Facial nerve schwannoma

Balasubramanian Thiagarajan (drtbalu)

Introduction:

Facial nerve schwannomas are very rare benign tumors originating along the course of the facial nerve. On imaging these masses present as enhancing CP angle mass making it difficult to distinguish from vestibular schwannoma and meningioma. These lesions begin from the schwann cell lining of the facial nerve. These tumors are ectodermal in origin. It is encapsulated. The mean growth rate of these tumors is about 1.4 mm / year.

The geniculate ganglion (facial nerve ganglion) is located in the temporal bone and contains cell bodies associated with facial nerve specialized taste and general somatic sensory fibers. In the absence of pathology, the geniculate ganglion is usually not visible under IV gadolinium based enhancement. In the presence of schwannoma the ganglion becomes visible due to enhancement.

Location:

Facial nerve schwannomas can involve any portion of the facial nerve. Commonly it is seen in the perigeniculate area and the tympanic segment. Skip lesions have also been reported causing involvement of multiple areas of facial nerve simultaneously.

Epidemiology:

Incidence of intratemporal facial schwannoma is about 0.8%. Peak incidence is between 3rd and 6th decades of life. It does not show any sexual preponderance. These tumors can arise from any segment of facial nerve from the CP angle to the extratemporal peripheral portion. Hence the lesion can be located intracranially, intratemporally or extratemporally.

Clinical features:

Features differ on the basis of the site of origin and also the variability of structures getting involved. Some of the presenting features include:
1. Facial nerve paralysis Most common symptom and is progressive in nature.

2. Hearing loss may be conductive, cochlear or retrocochlear.

3. Facial nerve pain

4. Hemifacial spasm

5. Decreased lacrimation

Facial palsy could be sudden or progressive in nature. Facial palsy is often preceded by bouts of facial twitching which is a very common complaint in these patients. Normal facial nerve function has been reported in nearly a quarter of these patients. Schwannomas can arise from any portion of the facial nerve from the CP angle to the neuromuscular junction. There is of course a predilection for the involvement of geniculate ganglion. From here the lesion can extend to involve the tympanic segment or labyrinthine portion of the facial nerve.

Uncommonly the facial nerve can involve middle cranial fossa by direct spread upwards through the temporal bone. If the schwannoma originates from the mastoid segment of the facial nerve then it could present as aural polyp coming from the posterior wall of external auditory canal. Ear discharge may also be present confusing the issue.

Facial nerve schwannomas have also been discovered in patients presenting with minor facial nerve symptoms, these symptoms could very well have been dismissed as due to Bell's palsy. T1 weighted MR imaging commonly shows intratemporal facial nerve enhancement of all or part of the facial nerve along with a tuft of enhancement of the fundus of the internal acoustic meatus in patients with Bell's palsy.

Imaging:

CT and contrast enhanced MRI should be performed. These two imaging modalities compliment each other in arriving at a final diagnosis. The presence of the mass and its precise extension can be accurately seen in imaging. CT bone window is considered to be superior to MRI. Enlargement of facial nerve should be suggestive of the diagnosis.

The normal size of the facial nerve canal is also used to distinguish normal facial nerve enhancement during Bell's palsy from facial nerve schwannoma. It should also be remembered that the prominent circumneural arteriovenous plexus surrounding the facial nerve causes segmental facial nerve enhancement distal to the labyrinthine segment in nearly 75% thin section T1 enhanced MR scans.

CT temporal bone aids in surgical planning as it clearly shows the relationship of the facial nerve canal to normal anatomical landmarks such as the ossicles which are not seen on MRI. MRI will help in identification of soft tissue facial nerve abnormalities. It helps in differentiating masses around the facial nerve like lipoma, hemangioma and cholesteatoma from tumors arising from the nerve. It can also help in determination of perineural spread of malignant lesions like parotid malignancies.

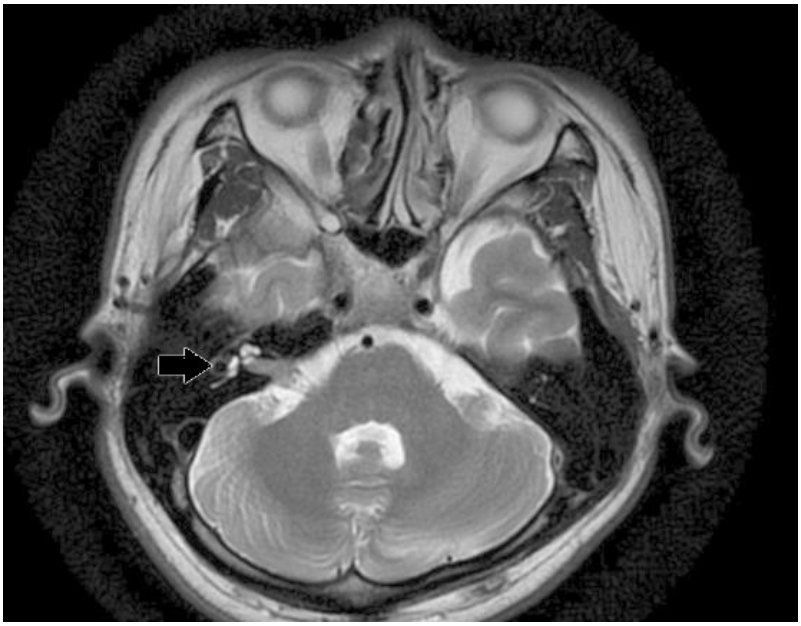
Facial nerve schwannomas involving CPA and IAC segments of facial nerve may be indistinguishable from acoustic schwannoma if there is no extension into the labyrinthine segment of the facial nerve is present. It is imperative that all cases of newly diagnosed acoustic schwannoma should be inspected for the presence of a labyrinthine tail that identifies them as facial nerve schwannoma in MRI images. When extensive a CPA-IAC facial nerve schwannoma can present with a distinct image apperarance i.e. "dumbbell" shape mass due to extension from IAC fundus through the labyrinthine segment and into the geniculate fossa. This feature along with T1 MR enhancement and sharply scalloped / fusiform enlargement of facial nerve canal is diagnostic of facial nerve schwannoma.

Lesions involving geniculate ganglion primarily or extending along the greater superficial petrosal nerve can present as a middle cranial fossa mass. When lesions of the middle cranial fossa are

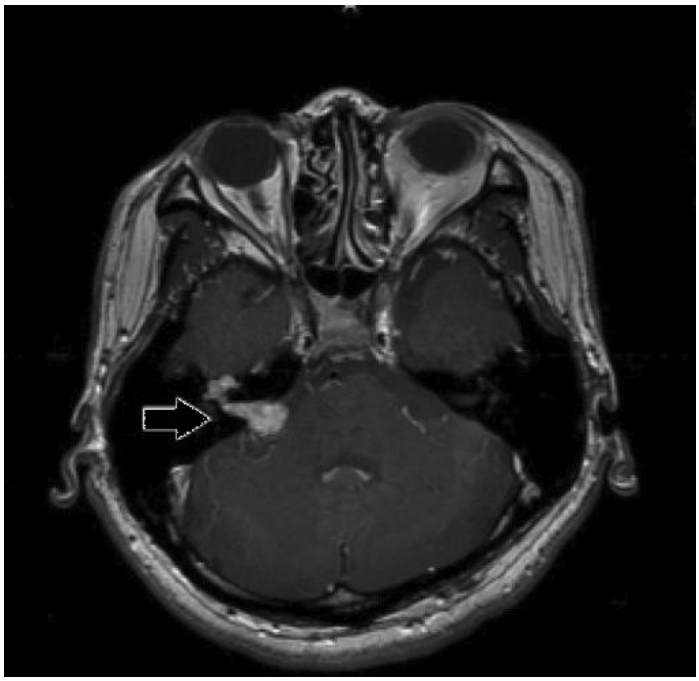
determined to be extra-axial, inspection of skull base and temporal bone should be done to rule out origin from the geniculate ganglion. Lesions involving geniculate ganglion will have a bulbous enlargement at the geniculate fossa itself. Facial nerve scwhannomas emanating from the greater superficial petrosal nerve scallops the anterior margin of the geniculate fossa and adjacent bony petrous apex.

The tympanic segment of the facial nerve lacks the thick surrounding bony architecture of the labyrinthine segment of the facial nerve canal. Facial nerve schwannomas arising from this segment often do not have the fusiform morphology, but instead are multilobular. When lobulation occurs superiorly or medially then a fistula of lateral canal may be caused. Inferolateral lobulation into the middle ear cavity cause lateral displacement of the ossicles causing conductive hearing loss in these patients.

Involvement of mastoid segment of facial nerve presents an interesting picture in MR Images. Since mastoid air cells contain thin walled bony septum, these septum are easily eroded by the mass and will be visible in MR images as irregular tumor margins with bone erosion. CT images especially with bone window settings explains these margins by showing facial nerve schwannoma breaking into surrounding mastoid air cells. If CT is not done in these patients then a misdiganosis will result.



Contrast enhanced T2 weighted image MR showing intracanalicular tumor in the region of CP angle indicated by black arrow



Contrast enhanced T1 weighted MR image showing enhancing mass at the CP angle and IAC with involvement of labyrinthine segment and geniculate ganglion (black arrow)

Electromyography:

This is helpful in quantifying the nerve's residual motor function and in predicting the post operative prognosis of facial recovery after nerve reconstruction.

Electroneurinography:

This test helps in predicting post operative prognosis of facial palsy. Results are considered to be better when the nerve is repaired if compound action potentials decrease by no more than 50%.

Management:

This involves surgical removal of the mass and reconstruction of facial nerve. The approach depneds on the site of the tumor, size of the tumor and hearing loss. Timing for surgery is again controversial,

because facial nerve neuromas are very slow growing tumors. Surgery should be deferred until the facial nerve function deteriorates.

To delay the surgery would allow the tumor to expand outside the natural bony confines. If the tumor becomes too large then surgery becomes that much difficult and likelihood of post op complications also increases. The facial nerve recovery is also poor.

If inner ear is invaded by the tumor then eventually the brain stem may become compressed due to the bulk of the mass. Hence surgical resection of facial nerve schwannoma should be resorted to without delay in patients with progressive facial palsy or in patients with large CP angle tumors that could compress the brain stem or produce hydrocephalus. Tumors involving the inner ear is also another indication for immediate surgery.

Tumor proximal to geniculate ganglion with serviceable hearing:

This should be approached via the middle cranial fossa provided the tumor does not extend far into the CP angle.

Tumor proximal to mid IAC with < = 1 cm of the CPA component an extended middle cranial fossa approach is ideal.

Lesions with CPA component > 1 cm: Retrosigmoid approach gives best access with added advantage of hearing preservation.

In lesions with non serviceable hearing a translabyrinthine approach is the best and most direct approach to the mass. This approach also provides the best access for facial nerve grafting.

Involvement of tympanic segment: This can be managed by transmastoid approach with facial recess opening.

Mastoid segment: Masses involving mastoid segment of the facial nerve can be accessed by mastoidectomy. Additional extratemporal extension may require following the nerve to the parotid gland.

It should be remembered that the tumor must be removed along with the affected segment of facial nerve and cable grafting will have to be resorted to in order to restore the function of facial nerve at least partially. Grater auricular nerve / sural nerve are excellent graft materials for grafting purposes.

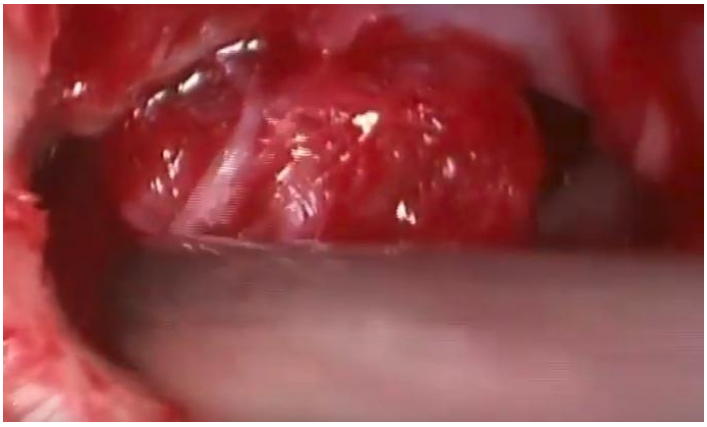
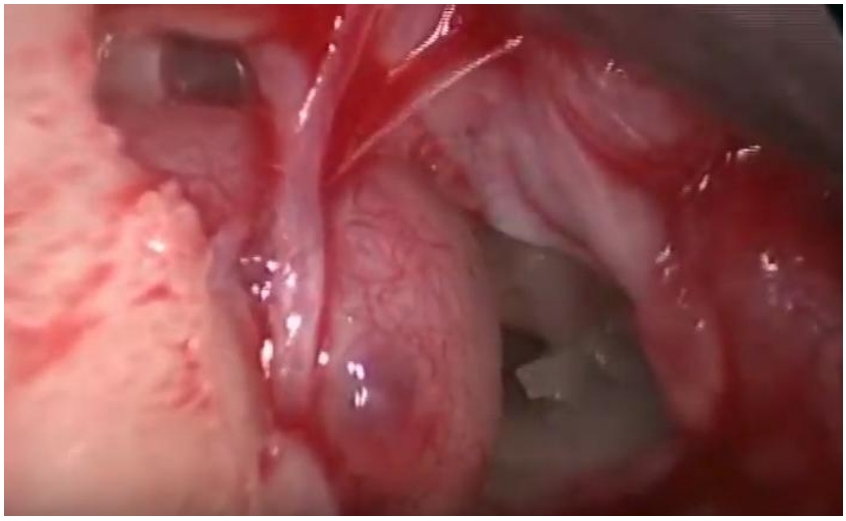


Image showing facial nerve schwannoma exposed



Facial nerve schwannoma clearly seen under chorda tympani nerve