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Case Report

Rare case of Multi-segment facial nerve schwannoma (MFNS) - A case report

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ABSTRACT

Multi-segment Facial Nerve Schwannoma (MFNS) is a very rare tumor, benign in nature and can arise anywhere along the course of the facial nerve from its origin in the cerebello-pontine angle to its extra cranial ramification in the parotid space of the extra cranial head and neck. It mimic like vestibular schwannoma and other cranial nerve schwannoma. The most common clinical presentation is facial palsy and depending upon the segment of the facial nerve involved. We report a rare case of multi-segment facial nerve schwannoma extending from cerebello-pontine angle to the parotid gland, presented with facial palsy, hearing loss, difficulty in swallowing and difficulty in walking. Radiological investigations was done and confirmed the diagnosis MFNS. Patient underwent near total excision of the tumor by combined approach and patient was improved clinically after the surgery.

1. Introduction

Multi-segment Facial Nerve Schwannoma (MFNS) is a very rare slow growing tumor arising from the facial nerve. Commonly most of the Facial nerve schwannomas are intratemporal, solitary, unilateral and sporadic in nature, with 9% of cases arising from the intraparotid portion.[1] But in Multi-segment Facial nerve schwannoma (MFNS), it involves all the segments of facial nerve, extending from cerebello-pontine angle intra cranially to parotid region extra cranially, which is very rare and not reported in literature so far. The signs and symptoms of the MFNS depends upon the mass effect over the adjacent cranial nerves, brain stem and other structures. Most common clinical presentation is facial palsy, hearing loss, lower cranial nerve palsy, brain stem dysfunction, cerebellar dysfunction and palpable parotid mass.[2] Facial nerve is very resistant to compression and often facial nerve paralysis or facial nerve weakness is not present in many FNS.[3] Magnetic resonance imaging (MRI) with contrast and Computed tomography (CT) of temporal bone are the key to diagnosis and for treatment plan. Combined approach including Subocciptal craniotomy, Translabyrinthine approach and Neck dissection are the key surgical procedures for the excision of MFNS in patients with non serviceable hearing.

2. Case Report

A sixty years old lady presented with progressive right sided facial weakness, reduced hearing, difficulty in swallowing and difficulty in walking for the past 3 years. On clinical examination patient had Profound Sensory neural hearing loss (SNHL) on right side, facial nerve palsy with House and brackmann grade IV on right side, reduced facial sensation on right side, lower cranial nerve (9,10,11,12) weakness, spastic weakness in all 4 limbs , cerebellar signs on right side, palpable mass in right side of the neck, parotid region and also seen in the right External auditory canal (Figure 1). MRI brain with contrast shows a well-defined smooth

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marginated solid cystic mass lesion in the right cerebello-pontine angle, extending through right internal auditory meatus, facial nerve canal, external auditory canal and extracranially into the infra-mastoid region and into right parotid salivary gland. Lesion appears heterointense in T2 with heterogenous post contrast enhancement. Intra cranial portion of the lesion measures 5.1cm*3.8cm*4.8cm. Extra cranial portion of the lesion measures 2.4cm*2.7cm*2.8cm. The lesion causes mass effect, compression of right cerebellar hemisphere, right cerebellar peduncle, brainstem, cervical-medullary junction, cerebral aqueduct and fourth ventricle outlet with displacement to right side and causing obstructive hydrocephalus involving bilateral lateral and third ventricle (Figure 2). On CT screening, the lesion causes widening of right internal auditory canal with extension into middle ear and mastoid causing destruction of bony structures including middle ear ossicles, reported as Right Facial nerve schwannoma with both intracranial and extra cranial component causing mass effect and obstructive hydrocephalus (Figure 3). Pre operative work up was done and surgical fitness obtained from anesthesia department. Surgery was planned, by combined approach including suboccipital craniotomy, translabyrinthine (non-serviceable hearing) and neck dissection. Subtotal excision of the tumor was done as the tumor was very adherent to the lower cranial nerves which was left behind. Brainstem decompression was adequate. Post operatively patient improved of symptoms like improved facial sensation, able to swallow and able to walk gradually over the period of two weeks. Histopathology reported as Facial nerve schwannoma.

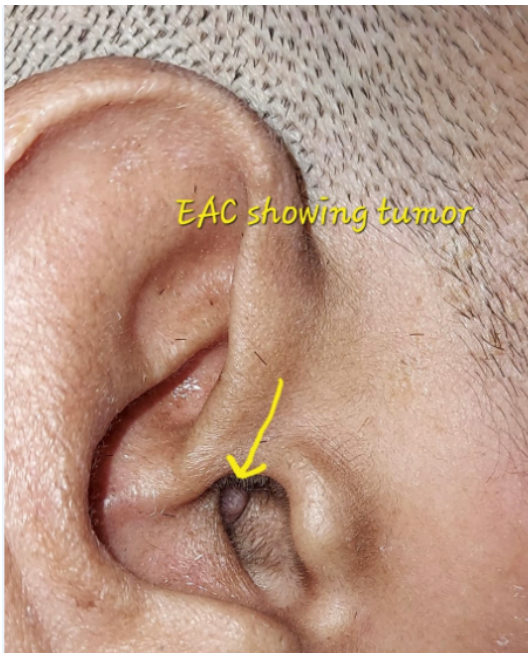

Fig. 1:

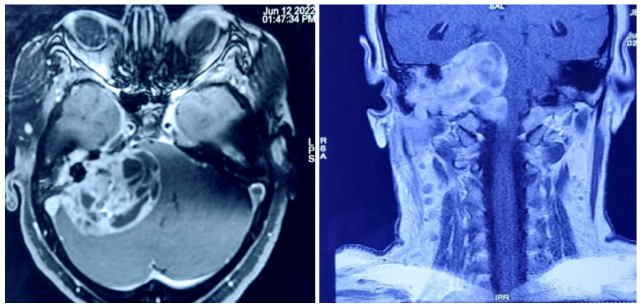

Fig. 2:

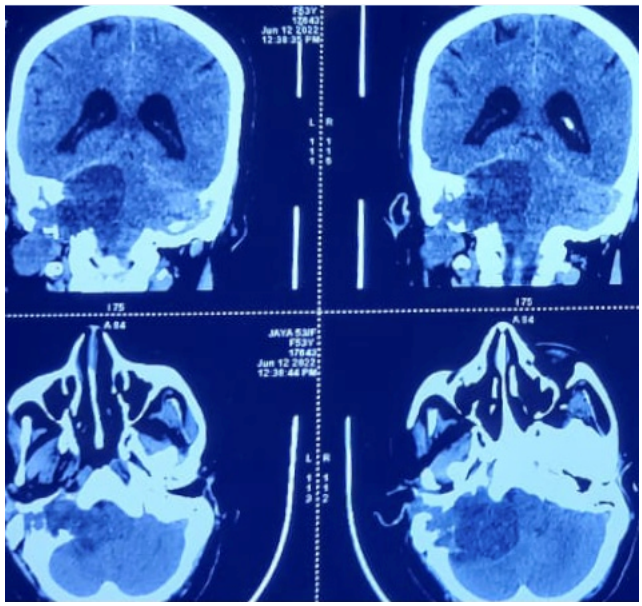

Fig. 3:

3. Discussion

Facial nerve has a long course extending from the glial-schwannoma cell junction at cerebello-pontine angle to the peripheral branches in the face.[4] FNS arises anywhere from the above course, most commonly from intra temporal compartment. The range of symptoms makes FNS difficult to diagnose preoperatively without radiological examination. FNS usually expands along the path of least resistant and remain asymptomatic until large without clinically presenting with 7th nerve dysfunction.[5] The most common clinical presentation is varying degree of facial paresis, conductive hearing loss, tinnitus, hemifacial spasm, otalgia, reduced lacrimation, sensory neural hearing loss and ear canal mass.[6] Early imaging is warranted in patient with progressive facial paresis with swelling in parotid region. FNS regardless of tumor locality, the basic imaging characteristic of schwannoma are the same. MRI image will show hypo to isointense in T1 WI, heterogenous hyperintense in

T2 WI with contrast enhancement. Cystic changes may seen in larger schwannoma.[7] Schwannoma exhibit "Target sign" in which hyperintensity in periphery due to loose myxomatosis Antoni B regions and hypointensity in centre due to compactly packed cellular Antoni A regions.[8] CT temporal bone to look for bony anatomy, bony erosions and breaking into surrounding mastoid air cells.[9] Ultrasonography of the neck and parotid region to be done to rule out other differential diagnosis like pleomorphic adenoma and adenoid cystic carcinoma. Other diagnostic work up included audiometry (audiological test) and auditory brain stem evoked response audiometry (ABER).[10] Surgical excision is the primary mode of treatment. Surgical approach for Multi-segment FNS depends not only the size and anatomical location, but also patients hearing status.[11] In our case, as patient with non-serviceable hearing we did surgical excision by combined approach including suboccipital craniotomy, translabyrinthine and neck dissection. Surgical excision of FNS in individual without delay, for patients with progressive facial palsy, large CP angle component compressing brain stem and hydrocephalus and tumor invading inner ear. Post operatively facial nerve rehabilitative measures like eyelid implant, lateral tarsorraphy depending upon the severity of the facial nerve palsy.[12]

4. Conclusion

Multi-segent Facial Nerve Schwannoma (MFNS) is a very rare tumor, involving all segments of facial nerve from cerebello-pontine angle to parotid gland, not yet reported in literature so far, our case being the first Multi-segment FNS extending from CP angle region to the parotid region. FNS usually mimics vestibular and other cranial nerve schwannoma. Early diagnosis and treatment leads to avoiding significant morbidity to the patients. Proper history, clinical examination and radiological investigations clinch in diagnosis. Pre op diagnosis of FNS is difficult to establish as rarity of tumour and non-specific clinical presentation. Multisegment FNS requires combined approach for achieving good surgical excision and proper rehabilitation to be provided to the patient post operatively to improve the quality of life.

5. Conflicts of Interest

None.

6. Source of Funding

None.

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