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Case Report

A rare case of cervical sympathetic schwannoma

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ABSTRACT

Schwannomas are benign neoplasms arising from myelinating Schwann cells. Nerve sheath tumours arising from the sympathetic chain are very sporadic, with very few cases reported that presented with Horner's syndrome. The authors describe its radiological and histological features and surgical management in this case report. A sixty-eight-year-old man presented with throat discomfort and hoarseness of voice for three months. He had a palpable mass on the left side in the submandibular region on neck examination. The patient had left recurrent laryngeal nerve palsy and Horner's syndrome on general examination. MRI with contrast was performed to see the extent of the tumour. Exploratory cervicotomy was performed; the tumour was seen originating from the lower cervical sympathetic chain posterior to the carotid sheath. Histopathological examination confirmed the diagnosis of cervical sympathetic schwannoma. Cervical sympathetic schwannoma is a benign, slow-growing neoplasm with varied presentation. Presentation with Horner's syndrome is a rare but reported presentation. Radiological imaging plays a vital role in the identification and surgical management. For symptomatic tumours, exploratory cervicotomy is the preferred approach for its wide exposure. Histopathological examination is confirmatory for the diagnosis.

Keywords: Cervical sympathetic schwannoma, Horner's syndrome, Histopathological examination

INTRODUCTION

Schwannomas are benign neoplasms arising from myelinating Schwann cells.[1] Although they occur throughout the body, up to 45% are found in the head and neck region, with many arising in the parapharyngeal space as either vagus nerve cervical schwannoma (VNCS) or sympathetic chain cervical schwannoma (SCCS).[2-4] Nerve sheath tumours arising from the sympathetic chain are sporadic, with very few cases described in the literature. On microscopy, schwannomas are encapsulated, solid or cystic tumours. They can be composed of two cellular zones: Antoni type A, densely arranged with spindle-shaped Schwann cells and areas of palisading nuclei, and Antoni B, characterised by a hypocellular arrangement and a large quantity of myxoid tissue.[5] This report presents a case that manifested with throat discomfort and hoarseness of voice for three months. A swelling was palpated in the neck region on the left side. The tumour was subsequently visualized on Gadolinium-enhanced MRI in the left carotid space and was confirmed on excision biopsy.

CASE REPORT

A sixty-eight-year-old man presented to our outpatient clinic with throat discomfort for three months. It was associated with a change in voice for two months. There was no difficulty with swallowing or breathing, no history of fever, and the patient had no loss of appetite or weight loss. The oral examination was unremarkable. On examining the neck, a swelling was palpated, on the left side in the submandibular region measuring 3×2 cm, firm in consistency, non-tender on palpation. On general examination, the patient had slight drooping of the left eyelid and miosis. The complete neurologic assessment

revealed left recurrent laryngeal nerve palsy and Horner's syndrome. Features of Horner's syndrome include pupillary miosis with ptosis, enophthalmos, facial anhydrosis of the ipsilateral face with loss of ciliospinal reflex. On fiberoptic examination of the larynx, left vocal cord palsy was documented (Figure 1). To further investigate, Gadolinium-enhanced MRI of the neck was requested, which showed a well-defined oval-shaped T1 iso to hypointense, T2 hyperintense mass lesion centered in the left carotid space posterior to the carotid artery and internal jugular vein. It measured 5.8×2.9×2.4 cm displacing the common carotid artery and internal jugular vein anteriorly. The lesion showed heterogeneous contrast enhancement and extended up to the level of carotid bifurcation superiorly (Figure 2). The multidisciplinary team discussed the MRI findings with the patient, and it was decided to proceed with an exploratory cervicotomy. A provisional diagnosis of cervical sympathetic schwannoma was made, along with a differential diagnosis of vagal schwannoma and paraganglioma. On surgical exploration, it was found that the tumour was originating posterior to the carotid sheath arising from the lower cervical sympathetic chain. The carotid artery and internal jugular vein were displaced anteriorly. The tumour specimen was sent for histopathological examination. It was composed of cellular (Antoni A) and hypocellular (Antoni B) areas. The tumour was composed of Schwann cells arranged in diffuse sheets and short fascicles intermingled with scanty fibrovascular stroma. The cells had an elongated pump nucleus with a moderate amount of cytoplasm. Focally the cells show nuclear palisading with Verocay bodies. In the tumour specimen, the sympathetic chain was visualised; thus, the diagnosis of schwannoma arising from the sympathetic chain was confirmed (Figure 3). In the postoperative period, the patient developed first bite syndrome. Hoarseness of voice improved, but Horner's syndrome persisted after the surgery. The drain was removed on 3rd postop day.

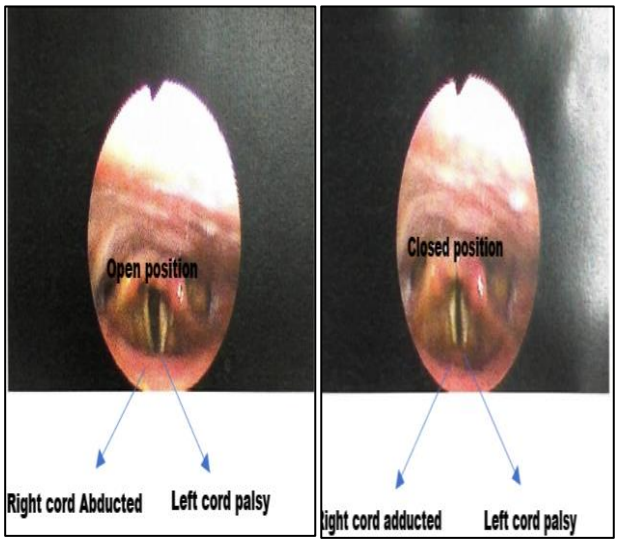

Figure 1: Showing fiberoptic laryngoscopic picture of left vocal paralysis.

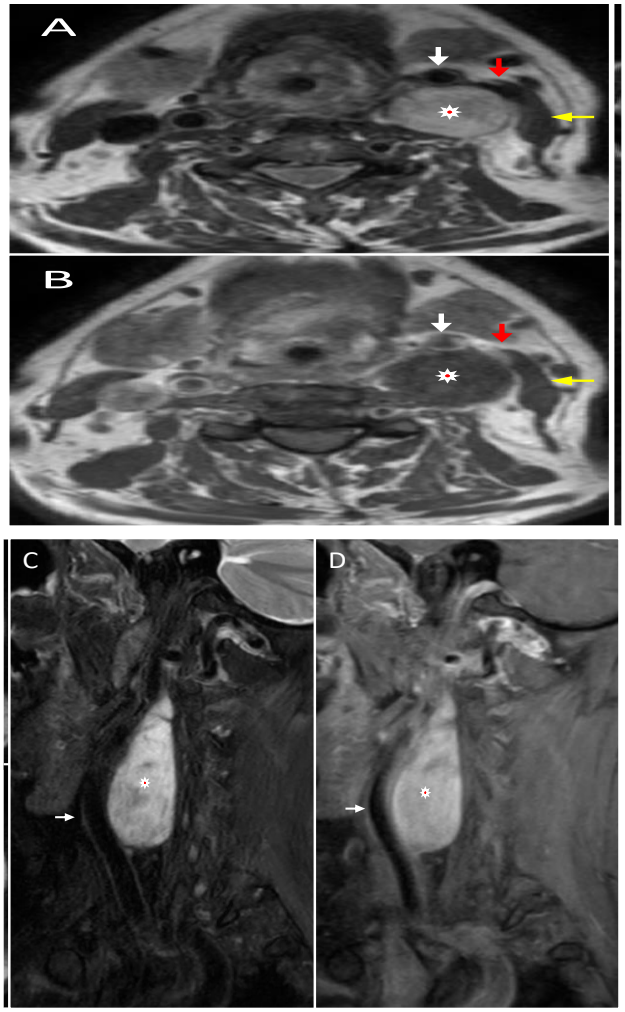

Figure 2 (A-D): MRI neck with contrast: axial T2, axial T1, sagittal STIR shows a well-defined oval-shaped T1 isointense, T2 hyperintense mass lesion (star) centred in the left carotid space posterior to the carotid artery (white arrow) and internal jugular vein (red arrow) and medial to the sternocleidomastoid muscle. The common carotid artery and internal jugular vein are displaced anteriorly by the lesion. Sagittal postcontrast T1: The lesion shows intense contrast enhancement.

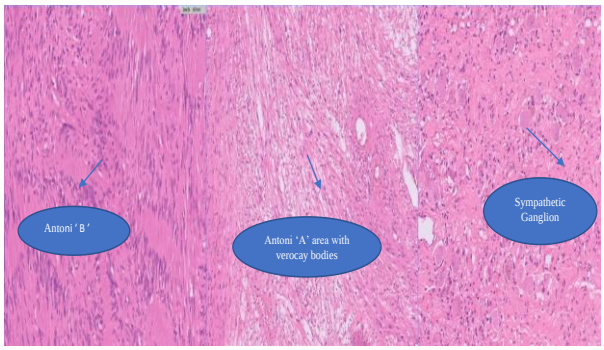

Figure 3: HPE Slide of resected tumour specimen showing Antoni 'B' area, Antoni 'A' area with verocay bodies, and sympathetic ganglion.

DISCUSSION

Ritter first reported cervical schwannoma in 1899.[6] Schwannomas are benign neurogenic tumours originating from Schwann cells. SCCS is generally adult-onset (20-70 years of age), with a balanced gender ratio.[7] The tumour may develop in the parapharyngeal space from the last four cranial nerves and cervical sympathetic chain, with the tenth nerve most frequently involved.[8,9] The clinical presentation is usually atypical with an isolated asymptomatic lateral cervical mass of progressively increasing volume.[7] There are very few cases described of SCCS in literature, and even among them, presentation with Horner's syndrome is seldom seen. Tenth nerve involvement and Horner's syndrome can be explained by the pressure effect of the tumour on neighbouring structures. Presentation with Horner's syndrome is infrequent as these tumours are non-infiltrating; also, the sympathetic chain runs in the loose fascial compartment, so injury due to compression is extremely rare. Imaging studies play a central role in diagnosing head and neck schwannomas. Regardless of the nerve of origin, schwannomas, in general, are hypodense in comparison to muscle on CT without contrast.[10] MRI of schwannomas reveals relatively low signal intensity on T1-weighted images and high signal intensity on T2-weighted images.[4] In MRI, split fat sign, target sign and fascicular sign can be seen. The presence of a fat plane between the parotid and the lesion truly differentiates a parapharyngeal lesion from an extension of the deep lobe of the parotid. The tumours of the deep lobe of the parotid can reach the parapharyngeal space through the stylomandibular tunnel. Surgery is the treatment of choice in schwannoma.[8,11] Several approaches have been described for the excision of tumours in the parapharyngeal space, including transcervical, trans-parotid, infratemporal, and transoral approaches. We preferred the transcervical approach as it offers wide exposure for the complete excision of the tumour in the parapharyngeal space. It is of utmost importance to identify and preserve all vascular and neural structures intraoperatively, namely glossopharyngeal, vagal, spinal accessory, hypoglossal and superior laryngeal nerves. By careful identification and preservation, postoperative morbidity can be avoided. Microscopically schwannomas have Antoni A and B areas. Antoni A regions are composed of more densely arranged cells with specific areas of palisading nuclei arranged in rows. In contrast, Antoni B regions tend to be more hypocellular, with a loose and disorderly arrangement. Verocay bodies are more distinctive of schwannomas than Antoni A and Antoni B pattern.[12] Verocay bodies are a component of "Antoni A", which are the dense areas of schwannomas located between palisading spindle cells found in neoplasms, two nuclear palisading regions and an anuclear zone makeup one Verocay body.[13] The proliferation index is low, and on immunohistochemistry, S-100 protein is positive in all schwannomas and neurofibromas.[14] Patients are usually extubated after surgery. Patients having an external approach can usually commence an oral diet after surgery. First bite syndrome is a known complication after parapharyngeal surgery. It presents as acute and intense pain in the parotid region with the first bite. Symptoms improve on subsequent bites. It is thought that first bite syndrome may be caused by loss of sympathetic input to the parotid gland after severing of the cervical sympathetic chain resulting in a denervation hypersensitivity of the sympathetic receptors that control myoepithelial cells in the parotid gland.[15]

CONCLUSION

Cervical sympathetic schwannoma is a benign, slow-growing neoplasm with varied presentation. Radiological imaging plays a vital role in the identification and surgical management. For symptomatic tumours, exploratory cervicotomy is the preferred approach for its wide exposure. Careful identification and preservation of vascular and neural structures are of utmost importance in decreasing postoperative morbidity and mortality. Recurrence is unusual after complete surgical removal. The presence of Antoni A, Antoni B, Verocay bodies and sympathetic chain on histopathological examination is confirmatory for diagnosis.

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