by a standard carotid incision in the left cervical region. Resection of adherent mass to the carotid bifurcation was done after taking proximal and distal control of both external and internal carotid arteries. The mass was appearing encapsulated and plane of dissection could be reached without any difficulty. Throughout the surgery hypotension was avoided and initial part of surgery till the dissection of tumorthe systolic blood pressure was kept more than 120mmHg. Patient had an uneventful recovery and she was discharged on 4th postoperative day. The presumed CBT was sent for histopathological examination which revealed a schwannoma (Figure 3). At 3 month follow up, patient is doing well.

DISCUSSION

Schwannomas are benign nerve sheath tumors originating from the lower four cranial nerves or the CSC in the parapharyngeal space.[1] Like CBT, Schwannomas present with long standing, slow growing mass, mostly asymptomatic and show nonspecific signs on clinical examination.[1] It presents as a pulsatile mass when it grows in close proximity to carotid bifurcation and very often misdiagnosed as CBT. Similar cases of CSC schwannomas have been reported in the literature, where it mimicks a CBT and post resection, the histopathological examination revealed a schwannoma.[1,2] In patients with presumed CBT the preoperative assessment includes history, physical examination, serum and urine estimation of catecholamines, ultrasound of

neck. Contrast CT scanned MRI of neck define the extent and plane of the swelling and rule out the presence of any contralateral tumour. Carotid angiography is useful to assess both the carotid circulation and vascularity of the mass.[3] Surgical excision is the standard treatment. The standard technique for surgery includes wide exposure, identification and preservation of the neurovascular structures, tumour dissection, shunting, and grafting when required.[4] Cranial nerve palsies and Horner's syndrome can occur due to neural infiltration or nerve damage caused during surgery, or edema after surgery.[5] The incidence of neurological damage is usually around 60% with cranial nerves VII, IX, X, and XII.[6] With CSC schwannoma resection, the incidence of Horner's syndrome is high.[7] Depending on the extent of the mass, the carotid artery may have to be clamped for the procedure. During this time, it is important to ensure the adequacy of contralateral circulation to prevent cerebral ischemia. Several methods for monitoring cerebral ischemia are useful in this situation, like electroencephalography, somatosensory evoked potential, transcranial Doppler, jugular venous bulb oxygen saturation, trans cranial non-invasive cerebral oximetry using infra-red light source and measurement of distal carotid stump pressure.[8] Raising blood pressure to improve collateral flow can prevent or reverse changes on the electroencephalogram and thus avoid ischemic injury.

CONCLUSION

In conclusion, CSC Schwannomas always pose a diagnostic challenge due to their nonspecific clinical signs. Most often the diagnosis is made postoperatively after histopathologic examination. The investigations and surgical management in patients presenting with a parapharyngeal pulsatile neck mass are similar to patients with CBT. The intraoperative measures play a major role in preventing the neurological complications related to CBT surgery.

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