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Radiology Section

Isolated Hypoglossal Nerve Schwannoma: An Uncommon Presentation of Schwannoma

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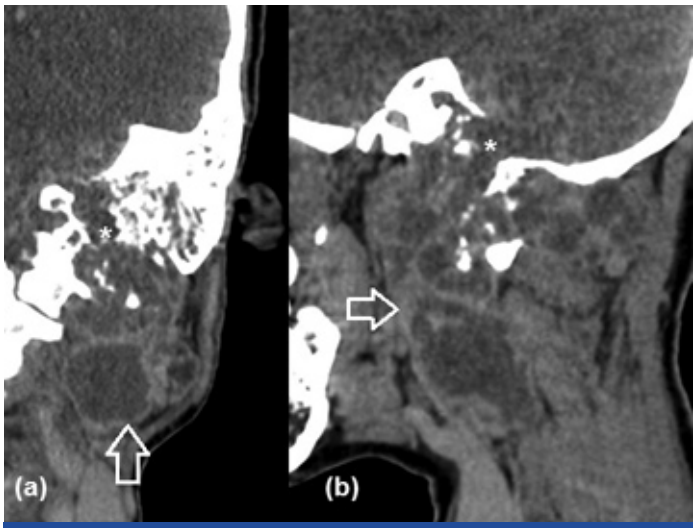
Keywords: CNS, Radio imaging

A 35-year-old lady presented with complains of difficulty in swallowing, mild hoarseness, deafness in left ear & swelling in left posterior triangle of neck for the 2 years. Patient had occasional occipital headache. Swelling was initially small in size then gradually increased. No complain of fever & past history of tuberculosis.

On examination approximately 6 x 6 cm sized well defined, lobulated swelling was present in left posterior triangle of neck. On palpation swelling showed both solid as well as cystic in consistency. It was immobile swelling. Overlying skin appeared normal. No evidence of any erythema or warmness. Tongue deviation to the left side when pushed out with fasciculation and fissuring in the left side of tongue was seen due to atrophy. Patient had dysarthria with distorted vowel & word flow without pauses. Patient had both sensory neural as well as conductive deafness. There was no evidence of any dysmorphic facial features. Other cranial nerves and systemic examination was normal.

On routine investigation Hemoglobin 10.1 gm%, total WBC count 8300 cell/cu.mm, ESR was within normal limit. Chest X ray was normal. CT scan revealed multiloculated hypodense cystic lesion present at the left cerebello-pontine angle region with peripheral enhancement. Lesion cause widening of the left hypoglossal canal with destruction of the adjacent clivus, left jugular foramen, left occipital bone & left occipital condyle [Table/Fig-1a,b]. Hypo density was seen in left sided of hemi-tongue suggestive of fatty infiltration [Table/Fig-2]. MRI scan revealed a large multiloculated 10 x 7.5 x 5.8 cm sized lesion present at the left cerebello-pontine angle region. Lesion was hypointense on T1 & hyperintese on STIR images with peripheral enhancement. The lesion was infiltrating left cerebellar hemisphere superiorly. Super medially, lesion was indenting on pons, medulla and middle cerebral peduncle. Inferiorly, lesion extends into the left parapharengeal space.



[Table/Fig-1]: (a) Coronal Post contrast Computed Tomography (CT) neck showing multiloculated hypodense peripheral enhancing cystic lesion (open white arrow) with widening of the left hypoglossal canal (white asterisk) with destruction of the left jugular foramen, left occipital bone & left occipital condyle. (b) Sagittal Post contrast CT scan showing multiloculated peripheral enhancing hypodense cystic lesion (open white arrow) at the left cerebello-pontine angle region widening of the left hypoglossal canal (white asterisk) with destruction of the adjacent clivus

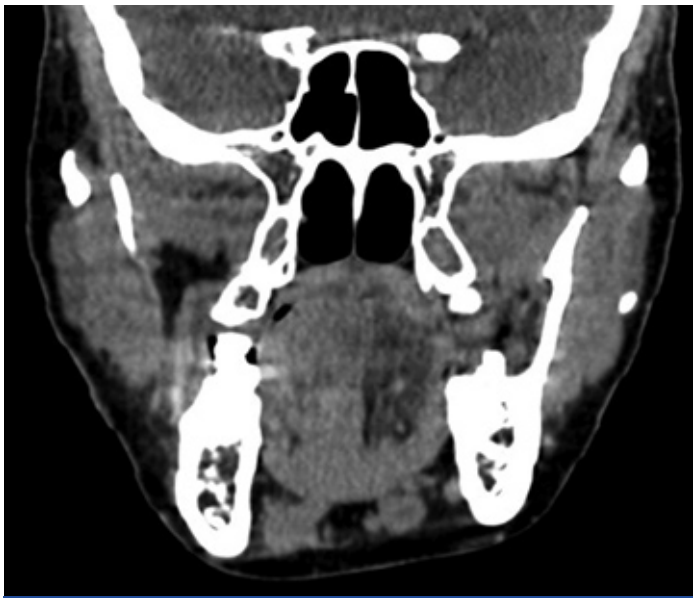
Anteriorly, lesion was reached up to the left pterygoid muscles. The lesion was extending into the left middle ear cavity, external auditory canal & internal auditory canal. Lesion causes widening of the left hypoglossal canal. Lesion was engulfing proximal part of left internal jugular vein and left sigmoid sinus. Cervical part of the left internal carotid artery was displaced anteriorly and left vertebral artery was displaced posteriorly. Lesion abuts left VII & VIII cranial nerve. High signal intensity was seen in left sided of hemi-tongue on T1 WI suggestive of fatty infiltration [Table/Fig-3,4].

On histopthological examination compact hyper cellular Antoni A areas and myxoid hypo cellular Antoni B areas were seen. Cells were narrow, elongate, and wavy with tapered ends interspersed with collagen fibers. Tumour cells showed ill defined cytoplasm, dense chromatin. Verocay bodies were seen in cellular areas [Table/Fig-5]. Large irregularly spaced vessels most prominent in Antoni B areas.

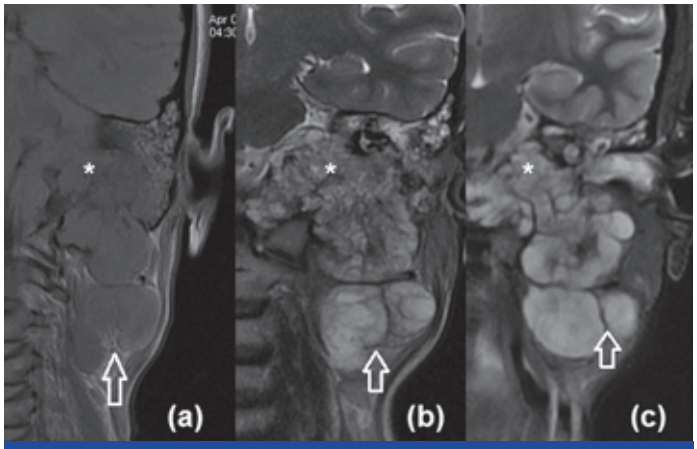
A clinical diagnosis of hypoglossal nerve Schwannoma was made based on the clinical, histopthological and imaging features. Then patient was referred to the higher centre for the further management. Where surgical excision of the tumour was done.

Schwannoma is benign, slow-growing neoplasm of the myelin-producing Schwann cells in the peripheral sensorimotor nervous system. Vestibular Schwannoma is the most common cranial nerve Schwannoma [1]. Pure motor nerve schwannomas is rare. Schwannomas of the hypoglossal nerve is accounting for only 5% of all nonacoustic intracranial schwannomas. Our case was classical involvement of the pure motor nerve.

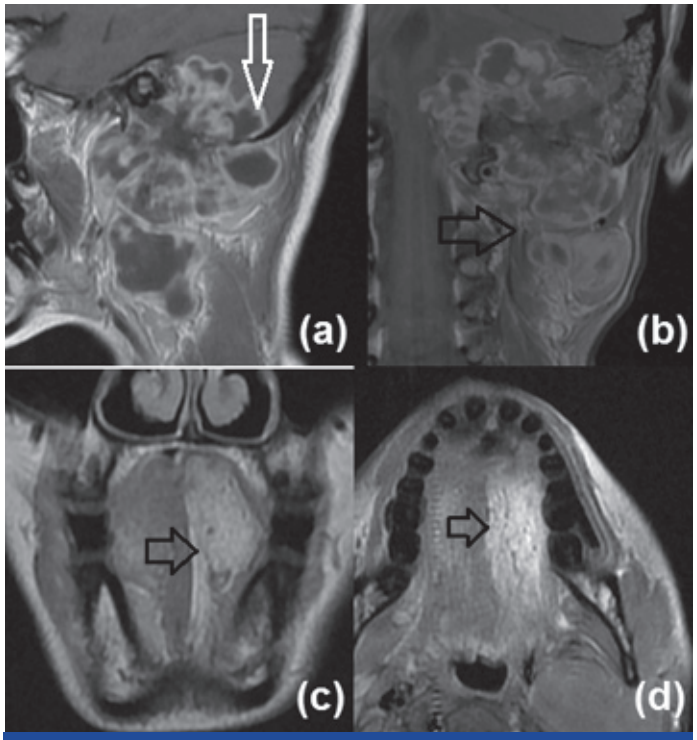
Cranial nerve schwannomas are usually isolated but some are associated with neurofibromatosis type 2 [2]. Hypoglossal schwannoma originates intra-cranially but it extends extra-cranially via the hypoglossal canal giving “dumbbell” appearance.



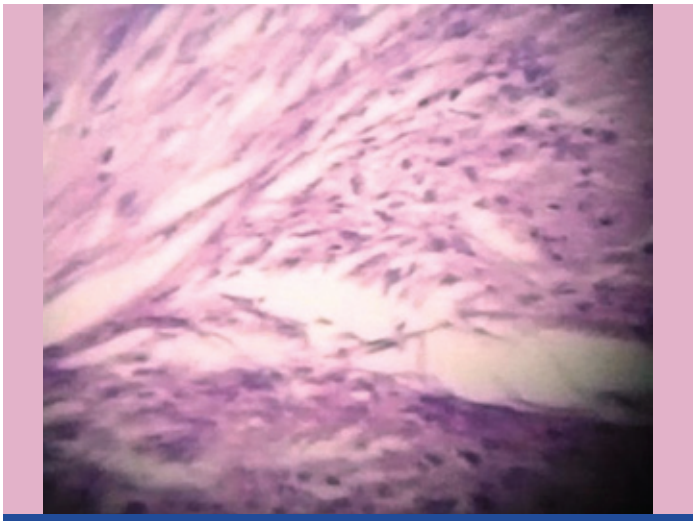
[Table/Fig-2]: Coronal postcontrats CT scan showing hypo density in left sided of hemi-tongue suggestive of fatty infiltration



[Table/Fig-3]: (a) Coronal T1 W MRI scan showing large multiloculated hypo to iso-intense lesion at the left cerebello-pontine angle region. (open white arrow) The lesion is infiltrating left cerebellar hemisphere superiorly. Inferiorly the lesion extends into the left parapharengeal space. Widening of the left hypoglossal canal (white asterisk) is seen. (b) Coronal T2 W MR scan showing high signal intensity lesion at the left cerebello-pontine angle region. (open white arrow) Super medially the lesion is indenting on pons, medulla and middle cerebral peduncle. Inferiorly the lesion extends into the left parapharengeal space. The lesion is extending into the left middle ear cavity, external auditory canal & internal auditory canal. Widening of left hypoglossal canal is present (white asterisk). (c) Coronal T2 W MR scan showing high signal intensity lesion at the left cerebello-pontine angle region (open white arrow) with widening of left hypoglossal canal (white asterisk)



[Table/Fig-4]: (a) Sagittal post contrast T1 W MRI scan showing a large multiloculated peripheral enhancing lesion (open white arrow) present at the left cerebello-pontine angle region. (b) Coronal postcontrast T1 W image showing peripheral enhancing multiloculated cystic lesion (open black arrow) in left cerebello-pontine angle region with widening of left hypoglossal canal. (c) Coronal T1 W MR image showing atrophy with high signal intensity in left hemi tongue (open black arrow) suggestive of fatty infiltration. (d) Axial T1 W MR image showing atrophy with high signal intensity in left hemi tongue (open black arrow) suggestive of fatty infiltration



[Table/Fig-5]: Haematoxylin and Eosin stain (40X) shows compact hyper cellular Antoni A areas and myxoid hypo cellular Antoni B areas. Cells were narrow, elongate, and wavy with tapered ends interspersed with collagen fibers. Tumour cells also show ill defined cytoplasm, dense chromatin

Hypoglossal nerve palsy is seen in 85.7% of patients with hypoglossal schwannomas [3]. Several reports have described hypoglossal schwannomas without hypoglossal nerve palsy. But the present case had features of the hypoglossal nerve palsy.

There is a no role exists for plain radiographic in evaluation of schwannomas. Non-enhanced CT scan; most schwannomas are iso-dense relative to brain parenchyma. Cystic changes or haemorrhage may see. On contrast-enhanced CT scans, it shows typically homogeneous enhancement or peripheral enhancement in cystic changes. Bone-window images can demonstrate expansion of the hypoglossal canal with destruction. Present case showed peripheral enhancement.

Schwannomas are typically iso-intense or hypointense relative to gray matter on T1-weighted images and slightly hypointense to CSF on T2-weighted images. On post contrast enhancement is typically homogeneous, but large tumour show areas of cystic degeneration.

Schwannoma is treated surgical through the suboccipital subtonsillar approach which was first described by Tatagiba et al., to treat a hypoglossal schwannoma [4]. Prognosis is good after surgical treatment. Schwannoma of the pure motor nerve is rare. The present case was classical involvement of the pure motor nerve. Imaging plays a vital role not in the diagnosis but also to know the extent of the hypoglossal nerve schwannoma so further management will plan accordingly.

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