ISSN No:-2456-2165

Case Series- A Radiological Spectrum of Cranial Nerve Schwannomas

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Abstract:- Schwannomas are common intracranial tumors that arise from the schwann cells of peripheral or cranial nerves. Understanding the anatomical location & types of presentation of these tumors assists in arriving at an appropriate clinical diagnosis. In this case series, we will discuss the various presentations of such tumors and the radiological anatomy of the concerning cranial nerve. The association between Schwannomas and neurofibromas is also briefly discussed.

I. INTRODUCTION

Schwannomas arise from the schwann cells of the nerve sheath of peripheral or cranial nerves [1]. The most commonly involved cranial nerve is the vestibulocochlear nerve followed by the trigeminal and facial nerves.

Schwannomas are also associated with a few conditions such as neurofibromatosis type 2 and schwannomatosis. Individuals with neurofibromatosis type 2 show a genetic predisposition to schwannomas, meningioma & ependymomas [2].

As it is a common cranial tumour, understanding their location and various types of presentations are important in clinical practice.

In our case series, we have presented an overview of schwannomas involving the various cranial nerves to provide a comprehensive spectrum. In addition to this, we have detailed the anatomy of cranial nerves to provide a holistic approach.

- *Imaging:*

As cranial nerves are not easily detected on spin echo sequences, in depth evaluation was made possible with usage of Steady-state free precession sequences (SSFP) /Constructive Interference in Steady State CISS - which employs heavily weighted T2W images.

SSFP is a gradient echo sequence with a short relaxation time & small flip angle.It has the ability to generate a strong signal in tissues with a high T2/T1 ratio – such as CSF & fat. REFERENCE SSFP makes the visualization of the cisternal segments of the cranial nerves due to high contrast between the fluid (CSF) & the nerves. The short acquisition time & high spatial resolution are also contribute to its value in evaluation of cranial nerves and cranial nerve lesions (tumors).

- *Imaging Features:*

Schwannomas are typically iso intense to hypointense on T1 weighted images, hyperintense of T2 weighted images & can show variable enhancement on post-gadolinium images.

The heterogeneity in patterns of enhancement due to presence of hemorrhage, necrosis & cystic degeneration to varying degrees– especially seen in larger lesions.

'Target sign' appearance has been described in literature [2] and is highly indicative of a schwannoma; however, it is rarely seen in practicality. It is more commonly seen in neurofibroma.

Clinical findings, such as denervation induced muscle atrophy or a sensory deficit in combination with imaging features helps in coming to a conclusive diagnosis of a schwannoma and the related cranial nerve.

ISSN No:-2456-2165

II. CASE SERIES WITH ASSOCIATED CRANIAL NERVE ANATOMY

Fig. 1: MRI axial section & pictorial representation of left facial & vestibulocochlear nerves

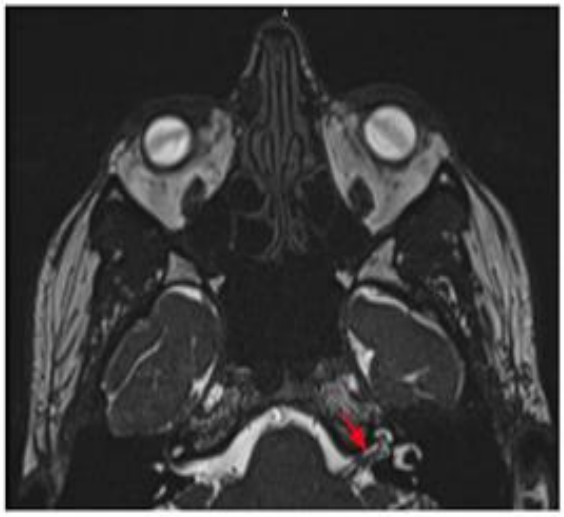

Fig. 1.1

Fig. 1.2

➢ *Facial Nerve & Vestibulocochlear Nerve*:

Facial nerve has motor & sensory functions. It is divided into a main nerve & an intermediate one.

The motor roots exit from the pontomedullary sulcus, traverses through the cerebellopontine cistern & enters the internal acoustic meatus. It then passes through the facial canal and gives rise to 3 segments:

- *Labyrinthine*
- *Tympanic*
- *Mastoid*

The vestibulocochlear nerve is formed by two nerves: the cochlear and the vestibular nerve.

Both nerves traverse the internal acoustic meatus, together with the facial nerve, and pass through the cerebellopontine cistern & the internal acoustic meatus.

The vestibulocochlear nerve enters the brainstem and reaches the dorsal and ventral cochlear nucleus and the four vestibular nuclei that are located in the pons [3].

➢ *CASE 1:*

A 23 year old male presented with complaints of right sided hearing loss since 1.5 years. Swaying gait and blurring of vision (both eyes) since 6 months.

Fig. 2

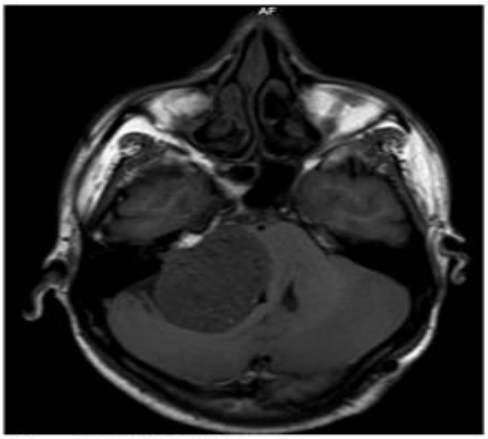

Fig. 2.1: T1W image

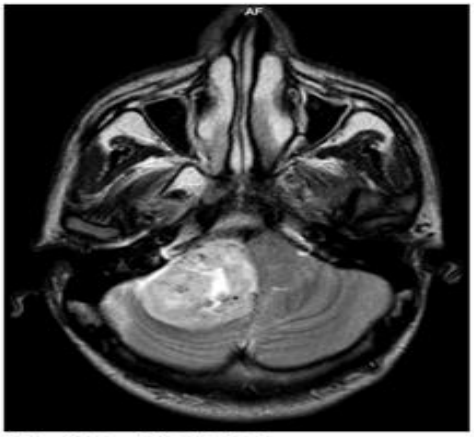

Fig 2.2: FLAIR image

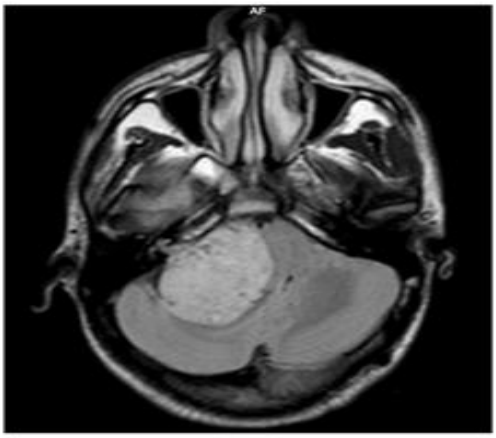

Fig 2.3: T2W image

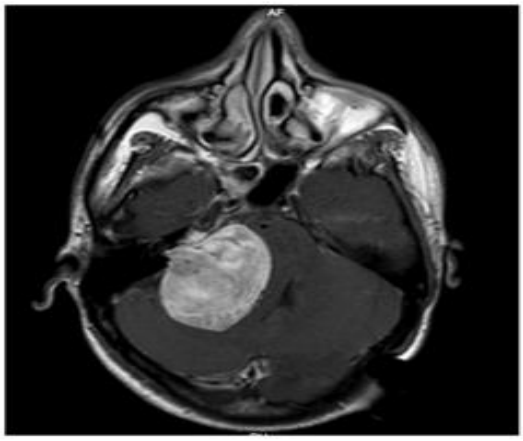

Fig 2.4: T1W post-GAD image

Imaging findings revealed a well-defined extra axial, heterogeneous, avidly enhancing lesion with areas of blooming & cystic areas within, in the right cerebellopontine angle extending into the IAC – s/o Acoustic Schwannoma.

- *Trigeminal Nerve:*

Fig. 3: MRI axial section & pictorial representation of left trigeminal nerve

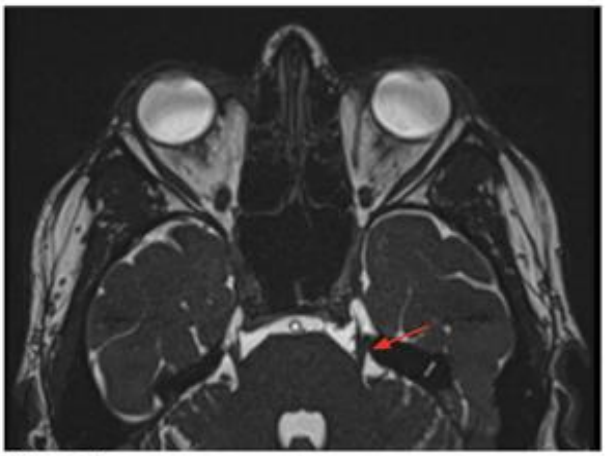

Fig. 3.1

Fig. 3.2

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The trigeminal nerve root emerges from the pons and goes through the prepontine cistern to enter the Meckel cave.

The ophthalmic and maxillary divisions of the trigeminal nerve course through the lateral wall of the cavernous sinus. The mandibular branch courses through the foramen ovale.

Trigeminal schwannomas can be unicompartmental or multicompartmental.

Due to the close proximity of the nerves that are present in this region, accurate assessment of the origin of the Schwannoma becomes difficult.[4]

- *Case 2:*

Fig. 4

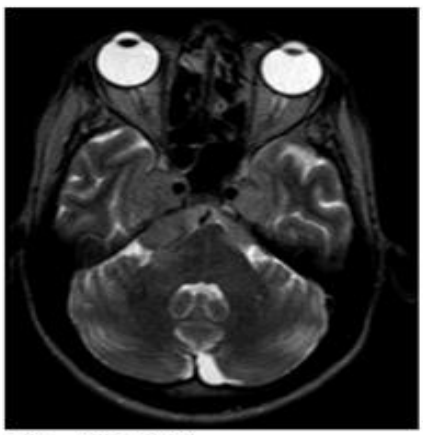

Fig. 4.1: T2W

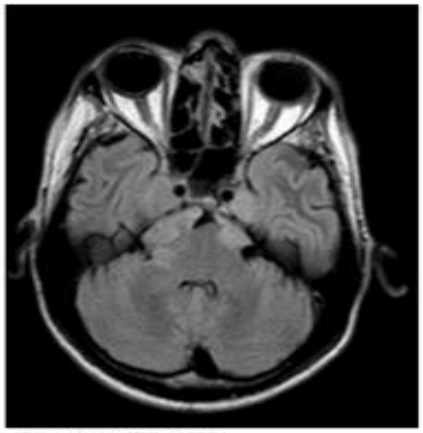

Fig. 4.2: FLAIR

Fig. 4.3:T1W POST GAD

Imaging findings of a patient showing two extra axial well defined lesions seen in the Meckel's cave extending into the prepontine and cerebellopontine cisterns bilaterally– S/o bilateral trigeminal schwannomas.

- *Case 3:*

A 21 year old male patient presented with complaints of multiple swelling on the body since birth and complaints of progressive loss of vision of the right eye since 3 years.

Fig. 5

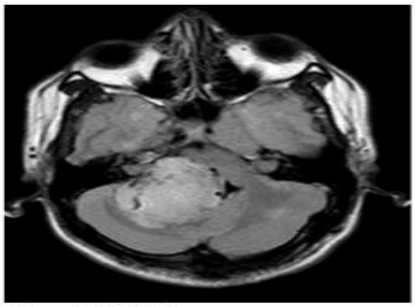

Fig. 5.1:T1W

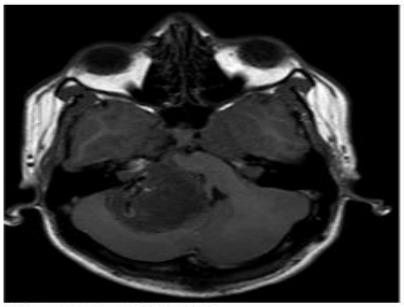

Fig. 5.2: FLAIR

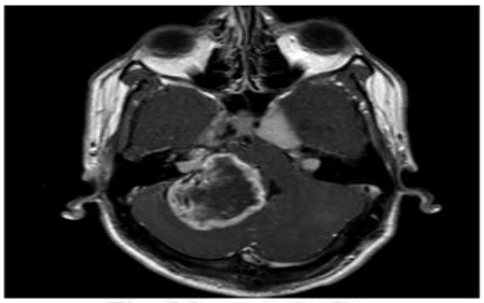

Fig. 5.3: post-GAD

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Imaging findings reveal multiple Schwannomas seen in bilateral Meckel's cave & both cerebellopontine angles involving the VII & VIII nerve complexes (largest on right cerebellopontine region) – likely Neurofibromatosis type II.

- *Case 4:*

A peculiar presentation of a cranial nerve Schwannoma occurred in a patient with complaints of a painless swelling in the region of the right eye.

Fig. 6

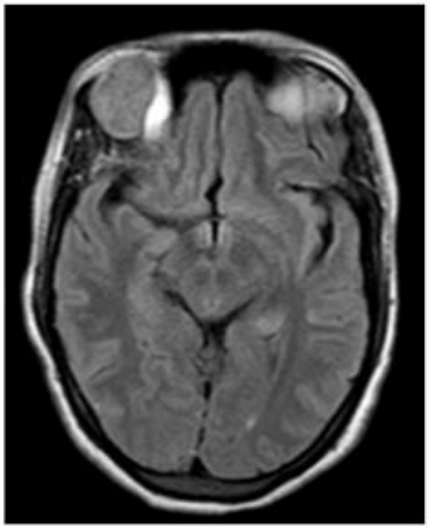

Fig. 6.1:T2W axial

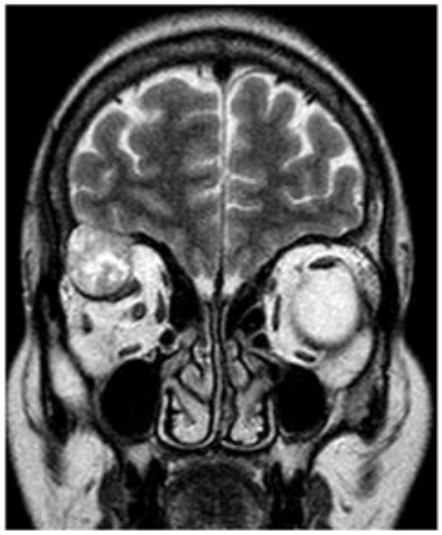

Fig. 6.2: T2W Coronal

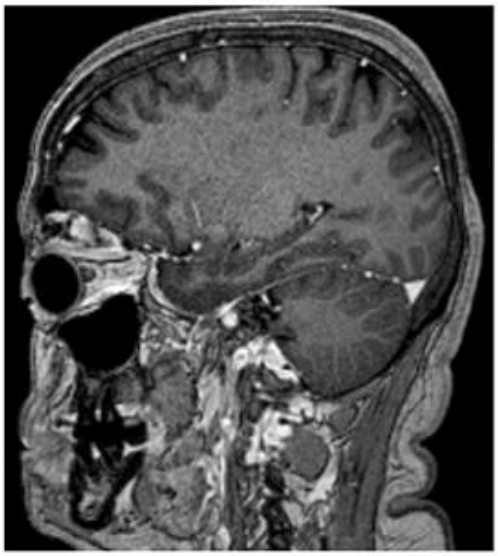

Fig. 6.3: post-GAD Sagittal

Well encapsulated right intraorbital, extraconal, lobulated solid lesion with cystic areas – likely arising from a branch of V1 segment of the trigeminal nerve.

Histopathology of this case revealed it to be a Schwannoma. It has been postulated that schwannomas do not commonly arise from cranial nerve II as it is myelinated by oligodendrocytes rather than Schwann cells.

➢ *Trochlear Nerve:*

Fig. 7: MRI axial section & pictorial representation of right trochlear nerve:

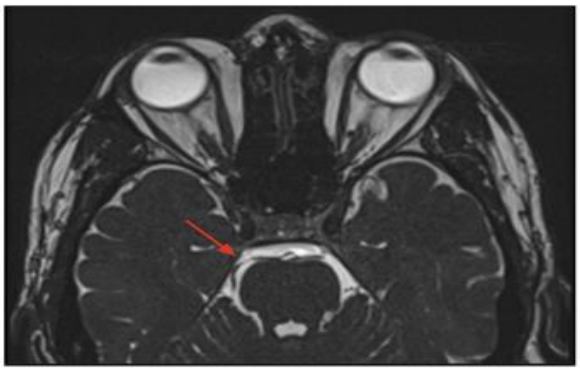

Fig. 7.1

Fig. 7.2

The root of the trochlear nerve arises from the pons, courses over the cerebellar peduncle & along the tentorium to enter the cavernous sinus.

Then, it traverses through the Superior orbital fissure.

➢ *Case 4:*

A 39 year old patient presented with complaints of swelling in the right side of the cheek associated with blurring of vision & superior oblique muscle weakness.

Fig. 8

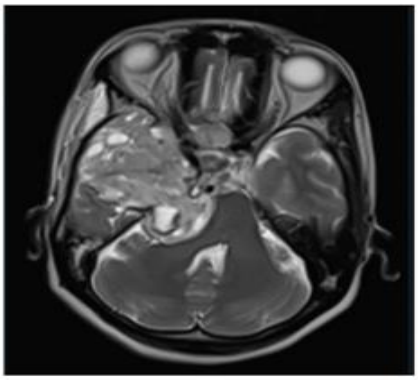

Fig. 8.1: T2W axial

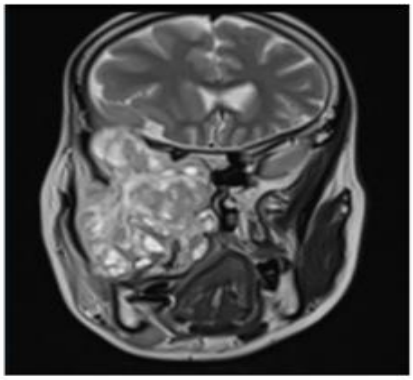

Fig. 8.2: T2W coronal

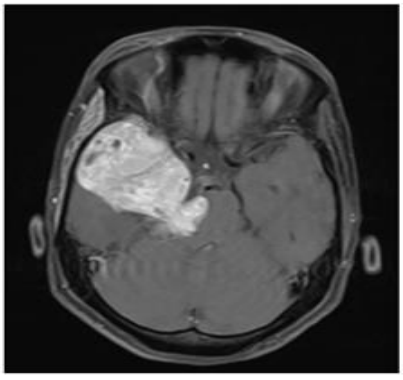

Fig. 8.3: post-GAD

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Imaging was performed and shows a fairly well-defined, lobulated lesion noted in the right cerebellopontine angle region seen causing mass effect on the pons and extending into the middle cranial fossa & further into the infratemporal fossa– Features suggestive of multi-compartmental schwannoma – likely arising from the Trochlear nerve.

- *Vagus Nerve:*

Fig. 9: MRI axial section & pictorial representation of right vagus nerve:

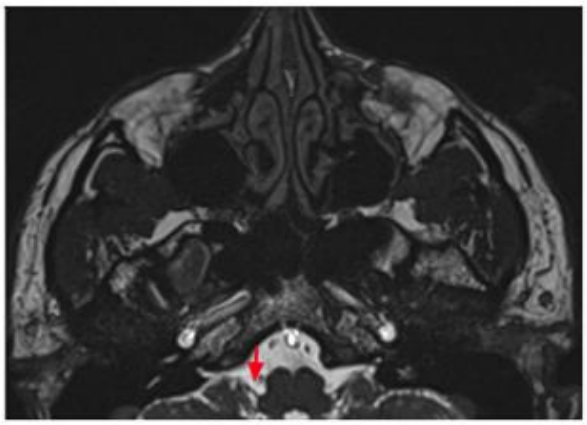

Fig. 9.1

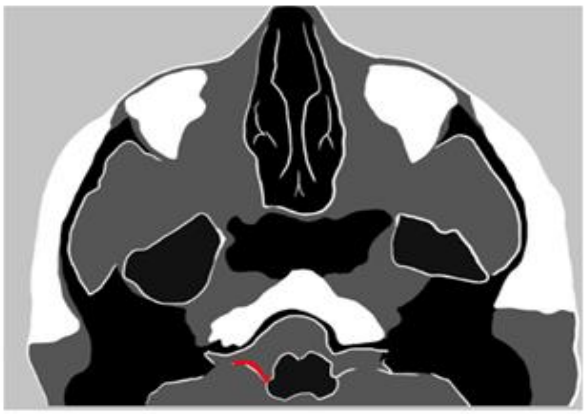

Fig. 9.2

The vagus nerve emerges from the lateral medulla, and the dorsal nucleus of the vagus.

It enters the lateral cerebellomedullary cistern and exits the skull through the jugular foramen between the glossopharyngeal nerve and the accessory nerve.

It forms a superior and inferior ganglion inferior to the jugular foramen.

It then descends vertically in the retrostyloid space together with the carotid artery & gives off the recurrent laryngeal nerve in the mediastinum.

Then, the vagus nerve enters the abdominal cavity through the esophageal hiatus.

- *Case 5:*

A 43 year old female presented with complaints of dizziness & headache on and off.

Fig. 10

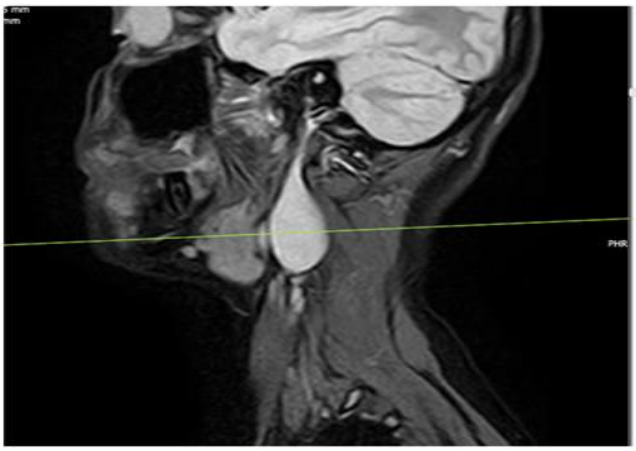

Fig. 10.1: T2W sagittal

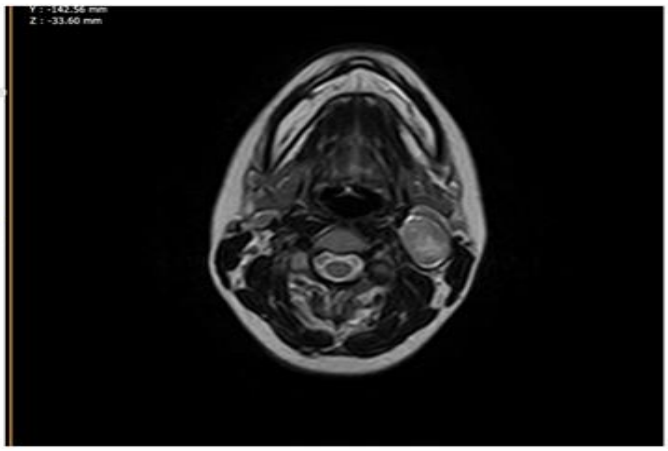

Fig. 10.2: T2W axial

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Imaging findings shows a well defined lobulated lesion in the left anterior paracervical neck space , posterior to carotid vessels and IJV.

Superiorly, the lesion shows focal, linear extension with suspicious extension upto jugular foramen at the level of cranial exiting of likely the left vagus nerve.

-likely vagus/sympathetic chain schwannoma

- *Hypoglossal Nerve:*

The hypoglossal nerve emerges from the medulla oblongata as a series of rootlets extending from the ventrolateral sulcus of the medulla into the lateral cerebellomedullary cistern.

It is near the vertebral artery and the posterior inferior cerebellar artery (PICA).

The nerve exits the skull through the hypoglossal canal.

- *Case 6:*

A 54 year old male patient presented with complaints of tongue atrophy, dysphagia & slurring of speech.

Fig.11

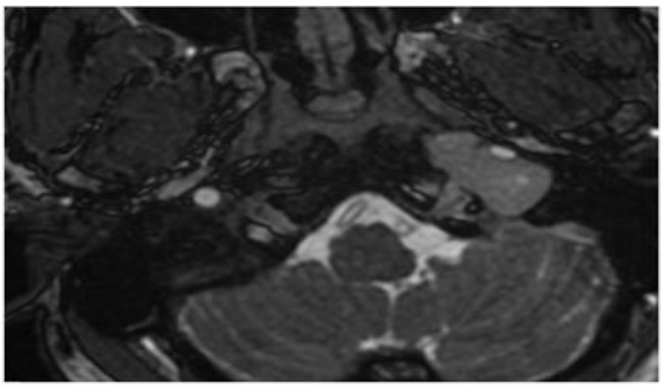


Fig. 11.1: T2W axial

Imaging findings revealed a well-defined homogenous lobulated extra-axial lesion noted in the left hypoglossal canal extending into left upper para and nasopharyngeal soft tissues with extensions as described- s/o neoplastic etiology- likely schwannoma.

- *Accessory Nerve:*

The accessory nerve has both cranial and spinal roots.

Cranial roots emerge into the lateral cerebellomedullary cistern, while spinal roots emerge from upper cervical segment of the spinal cord (from C0 to C5) and pass superiorly through the foramen magnum into the cisterna magna.

Within the jugular foramen, cranial roots mix with vagus nerve at the level of the superior vagal ganglion, while spinal roots descend laterally to the internal carotid artery and internal jugular vein.

- *Olfactory, Optic & Oculomotor Nerves:*

The olfactory nerve conveys stimuli from the nasal cavity to the brain. It is not surrounded by Schwann cells & instead has olfactory receptors and olfactory filiae that represent its axons.

The enter the anterior cranial fossa through the cribiform plate, terminating at the olfactory bulb.

Schwannomas are infrequently associated with the olfactory nerve and when they do occur, they generally present as a midline lesion & are extra-axial in origin. [5]

ISSN No:-2456-2165

The origin of these schwannomas has been debated. A developmental theory postulates that they arise from aberrant Schwann cells within the CNS.

Other theories suggest a transformation of mesenchymal pial cells to Schwann cells.

Fig. 12:

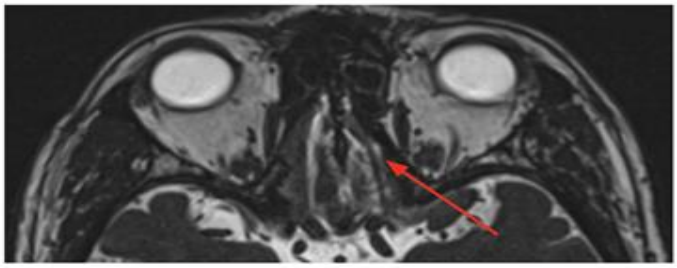


Fig. 12.1: Left olfactory nerve

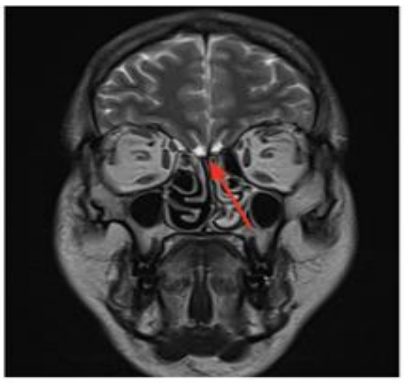


Fig. 12.2: Olfactory bulb

➢ *Optic:*

The optic nerve is considered to be an extension of the CNS and is lined by oligodendrocytes.

From the posterior aspect of the globe, the nerve is divided into 4 segments:

- *Intraocular*
- *Intraorbital*
- *Intracanalicular*
- *prechiasmatic*

Both nerves then join together and form the optic chiasma and then traverse to the lateral geniculate nuclei. From there, they reach the primary visual cortex which is located in the occipital lobe (Brodmann area).

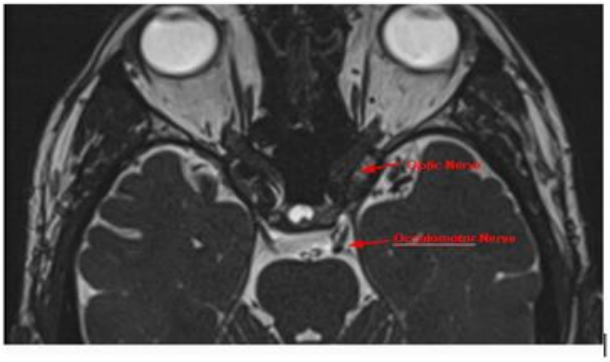
➢ *Occulomotor:*

The somatic motor functions of the oculomotor nerve originate from teh superior colliculi of the midbrain & the parasympathetic fibres arise from the Edinger-Westphal nucleus.

The nerve root originates within the interpeduncular cistern and traverses superior to the posterior cerebral artery.

The nerve enters the cavernous sinus and passes through the superior orbital fissure (SOF) to reach the orbit.

Fig. 13: MRI axial section of left optic & occulomotor nerves:



➢ *Glossopharyngeal Nerve:*

Fig. 14: MRI axial section & pictorial representation of left glossopharyngeal nerve:

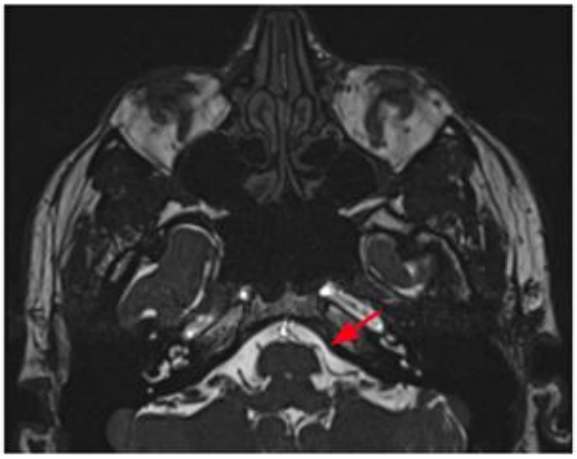


Fig. 14.1

Fig. 14.2

The glossopharyngeal nerve emerges from the lateral medulla & into the lateral cerebellomedullary cistern. It exits the skull through the jugular foramen and enters the carotid space. Its lingual branch ends in the posterior sublingual space.

Other nervous branches of the extracranial segment of IX nerve are pharyngeal, sinus nerve, stylopharyngeus, and the tympanic branch.

➢ *Abducens Nerve:*

Fig. 15: MRI axial section & pictorial representation of left abducens nerve:

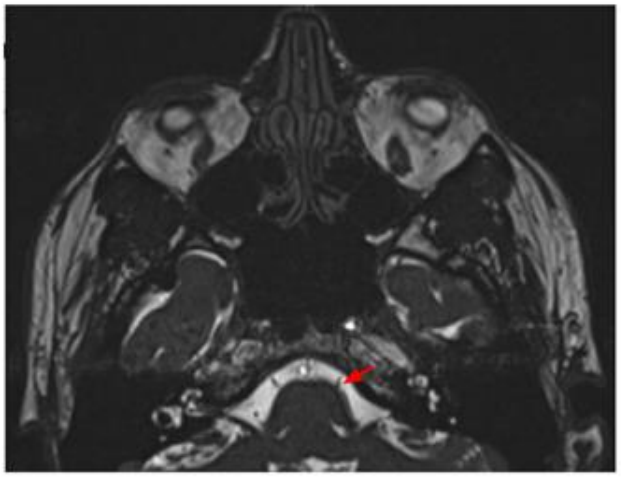


Fig. 15.1

Fig. 15.2

Abducens nerve emerges from its nucleus beneath the floor of the fourth ventricle, traverses the prepontine cistern, descends posterior to the clivus and enters the cavernous sinus & the superior orbital foramen.

It is uncommon to find a schwannoma associated with this nerve with very few recorded cases. [6]

III. CONCLUSION

Schwannomas are common intra-cranial tumors that arise from schwann cells of peripheral or cranial nerves. Though they tend to present with classical features, few of these lesions may show an atypical presentation or may be seen in uncommon locations. This case series encompasses typical and atypical presentations of cranial nerves and elaborates on the radiological anatomy of the associated cranial nerves.

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