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Case Report

Cervical sympathetic chain schwannoma: a case report

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ABSTRACT

Neurogenic tumors including Schwannoma are the rare cause of the lateral neck swelling. We present a case of cervical sympathetic plexus schwannoma that was successfully managed by surgical excision. A 30-year-old male presented with complaints of painless slow growing swelling on the right side of neck of 6 months duration. On examination a single, non-tender, pulsatile swelling of 4×3 cm in the right lateral side of neck was seen beneath the anterior margin of the sternocleidomastoid and just below the angle of the mandible. Magnetic Resonance Imaging (MRI) neck showed a well-defined lesion of size 4.5×4.6×6 cm in the right carotid space with features suggestive of neurogenic tumour arising from sympathetic plexus, likely Schwannoma. Surgical excision of the tumour was done. Intra-operatively a well encapsulated lesion was present in the right carotid sheath arising likely from right cervical sympathetic plexus. Histopathological examination confirmed the diagnosis of Schwannoma. Post-operatively patient developed mild ptosis of right eye; however at 6 months follow-up patient is doing fine without any evidence of residual neurological loss or recurrence.

Keywords: Schwannoma, Neurilemmomas, Sympathetic chain, Neck tumors

INTRODUCTION

Lateral neck swellings in patients are usually thought to be lymph nodes from metastatic carcinomas of the oral cavity, pharynx, larynx, skin or the salivary glands, or due to inflammatory lymphadenopathy. Differential diagnosis also includes the lipoma, cysts, hygroma and aneurysm. Rarely the cause of the lateral neck swelling can be the neurogenic tumour including Schwannoma.

Majority of the neck Schwannoma's arise from the cervical sympathetic chain and the vagus nerve.[1] Schwannoma's, in majority of cases present with asymptomatic swelling in the lateral neck. They may also present with the compressive symptoms such as dysphagia or Horner's syndrome.

We present a case of right cervical sympathetic chain Schwannoma (CSCS) in a 30-year-old gentleman that was successfully managed by surgical excision. We are presenting this case as the CSCS are very rare and the anatomical location of these tumors create diagnostic and surgical challenge to the treating surgeon.

CASE REPORT

A 30-year-old gentleman presented to surgical outpatient department of our hospital with complaints of a painless swelling in the right lateral side of neck since 6 months which was gradually increasing in size. There were no neurological signs and careful evaluation revealed no evidence of primary neoplastic lesions in upper aerodigestive tract. On examination, a single, non-tender pulsatile swelling of 4×3 cm was present in the right lateral side of neck beneath the anterior margin of the sternocleidomastoid and just below the angle of the mandible. No other neck swelling or palpable cervical lymphadenopathy was seen. Videoendoscopic evaluation of upper aerodigestive tract revealed fullness and medial

displacement of right palatine tonsil with mild narrowing of oropharyngeal airway with normal rest of the upper aerodigestive tract and normal movement of vocal cords.

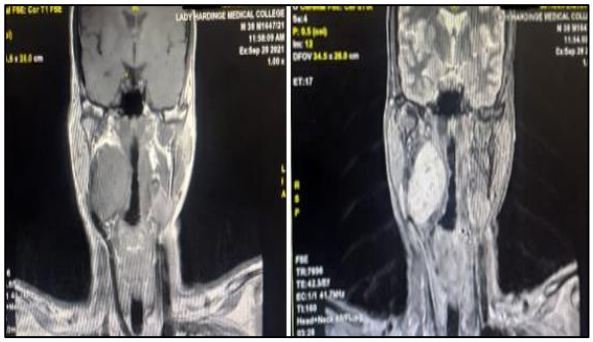

Figure 1. MRI images showing lesion in the right parapharyngeal space causing narrowing of the oropharyngeal airway. (A)coronal T1 image showing isointense lesion (B) coronal T2/STIR image showing hyperintense lesion.

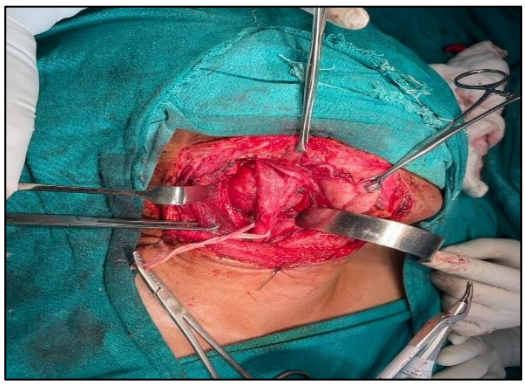

Figure 2: Lesion in the right carotid sheath displacing carotid vessels anterolaterally, IJV posterolaterally, cranially upto the base of skull.

On ultrasonography, (USG) single well defined, hypoechoic lesion with internal vascularity showing arterial wave form lesion of size 4.4×3.5 cm size was present on the right side of neck below the angle of mandible. There was no significant cervical lymphadenopathy on sonography. Magnetic Resonance Imaging (MRI) neck showed a well-defined oblong fusiform shaped lesion of size 4.5×4.6×6 cm in the right carotid space appearing heterogeneously isointense on T1 and heterogeneously hyperintense on T2/STIR with anterolateral displacement of internal and external carotid arteries by the lesion with features suggestive of neurogenic tumour arising from sympathetic plexus likely Schwannoma (Figure 1). The lesion was extending from C1 to inferior aspect of C4 vertebra cranially, anteromedially pushing the parapharyngeal fat and posteromedially, it was abutting the right longus colli muscle. There was no obvious evidence of any perineural invasion or any extension into neural foramina; with well-defined fat planes with surrounding structures and mass effect noted in the form of medial displacement of right palatine tonsil, complete non visualisation of right internal jugular vein (IJV) and mild narrowing of oropharyngeal airway. Contrast enhanced computed tomography (CECT) scan of neck and thorax complimented the findings of MRI without evidence of any other primary / metastatic lesions in the neck or thorax. USG guided fine needle aspiration cytology (FNAC) yielded only blood elements on smear.

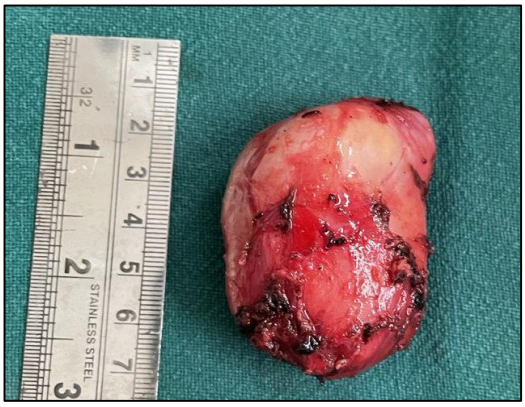

Figure 3: Excised schwannoma.

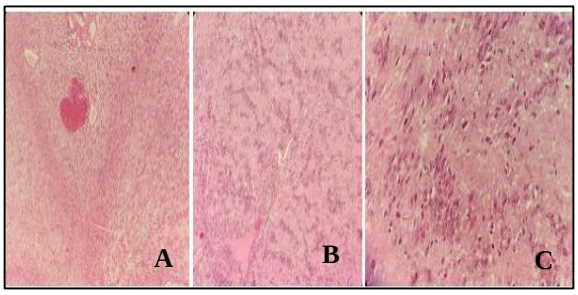

Figure 4 showing microscopic section of the specimen. (A) H and E section showing both hyper and hypo cellular areas; (B) H and E section showing presence of multiple verocay bodies; and (C) high power view of verocay bodies.

Surgical excision of the tumour was done. Intra-operatively, the well encapsulated lesion was present in the right carotid sheath displacing carotid vessels anterolaterally, IJV posterolaterally, medially extending into parapharyngeal space and cranially up to the base of skull without obvious gross involvement of Vagus or Hypoglossal nerve likely arising from right cervical sympathetic plexus (Figure 2). Gross examination of the specimen showed well encapsulated globular greyish soft tissue mass measuring 3.5×4×5 cm. On microscopy both hypercellular and hypocellular areas were seen showing verocay bodies; individual cells were long, slender and wavy with tapered ends. Mitosis was infrequent suggestive of Schwannoma. Post-operatively patient developed mild ptosis of right eye without any other noticeable neurological deficit. However, at 6months follow-up patient is doing fine without any evidence of residual neurological loss or recurrence.

DISCUSSION

Benign nerve sheath tumors constitute approximately 20-27% of tumors in the parapharyngeal space are.[1] Schwannoma's are slow growing peripheral nerve tumors arising from the nerve sheath. 25-45% of all schwannoma's are found in the head and neck region.[2] Schwannomas originate in the cranial (eg, V, VII, IX, X, XI, and XII), sympathetic, or peripheral nerves.[3] The most common presentation of the CSCS is asymptomatic mass in the lateral neck (69.7%). The other common presentation include the vascular disruption(38.2%), cranial nerve deficits(10%), Horner's syndrome(14.5%), pain(5.3%), dysphagia(5.3%).[4]

The reported size in the literature for CSCS has been between 2 and 7.5 cm in major diameter.[5] These tumors grow at least to 2.5-3 cm before they are detected.[6]

Imaging of the neck include CECT and MRI. A mass on CECT pushing the internal carotid artery or common carotid artery anteriorly is suggestive of Schwannoma originating from the sympathetic chain or vagus nerve.[6] On MRI, a Schwannoma may show hypointense/isointense on T1 imaging and shows hyperintense imaging on T2 imaging.

FNAC done in the Schwannoma's usually results in the bloody aspirate and does not have the adequate sensitivity and specificity in diagnosing the Schwannoma's.[7]

The en-bloc surgical resection of the tumour is the treatment of choice as the Schwannoma's are usually benign and rarely has recurrence.[8] The surgical resection of the CSCS is associated with the Horner's syndrome (91.1%), or first bite syndrome (21.1%) or both (15.9).[4] The surgical excision in most of the cases entails extracapsular excision which has a significant morbidity. Intracapsular enucleation of the tumour provides effective long term oncological outcome with improved functional outcomes.[9]

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