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Case Report

Lower Cranial Nerves Schwannoma; A Rare Case Report

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ABSTRACT

A schwannoma, also known as a neurilemoma, is a tumour that arises from Schwann cells, which are responsible for the formation of the myelin sheath covering peripheral nerves. Schwannomas typically present as single, benign tumours of the nervous system. They are most commonly found in the head and neck region, particularly along the eighth cranial nerve (vestibulocochlear nerve) in the internal auditory canal.

However, in some cases, Schwannomas can present in multiple forms. This condition is known as multiple schwannomas or schwannomatosis. It is less common than a single, isolated schwannoma. In these cases, tumours can arise from various points along the peripheral nervous system, including cranial nerves, spinal roots, brachial and lumbosacral plexuses, or major peripheral nerves.

We have an unusual case involving bilateral Schwannomas that are likely originating from the lower cranial nerves on right side with in involvement of hypoglossal canal and on the left side, it's suggested to be originating from the trigeminal nerve.

Key Words: *schwannomatosis, lower cranial nerves, rare case report*

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INTRODUCTION

Schwannoma arises from the myelinating cells of the cranial or peripheral nervous system, specifically composed of Schwann cells that typically produce the myelin sheath covering the nerve. Additionally, Schwannomas grow eccentrically within a capsule formed by the parent nerve[1,2]. Due to their slow growth, symptoms can range from none at all to cranial nerve palsy. Additionally, symptoms may arise from the mass effect exerted on nearby structures, denervation injury, or even hydrocephalus. In this context, we are presenting a case involving schwannomas affecting most of the lower cranial nerves[1].

CASE PRESENTATION

We present a case involving a 31-year-old woman with no known co-morbidities who presented to her primary physician with complaints of progressive right sided weakness and slurring of speech for the last 1 year. Upon physical examination, the patient was fully alert. Further assessment revealed lower cranial nerve dysfunction. She was advised an MRI scan by her attending doctor after which she presented in our department and her MRI brain was done. Her MRI showed a lesion with large lobulated cystic mass in right cerebellopontine angle, that was extending cranially upto aqueduct of sylvius and caudally upto right cerebellomedullary cistern with infiltration into the right hypoglossal canal causing its expansion .It was causing compression of fourth ventricle resulting in dilatation of lateral and 3rd ventricle. Another solid cum cystic mass was seen in left cerebellopontine angle, extending into pre pontine cistern and meckel's cave causing its expansion. findings were concluded as bilateral schwannoma likely arising from lower cranial nerves possibly hypoglossal or vagus nerve on right side and from trigeminal nerve on left side.

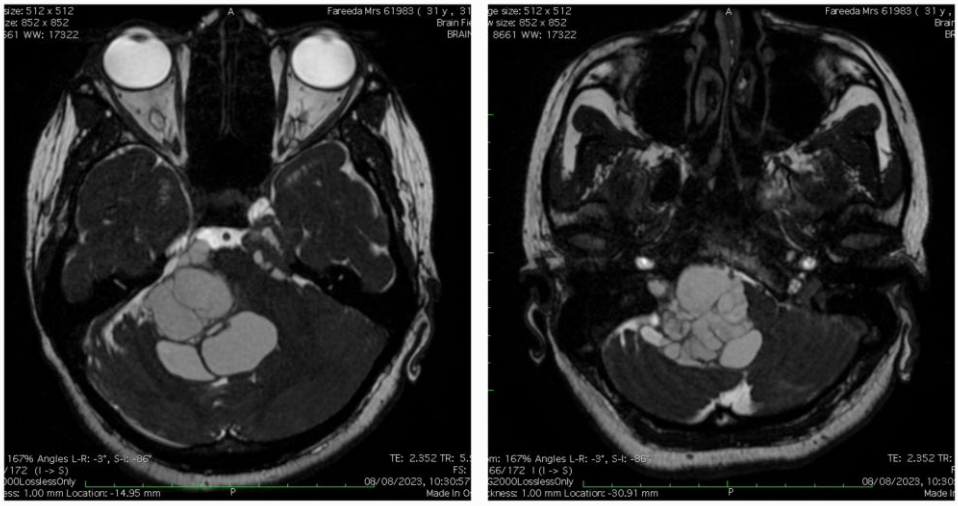

Figure 1 MRI brain showing cystic lobulated mass in rightprepontine and cerebellomedullary cisterns with infiltration into righthypoglossal canal

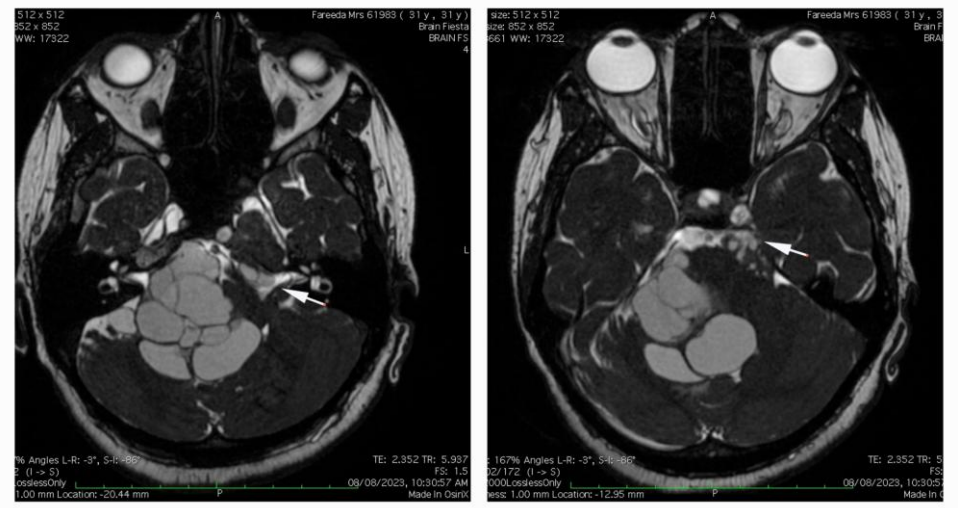

Figure 2 MRI brain showing small solid cum cystic mass in left prepontinecistern with infiltration into left Meckel's cave

DISCUSSION

A nerve sheath tumor primarily composed of Schwann cells, which create a protective layer around peripheral nerves. Schwannomas represent approximately 7% to 10% of all primary intracranial tumors[1,2,3].

Schwannomas derived from lower cranial nerves (IX–XII) in the cervical region, jugular foramen, and hypoglossal canal is relatively infrequent. In cases involving the cervical region, the originating nerves were identified in 62% of cases, which included the brachial plexus, facial nerve, trigeminal nerve, vagus nerve, and sympathetic chain[4].

In over 95% of patients with NF2, a defining characteristic is the eventual development of bilateral vestibular Schwannomas[1]

These tumors can indeed lead to symptoms due to their local mass effect, such as nausea and headache resulting from compression of the brainstem. Alternatively, they may cause specific cranial nerve deficits that can guide a targeted examination of the affected cranial nerves. For instance:

- Diplopia may lead to an examination of CN III, IV, and/or VI.
- Facial numbness, paresthesia, or pain may prompt an evaluation of CN V.
- Facial paresis may indicate a need to assess CN VII.
- Vertigo or hearing loss may warrant an evaluation of CN VIII.
- Dysphagia, dysphonia, or dysarthria may prompt an examination of CNs IX–XII.

Vestibular schwannomas are the most common, comprising 90% of cases, followed by schwannomas originating from the trigeminal, facial, and other lower cranial nerves.

On magnetic resonance imaging (MRI), Schwannomas typically appear isointense to hypo intense on T1-weighted

images, hyper intense on T2 and FLAIR images, and exhibit variable enhancement after contrast administration.(2)The heterogeneous appearance and enhancement pattern can be attributed to factors like necrosis, cystic degeneration, and hemorrhage, which is visualized as a loss of signal on gradient imaging[2,5], in our case large lobulated cystic mass in right cerebellopontine angle, another mass solid cum cystic is seen in left cerebellopontine angle.

Schwannomas can also display a target appearance on T2-weighted images, characterised by central hypo intensity and peripheral hyper intensity, a feature that is typically associated with neurofibromas[2].

On computed tomography (CT), associated bony changes, such as bone remodelling and widening of neural foramen, are better visualized[2].

In the view of literature, Trigeminal Schwannomasrank as the second most prevalent form of intracranial Schwannomas according to literature. These tumors can affect distinct segments of the trigeminal nerve, such as the preganglionic (found in the cisternal region), ganglionic (located within Meckel's cave), postganglionic (within the cavernous sinus), or foraminal segments. In cases where both preganglionic and ganglionic segments are involved, they often exhibit a characteristic dumbbell-shaped appearance. Symptoms of trigeminal Schwannomas may encompass facial numbness, pain, and tingling sensations. If the lesion impacts the mandibular division, it can lead to observable changes indicative of muscle denervation in the masticatory muscles.

Distinguishing trigeminal Schwannomas from other conditions can be challenging, as potential differential diagnoses span cerebellopontine angle meningioma, large vestibular schwannoma, ependymoma, perineural metastasis spread, atypical pituitary macro adenoma, internal carotid artery aneurysm, and various skull base lesions like chondrosarcoma and chordoma[2].

Schwannomas of the vagus nerve in the view of literature can involve various anatomical regions, including the cerebello-medullary cistern, jugular foramen, suprahyoid, or infrahyoid carotid spaces. They can displace the carotid bifurcation, carotid arteries, and internal jugular vein. Clinical presentations can range from no symptoms to potential vocal cord paresis. In cases of compression, clinical features of other lower cranial nerve paresis may also manifest.

The potential differential diagnoses for vagus nerve Schwannomas align with those for glossopharyngeal nerve Schwannomas, encompassing Schwannomas of the 9th and 11th cranial nerves, as well as paragangliomas located within the jugular foramen, suprahyoid, and infrahyoid carotid spaces.

Clinical signs of hypoglossal nerve schwannoma encompass dysarthria, tongue movement difficulties, tongue deviation to one side, and hemi atrophy. In cases of hypoglossal schwannoma, involvement can occur in the cerebello-medullary cistern, hypoglossal canal, suprahyoid neck, and the floor of the mouth. The presence of hemi atrophy and the appearance of one half of the tongue with fat intensity on MRI can provide a definitive diagnosis.

Differential diagnoses to consider include paragangliomas located in the carotid space, squamous cell carcinoma, and tumors of the salivary glands affecting the lingual space and floor of the mouth.

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CONCLUSION

These tumors are indeed exceptionally rare and can manifest with deficits related to lower cranial nerves. Magnetic resonance imaging proves to be a valuable diagnostic tool in such cases. The intention of this case report is to contribute additional data on the clinical presentation, affected patient population, and the diagnostic as well as surgical approaches in managing this exceptionally uncommon tumor. This report is likely to be of significant interest for medical professionals dealing with such cases.

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