CASE REPORT

Synchronous cervical plexus schwannomas: A rare presentation

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Abstract

Schwannomas are benign neural sheath tumours composed of neoplastic Schwann cells, 25-45% of which occur in the head and neck region. Clinical presentation varies according to the anatomical site but the most common presenting symptoms are a slow growing lesion, painless mass, or neurological deficits. Preoperative workup includes imaging studies e.g., ultrasound, computed tomography (CT) and magnetic resonance imaging (MRI). Management options include observation, surgical excision, intracapsular enucleation and radiotherapy. We present a rare case of synchronous cervical plexus schwannomas in a 13-year-old female. The patient presented with a slow growing mass on the right side of the neck, which had been present since early childhood. A percutaneous biopsy confirmed diagnosis of a schwannoma. Intracapsular enucleation was performed, and perioperative findings revealed two synchronous schwannomas in the right supraclavicular region. Synchronous schwannomas of the cervical plexus is a very rare presentation, with no prior cases reported in the literature. Surgical excision remains the treatment of choice in the management of such cases.

Keywords: cervical schwannoma, neck tumour, peripheral nerve sheath tumour, schwannoma, synchronous.

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Introduction

Schwannomas are slow growing benign neural sheath tumours composed of neoplastic Schwann cells,[1] of these, 25-45% occur in the head and neck region, most commonly in the lateral neck, parapharyngeal space and temporal bone.[2] These tumours have been reported to arise from cranial, peripheral or autonomic nerves.[1] Clinical presentation may vary according to the anatomical site of the tumour, however the most common symptoms include a slow-growing lesion, painless mass or neurological deficits.[2] The pre-operative workup includes imaging modalities such as ultrasound, CT, and MRI scans. Surgical management options include total excision, subcapsular excision and intracapsular enucleation of the tumour,[3] followed by post-operative histopathological examination to confirm the diagnosis. Post-operative Horner's syndrome is the most common complication following the excision of masses along cervical sympathetic chain. Dissection in the parapharyngeal space may also cause lower cranial nerve injury, which can manifest as vocal cord paralysis, dysphagia or shoulder weakness.[4]

We present a rare case of synchronous cervical schwannomas that were successfully managed by intracapsular enucleation without motor deficits. To our knowledge such a case has not yet been reported in the literature.

Case Report

A 13-year-old female presented to the ENT and head and neck out-patient department at Northwest General Hospital and Research Centre, Peshawar, on 13th March 2023. She complained of a slow growing mass on the right side of her neck, which had been present since childhood and had increased in size over the past few years. Fine needle aspiration cytology (FNAC) was performed but was inconclusive. Therefore, percutaneous biopsy had to be undertaken. After which the patient developed sharp paroxysmal stabbing pain exacerbated by movement of the right hand. The biopsy confirmed the diagnosis of a schwannoma. On clinical examination, there was a 5 x 6 cm mass noted in the right supraclavicular region. The mass was firm in consistency, tender on palpation with no fluctuation and mobility. There were no signs of No neurosensory or motor dysfunction. The patient was advised to undergo an MRI of the neck with contrast, which revealed a large well-defined lobulated mass in the supraclavicular fossa measuring 4.5 x 2.6 x 2.6 cm. This mass was found to be in continuity with the right C4-C5 exiting nerve, which appeared also thickened at the level of neural foramen. Additionally, the scans identified another lesion just below the first, measuring 6.3 x 1.9 x 2.5 cm and in continuity with the right C5-C6 exiting nerve. The two lesions showed heterogeneous high signals on T2 and isointense on T1 (Figure 1-2).

The management plan was discussed during a multidisciplinary meeting, and surgical excision with

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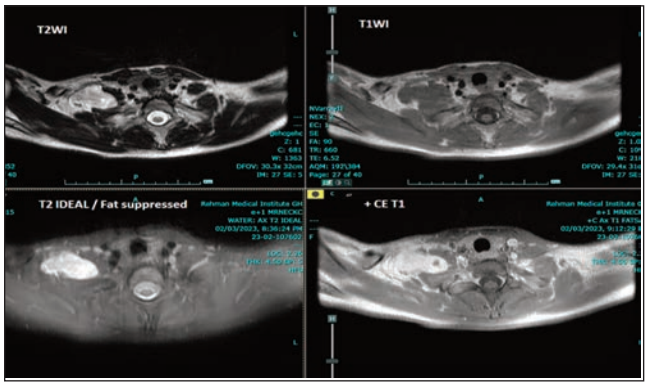

Figure-1: Multisequential MR axial images showing large lobulated ovoid lesion in right supraclavicular fossa appearing hyperintense on T2WI, isointense on T1WI, intensely hyperintense on WTAR/ Fat suppressed image and shows post contrast avid heterogeneous enhancement. It has smooth margins with displacing rather than invading regional soft tissue.

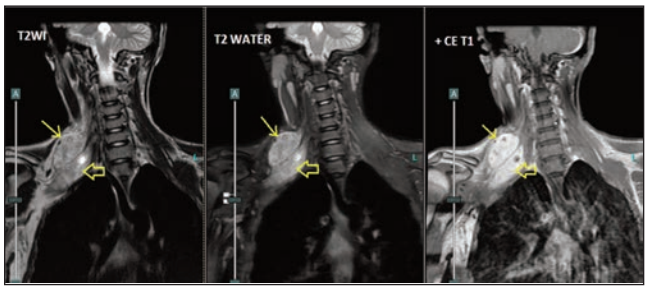

Figure-2: Coronal MR images showing two large ovoid schwannomas in right supraclavicular fossa. Note the long axis of the lesions is parallel to long axis of C4-C5 and C5-C6 nerves. Few tiny areas of necrosis seen in both upper and lower lesions appearing as non-enhancing intralesional foci. No extension into spinal canal seen.

adjuvant treatment, in case of residual or recurrent disease was planned. The Patient consent to publish the case report was taken at the time of surgery. Intracapsular enucleation was performed through a right cervical crease incision. Perioperative findings included two synchronous schwannomas in the right supraclavicular region. One mass was located deep and anterior to the sternocleidomastoid muscle, while the other was positioned anteriorly extending deep to the clavicle. The cervical nerve roots and sheath were was preserved during the procedure. Histopathologic examination confirmed both masses as schwannomas, showing a circumscribed, nearly encapsulated tumour composed of uniform spindle cells with a fascicular arrangement. The cells showed ill-defined cytoplasm and nuclei with dense chromatin and no evidence of mitotic figures or necrosis.

Post-operatively, the patient recovered without any neurosensory deficits or loss of motor function and was advised to undergo physiotherapy. During follow up, the patient reported significant improvement in pain. She was referred to a radiation oncologist for further assessment and administration of radiation therapy, if necessary. A post-operative MRI showed no signs of residual or recurrent disease. The patient is under regular follow up with no complaints at three months post-surgery.

Discussion

Neurogenic tumours, initially referred to as 'Neurinomas' by Verocay in 1910 and later termed 'Neurilemmomas' in 1935, are now known to originate from Schwann cells.[1] Schwannomas are slow growing, benign neural sheath tumours, 25-45% of which occur in the head and neck region, most commonly in the lateral neck, parapharyngeal space and temporal bone.[2] Other reported sites include the scalp, face, pharynx, parotid gland, middle ear, and external acoustic canal. Intraoral lesions show a predilection for the tongue, followed by palate, buccal mucosa, lip, and gingiva.[5] Schwannomas usually arise from the sensory divisions of cranial nerves, most commonly the vestibular nerve. Vagus, trigeminal, and facial nerves are less commonly affected and involvement of motor nerves is uncommon. Extracranial head and neck locations are unusual for non-vestibular schwannomas. Schwannomas of the cervical nerve plexus are equally rare, and synchronous tumours are even rarer.[6]

The most common age group affected is generally reported between 20-60 years, with no gender predilection.[5] These tumours are solitary, although they can occur in multiple areas, often associated with neurofibromatosis II and schwannomatosis.[2]

Cervical schwannomas usually present as painless masses without neurological deficits. Boumaza et al.[7] reported that 43% of patients presented with a painless isolated neck mass and while 8% experienced associated symptoms related to either compression by the mass or motor weakness of the affected nerve. In 49% of cases, no neck mass was reported and patients presented with symptoms such as pain, dysphagia, dysphonia, parapharyngeal discomfort, or Horner syndrome.

These tumours present a diagnostic challenge due to their slow growing nature and non-specific clinical presentation. They remain asymptomatic for a long time. Clinically, they are often misdiagnosed as other common benign lesions such as pleomorphic adenomas, fibromas or mucous retention cysts.[8] On clinical examination, they appear as benign masses and are sometimes tender. CT and MRI are essential for diagnostic tools, while ultrasound and FNAC may also be used. However, although FNAC has limited sensitivity and specificity in diagnosing schwannomas.[9] Histopathological examination remains the gold standard for diagnosis.

In elderly or severely ill patients, schwannomas may be monitored. For others complete surgical resection is the treatment of choice. Yasumatsu et al.[4] reported a 100% nerve palsy rate at 6 months post-complete resection compared to 31% with intracapsular enucleation. Neural monitoring during surgery helps to preserve motor nerve fibres on the tumour surface and guides surgical decisions.[10]

Conclusion

Cervical schwannomas are rare tumours of the head and neck region. To the best of our knowledge, synchronous cervical schwannomas have not been reported in the literature. Accurate pre-operative clinical diagnosis, including identification of the nerve of origin, is essential for ensuring optimal management. MRI remains the most essential imaging modality for diagnosis and surgical planning. Intracapsular enucleation is the treatment of choice, as it significantly improves motor function while minimizing the risk of neurological deficit, especially when combined with neural monitoring.

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Author Contribution:
HZ: Literature review, writing and final approval.
HM: Writing, drafting and revision.
IMK: Diagnosed and managed the case, supervision and final approval.
HS: Acquisition of clinical details.