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A RARE CASE OF INTRATEMPORAL FACIAL NERVE SCHWANNOMA: CASE REPORT

Radiology

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ABSTRACT

Schwannomas, are benign tumours arising from the nerve sheath. They are uncommon in the facial nerve and account for less than 1% of tumours of temporal bone. (3) They can arise anywhere along the facial nerve from its origin in the cerebellopontine angle to its extra cranial ramifications in the parotid space. (1,2) Most commonly they involve the intra-temporal course of the nerve, with propensity for the geniculate ganglion.

KEYWORDS

CASE REPORT:

A 24 years old patient presented in the out-patient of oto-rhinolaryngology department in a tertiary care hospital of Navi Mumbai, with complaints of right sided facial weakness since two months, drooling of saliva from left side of the mouth and drooping of angle of mouth to the right side. Patient was unable to close the left eye completely and loss of frontal crease since two months.

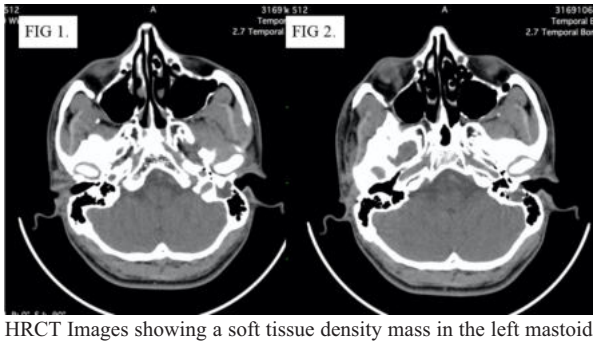
There was history of left sided ear ache preceding the facial weakness, which was intermittent and spontaneous radiating to the left side of the neck.

High resolution computed tomography of temporal bone was done and revealed a soft tissue density mass involving the mastoid air cells causing erosive bony changes, with a defect in the posterior wall of the external auditory canal causing protrusion of these contents into the bony external auditory canal. The semicircular canals and inner ear appeared normal. Further evaluation with MRI was suggested.

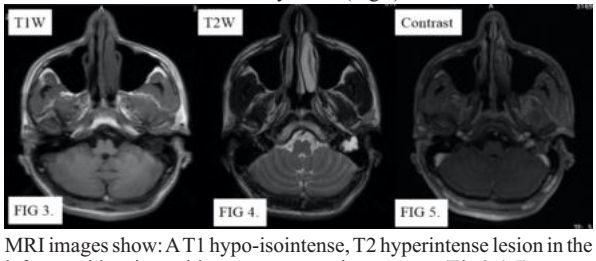
On MRI, the lesion appeared as a well defined enhancing altered signal intensity lesion involving the left mastoid region along the mastoid segment of the left facial nerve, which was iso-hypointense on T1W images and heterogeneously hyperintense on T2W and FLAIR images.

Radiological differentials were paraganglioma and facial neuroma.

The patient was operated, tumour excision with reconstruction of the facial nerve was done. Histopathology revealed the final diagnosis to be facial nerve schwannoma.



HRCT Images showing a soft tissue density mass in the left mastoid region (Fig1) with a small defect in the posterior wall and protrusion of contents into the external auditory canal. (Fig 2)



MRI images show: A T1 hypo-isointense, T2 hyperintense lesion in the left mastoid region, with post contrast enhancement. (Fig 3,4,5)

DISCUSSION:

SEGMENTAL ANATOMY OF THE FACIAL NERVE:

The course of the facial nerve is divided into six segments and two genua:

1. Intracranial, the nerve arises from the pons, travelling through the cerebellopontine angle to the internal acoustic meatus.
2. Meatal/canalicular: lies in the anterior superior quadrant of the internal auditory canal.
3. Labyrinthine segment, As the nerve passes the internal acoustic meatus it enters the fallopian canal passing anterolaterally between the cochlea and vestibule and bends at the geniculate ganglion (Genu 1) and gives off two branches.
4. Tympanic segment. Beyond the genu the nerve is now called the tympanic segment of facial nerve, lying immediately beneath the lateral semicircular canal in the medial wall of the middle ear cavity. Just distal to the pyramidal eminence the nerve makes its second turn (Genu 2) passing vertically downwards.
5. Intramastoid segment. The vertical portion of the nerve is now called the mastoid segment which extends upto the stylomastoid foramen.
6. Extratemporal segment. The nerve exists through the stylomastoid foramen and lies between the superficial and deep lobe of the parotid gland, where it gives of its terminal branches to form the parotid plexus. (4)

Schwannomas are tumours of ectodermal origin, arising from the Schwann cells that form the nerve sheath.

Facial nerve schwannomas are rare, slow growing tumours. Typically they are solitary, unilateral and sporadic in nature. (3)

Bilateral involvement may occur as a part of neurofibromatosis -1 spectrum (6)

They can occur at any age but more frequently occur in the fifth and sixth decade with no gender predilection.

Facial nerve schwannomas may involve multiple segments simultaneously or show skip lesions. (7) These are mainly eccentric in location, with displacement and splaying of the nerve fibres. They usually cause widening of the facial canal when small in size and if large can cause erosive changes of the adjacent mastoid. The mastoid segment of facial canal is surrounded by thin fragile bony septations which separate it from the mastoid bone itself. Therefore, it appears as a tumour with irregular margins with erosive changes involving the adjacent bone breaking into it, better visualised on CT images. (5) Malignant transformation of schwannomas is extremely rare (8)

A wide variety of symptoms ranging from facial nerve palsy, hearing loss, otalgia , to vague symptoms like imbalance, ataxia, vertigo, headache, make the clinical diagnosis challenging. Facial nerve dysfunction is regarded to be associated with facial nerve neuromas in only 5% of cases (3) and other non specific symptoms are more common. (1)

Normal facial nerve function was however reported in 27% patients (9). In rare cases where the mass is seen eroding the posterior bony wall of the external auditory canal, 26%-50% patients had normal facial nerve function. (10)

Although symptoms like, facial nerve paralysis and hearing loss can signal the presence of facial nerve schwannoma, the range of symptoms associated with facial nerve schwannoma, make it difficult to diagnose it preoperatively. Therefore, adequate radiological investigation and histology there after is essential for to make the correct diagnosis.

TREATMENT:

Therapeutic options of these tumours depend on the facial function, tumour size and symptoms of the patients.

Microsurgical excision with facial nerve repair is done in most cases. However, conservative techniques with involve the preservation of facial nerve are being adopted as treatment option, like watchful waiting, fallopian canal decompression and stereotactic radiosurgery. (11,12)

Sterotactic radiosurgery allows preservation of facial nerve function in most cases, by avoiding surgical excision and also causes growth arrest of the tumour. This could be very useful in cases of small tumours without any significant bony involvement and mild symptoms.

However if patient presents with worsening clinical symptoms and tumour causing erosion of adjacent bone or compressive changes on brain stem in case of cerebellopontine tumours, more aggressive surgical options need to be considered.

CONCLUSION:

For diagnosis of a facial nerve schwannoma, a combined approach of clinical, radiological and histopathological correlation is essential. Extensive evaluation of the facial nerve on HRCT temporal bone studies should always be done where erosive bony changes of mastoid air cells and facial canal are seen. MRI and contrast studies help to differentiate it from benign and more common lesions causing erosive bony changes like cholesteatoma and chronic infective changes, which does not show enhancement on post contrast studies.

The absence of clinical symptoms of facial nerve paralysis does not rule out the possibility of facial nerve schwannoma owing to its wide range of symptoms, Therefore, highlighting the importance of radiological examination.

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