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RESEARCH ARTICLE

INTRACRANIAL HYPOGLOSSAL SCHWANNOMA IN A 46-YEAR-OLD FEMALE: A RARE CASE

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ABSTRACT

Schwannomas are benign tumours that originate from the myelin sheath of peripheral nerves. They may be found in any part of the body. In over 90%, they present as vestibular schwannomas, arisng from 8th cranial nerve . Schwannomas that arise from the hypoglossal nerve (12th cranial nerve), are very rare, accounting for only 5% of all nonacoustic intracranial schwannomas. Till date, very less cases of hypoglossal schwannomas have been reported in the literature.Due to their scarcity, early diagnosis of these lesions is difficult and important for further management . Thus , Magnetic resonance imaging (MRI) is the routine imaging protcol for it. They are usually seen between the third and fifth decades of life, with no sex predilection. In the present case report, the case of a 46-year-old female presented with both eyes swelling , nystagmus , Imbalance while walking since 1month. After permorming MRI and diagnosing it ,the patient underwent excisional surgery, and pathological evaluation of the specimen confirmed the diagnosis of benign schwannoma with Antoni areas A and B.

INTRODUCTION

A schwannoma is a benign nerve sheath tumor composed of spindle cells in a variably compact (Antoni type A neurilemoma pattern) or loose (Antoni type B neurilemoma pattern) .This tumor may arise along the course of any nerve sheath of the peripheral nerves or cranial nerves . Intracranial schwannomas have rarely been reported .The majority (approximately 90%) of CN schwannomas arise from the vestibulocochlear nerve (CN VIII), with the next most commonly involved nerves being the trigeminal (CN V) and facial (CN VII) nerves, Hypoglossal schwannomas were reported for the first time by De Martel in 1933. These tumors can be completely intracranial, intracranial/extracranial, or completely extracranial. Schwannoma of the hypoglossal nerve is infrequent because of their rarity, thus early diagnosis of these lesions is essential for further management. Most common symptoms of the hypoglossal schwannoma include headache and dizziness. While the most distinguishing clinical findings of patients with extracranial hypoglossal nerve schwannoma are unilateral tongue atrophy and fasciculations but they can also be asymptomatic initially. Early diagnosis and surgical intervention can preserve the function of nerve.

CASE HISTORY: In our present report, a 46-year-old Indianfemale patient presented to OPD with chief complaints of slurring of speech, eyes swelling, nystagmus, imbalance while walking since 1month. Her past medical and family history were unremarkable. Patient was then advised MRI brain and orbit (Plain + contrast) for further evaluation.

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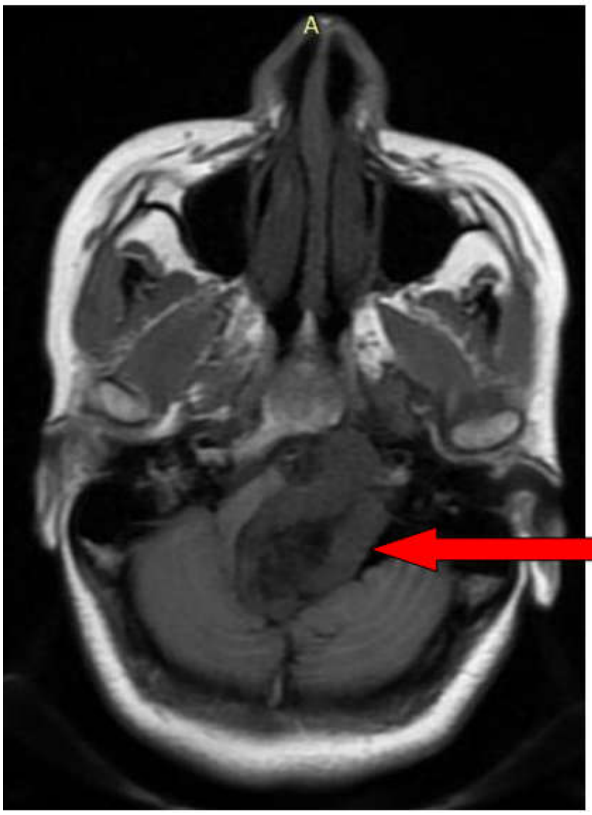
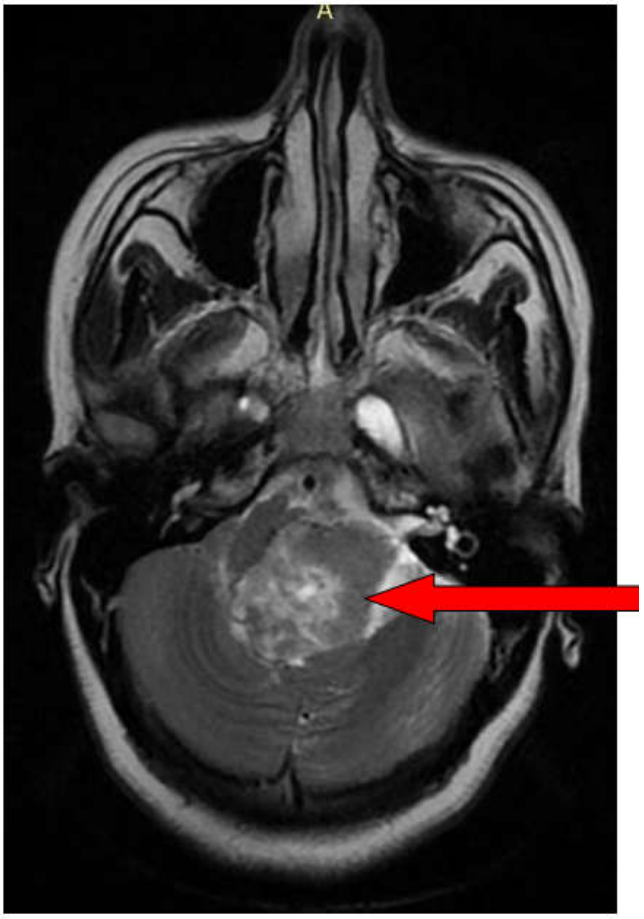
MRI orbit revealed no significant abnormality , however brain revealed a well defined extra axial altered signal intensity in the left anterior and inferior portion of posterior fossa adjacent to the left cerebello-medullary angle cistern which measured approximately 3.5 x4.3 x 5.2 x cm (CC x APx TR) . It appeared heterogeneously hyperintense on T2W and FLAIR and hypointense on T1W. No evidence of restriction is noted on diffusion weighted images. Few hypointense foci of blooming were noted on gradient images suggestive of calcifications. The lesion showed strong and heterogeneous peripheral enhancement on post contrast study with central non enhancing cystic / necrotic portion. Inferiorly the lesion was extending into the foramen magnum and laterally into the hypoglossal canal of skull base with its widening. Superiorly it was reaching up to the internal auditory meatus without any extension into it. Medially it was extending posterior to pons and medulla and significantly compressing the pons and medulla and displacing them right anteriorly. It was also displacing the 4th ventricle posteriorly and towards left without any obvious intraventricular extension. No evidence of hydrocephalus was noted. Medially the lesion is also crossing midline towards right side and indenting the bilateral cerebellar hemispheres and vermis.

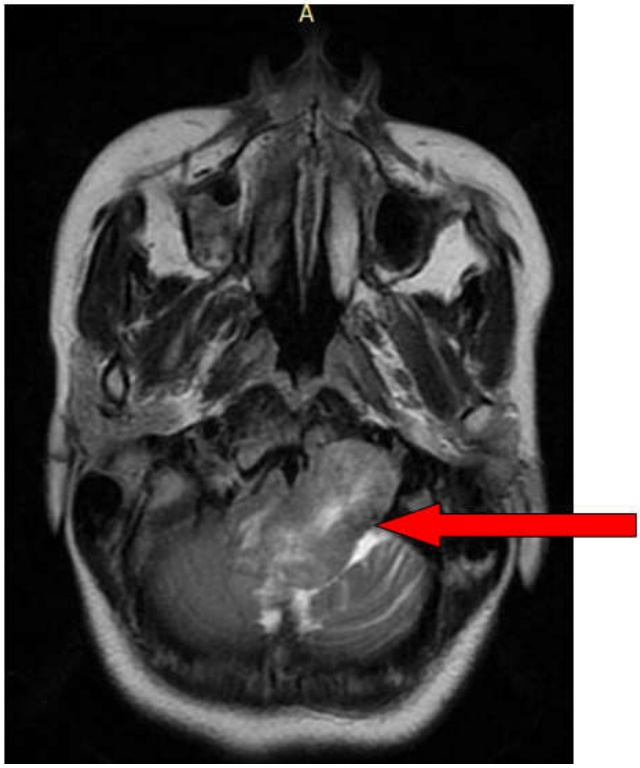
Based on imaging characteristics possibility of neurogenic lesion like Schwanomma of the left hypoglossal nerve or paraganglioma was likely. Suspecting that this tumor was either schwannoma or a paraganglioma, surgical excision of this tumour was performed which revealed an oval-shaped mass measuring 3.5 × 4 cm which was extending into the hypoglossal canal. The mass was in continuity with the hypoglossal nerve. The specimen was then sent for pathological examination.



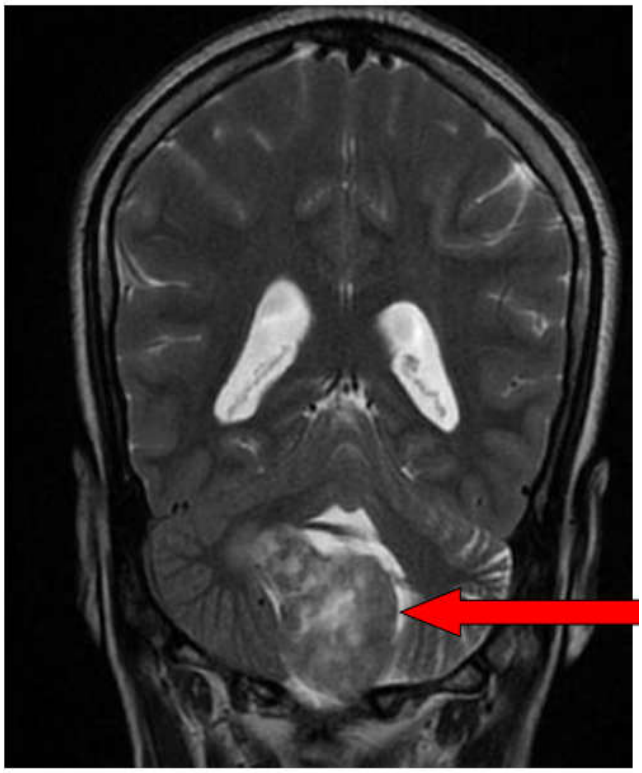
T1 W axial image of the brain showing well defined extra axial heterogeneously hypointense lesion in the left anterior and inferior portion of posterior fossa adjacent to the left cerebello-medullary angle cistern. Inferiorly the lesion is extending into the foramen magnum and laterally into the hypoglossal canal of skull base with its widening.



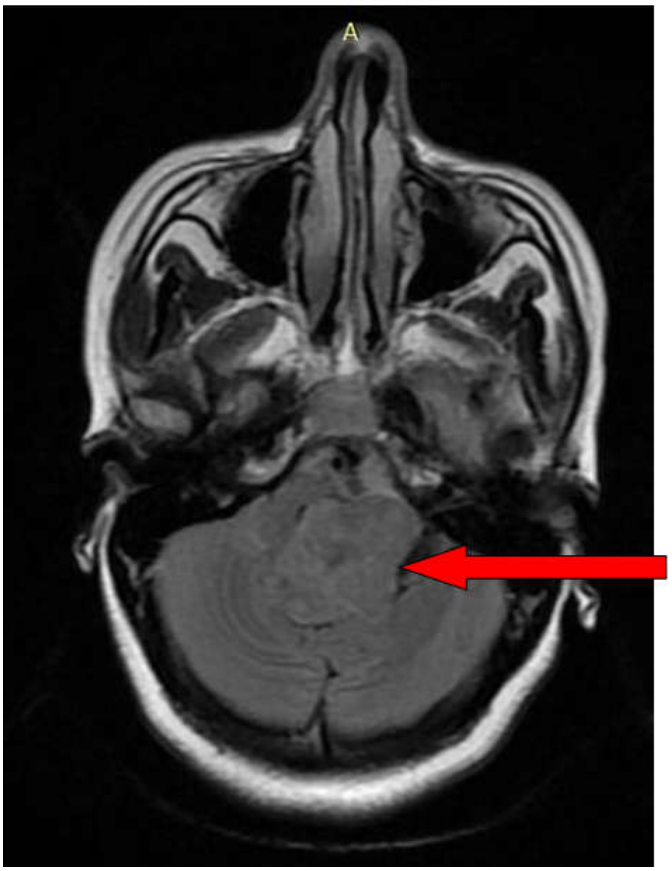
T2 W axial image of the brain with its extension superiorly upto the internal auditory meatus and medially extending posterior to pons and medulla and significantly compressing the pons & medulla and displacing them right anteriorly.



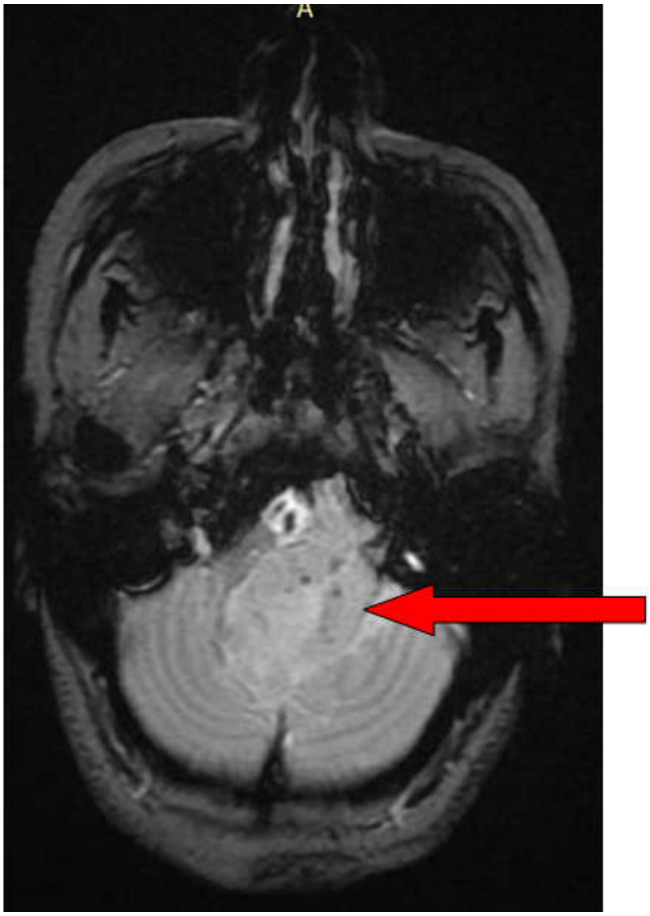
T2 W axial image of the brain showing well defined extra axial heterogeneously hyperintense lesion in the left anterior and inferior portion of posterior fossa adjacent to the left cerebello-medullary angle cistern.



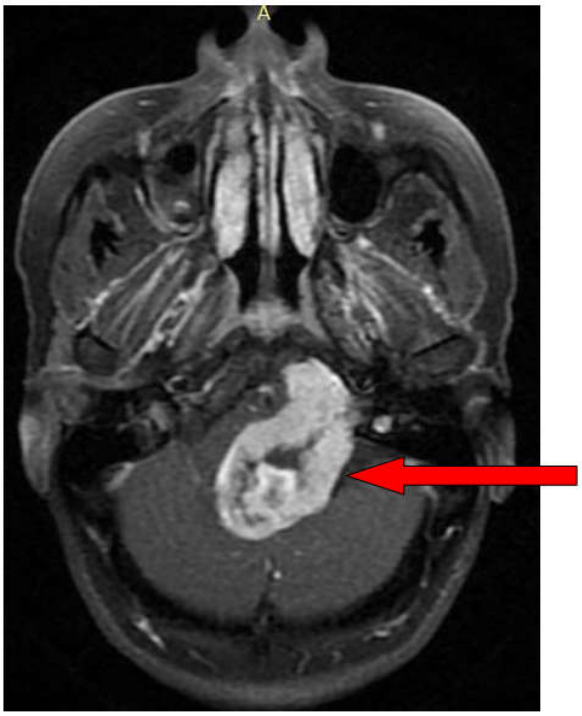
T2 W coronal image of the brain showing, medially extending posterior to pons & medulla and significantly compressing the pons & medulla , displacing them right anteriorly



FLAIR axial image of the brain



GRE axial image of the brain showing few hypointense foci of blooming were noted on gradient images suggestive of calcifications



T1 W post contrast axial image of the brain showing strong and heterogeneous peripheral enhancement on post contrast study with central non enhancing cystic / necrotic portion

It revealed high cellular density (Antoni A areas) and low cellular density areas embedded in a connective matrix (Antoni B areas). This, in addition to immunohistochemical positivity to S-100 protein, confirmed our diagnosis of a benign schwannoma .

DISCUSSION

Intracranial schwanomas are extremely rare tumours. Clinically they tend to be asymptomatic, until they are of a small size (<1–2 cm). When they increase in size (~2–3 cm) they cause pain and paraesthesias; at time they may grow upto 4–5 cm, as seen in this case (although rarely more than this). Regarding the location, most frequently, these lesions have an intracranial location, either developing from the cerebellopontine angle from the origin of the nerve or in the hypoglossal canal, and a secondary extra cranial extension, most commonly in the parapharyngeal space, presenting as neck mass.Early diagnosis and surgical intervention can preserve the function of nerve. Total excision of the tumor via an external approach is the best treatment for this condition.

Differential diagnosis: The differential diagnosis of the tumor involving hypoglossal canal includes:- chemodectoma, chordoma, meningioma, lymphoma, and metastatic tumors.

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