Review Article

Vestibular Schwannomas

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Introduction

Vestibular schwannoma also called Acoustic neurinoma is the commonest tumour occurring in the cerebello-pontine angle and accounts for about 80 to 90% of the cases. Cerebellopontine angle is formed by the petrous part of the temporal bone, pons and cerebellum and floor of the space is formed by the middle cerebellar peduncle. The trigeminal and trochlear nerves located near the superior limb, glossopharyngeal, vagus and accessory nerves are located near the inferior limb and the spinal accessory nerve near or within the angular space between the two limbs. The facial and vestibulocochlear nerve which are farther apart in the brainstem converge and meet at the internal auditory meatus. Cerebellopontine angle tumors are challenging for neurosurgeons because of the vital structures lying there and small space of the region. Anterior inferior cerebellar artery lies close to the seventh and eighth nerves.

Incidence

The commonest lesion in the cerebellopontine angle is vestibular schwannoma accounting for about 80% to 90% of the cases, with meningioma being the next common accounting for 5% to 10% of the cases followed by epidermoid (5%-7%), dermoid, arachnoid cyst, neuroenteric cyst, extension of brainstem gliomas and craniopharngiomas into the cerebellopontine angle, trigeminal schwannomas and lower cranial schwannomas. Most of the lesions are benign. 80% of eighth nerve schwannomas arise from the vestibular component and 5%-7% arise from the cochlear component[1,2].

Pathology

Vestibular schwannomas arise most commonly from the vestibular component of eighth cranial nerve. The tumor originates from the junctional zone where central and peripheral myelin meet. Grossly, the tumor is rubbery and pale, variable consistency with a well defined capsule with the varying degrees of vascularity. Microscopically tumor is composed of spindle cells with elongated nuclei and fibrillary cytoplasm, arranged in two patterns - Antoni A and Antoni B. Antoni A tissue is compactly arranged with elongated bipolar cells, Antoni B is less structured and loosely arranged. Antoni B type of tissue is seen mostly in large tumors with nuclear pleomorphism and mitotic figures are rare.

Neurofibromatosis-1(NF-1) : NF-1 is also known as peripheral neurofibromatosis. The genetic abnormality is thought to be in chromosome 17. The patients usually present with peripheral neurofibromatosis with cutaneous lesions, lisch nodules, optic glioma and axillary or inguinal freckling and not associated with vestibular schwannoma.

Neurofibromatosis-2(NF-2): NF-2 patients commonly present with bilateral vestibular schwannoma, multiple meningiomas and posterior subcapsular cataract. The genetic abnormality is in chromosome-22.

Vestibular schwannomas start within the internal auditory meatus (intracanalicular stage) and as they grow erode the internal auditory meatus and extend into the cerebellopontine angle cistern (cisternal stage). As the tumor enlarges further compresses the brainstem and because of fourth ventricular obstruction. Obstructive hydrocephalus ensues (brainstem stage).

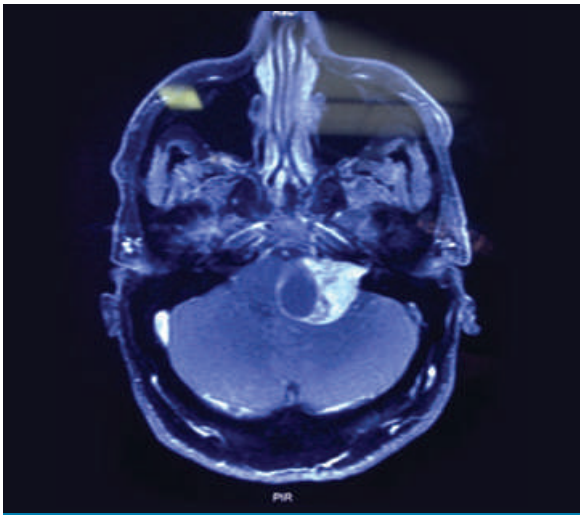

Fig 1: Pre-operative MRI(T1 contrast) showing left vestibular schwannoma.

Clinical features

Sensorineural hearing loss with impaired speech discrimination is the earliest symptom in vestibular

schwannoma. Some patients have preceeding tinnitus and transient vestibular symptoms. As the tumor progresses, patients develop features of other cranial nerve involvement including trigeminal, facial and lower cranial nerves depending on the extension of the tumor. Later, features of cerebellar and brainstem involvement and features of raised intracranial pressure occur.

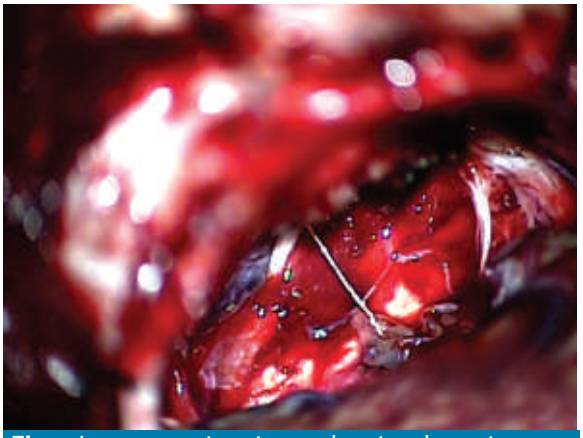

Fig 2: Intra-operative picture showing the various structures after excision of vestibular schwannoma.

Investigations: (Neuro-otological evaluation):

Neuro-otological tests: Pure tone audiometry shows features of sensorineural deafness with elevation of both air and bone conduction thresholds.

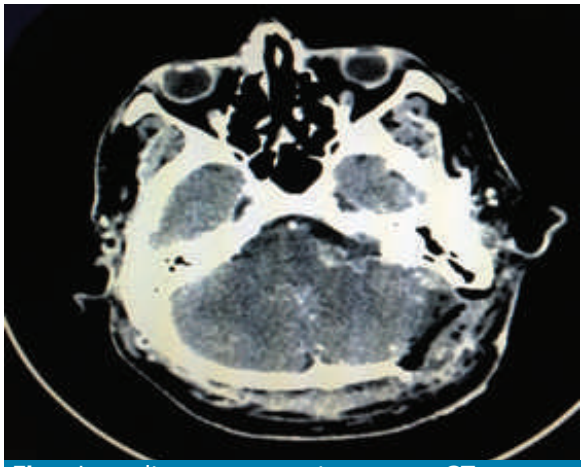

Fig 3: Immediate post-operative contrast CT scan showing complete tumor exicision.

Speech discrimination audiometry shows low speech discrimination scores. Retrocochlear lesions have lower scores than cochlear lesions.

Impedance audiometry is a sensitive index of retrocochlear disease. It consists of static compliance, tympanometry, acoustic or stapedial reflex. Static compliance and tympanometry are done to assess the middle ear disease and stapedial reflex is an indicator of retrocochlear disease. Patient with retrocochlear lesions show absent stapedial reflex.

Age	Sex	Diagnosis	Approach	Facial nerve preservation
47 yrs	Female	Right vestibular schwannoma	Retrosigmoid suboccipital approach	Yes
24 yrs	Male	Right vestibular schwannoma	Retrosigmoid suboccipital approach	No
55 yrs	Female	Left vestibular schwannoma	Retrosigmoid suboccipital approach	No
57 yrs	Female	Left vestibular schwannoma	Retrosigmoid suboccipital approach	No
30yrs	Male	Right vestibular schwannoma	Retrosigmoid suboccipital approach	Yes
25 yrs	Male	Right vestibular schwannoma	Retrosigmoid suboccipital approach	Yes
26 yrs	Male	Left vestibular schwannoma	Retrosigmoid suboccipital approach	Yes
52 yrs	Male	Left vestibular schwannoma	Retrosigmoid suboccipital approach	No
42 yrs	Female	Left vestibular schwannoma	Retrosigmoid suboccipital approach	No
59 yrs	Male	Left vestibular schwannoma	Retrosigmoid suboccipital approach	Yes
40yrs	Female	Left vestibular schwannoma	Retrosigmoid suboccipital approach	Yes
40 yrs	Female	Left trigeminal schwannoma (dumbbell schwannoma)	Combined middle fossa and retrosigmoid approach	Yes
35 yrs	Female	Right cerebellopontine angle meningioma	Retrosigmoid suboccipital approach	Yes
24 yrs	Male	Right vestibular schwannoma	Retrosigmoid suboccipital approach	No
35 yrs	Male	Right trigeminal schwannoma	Retosigmoid suboccipital approach	No

Table 1: Details of the cases of cerebellopontine angle tumors operated

Brainstem auditory evoked response is the most sensitive and specific audiological test. Increased interpeak latencies between wave I to III (> 2.5 ms), III to V (>2.3ms) and I to V (>4.4ms) and interaural wave V latency > 0.2ms are strongly suggestive of retrocochlear pathology (vestibular schwannoma). Other abnormalities include absent or poor wave form and absence of all waves except wave I.

Imaging

Magnetic Resonanace Imaging (brain): Gold standard for diagnosis, tumor is usually hypointense in relation to the csf in T1 images and iso-intense to midly hyperintense on T2 images and will enhance homogenously with gadolinium in vestibular schwannoma.

Computed Tomography (brain) with thin posterior fossa slices is done to identify bone destruction, expansion of internal auditory meatus. Position of labyrinth in relation to the fundus of the tumor and high riding jugular bulb.

Management

Observation.

Surgery.

Stereotactic Radiosurgery.

Radiation therapy.

Observation: For patients whom surgery carries risk and this is adopted in elderly patients without any neurological symptoms as these tumors are generally slow growing.

Stereotactic radiosurgery

Generally, tumors less than 3cm size are treated with stereotactic radiosurgery. The goal here is to stop the tumor growth and not to shrink or remove the tumor.

Surgery:

Surgery for cerebellopontine angle tumour is challenging because of the narrow space and the surrounding vital structures. The aim of the surgery is total excision of the tumor with preservation of cranial nerve function to the possible extent with good quality of life.

The main surgical approaches are retromastoid suboccipital transmeatal approach, middle fossa approach and translabyrinthine approach. Other approaches are retrolabyrinthine approach, trans canal approach, subtemporal transtentorial approach.

Retrosigmoid suboccipital approach is the most commonly used approach especially for the medium and large sized tumours. The advantages of this approach are the possibility of hearing preservation and better visualization of brainstem and other cranial nerves. But this is more suited for larger tumors; the need for cerebellar retraction and the risk of air embolism especially with sitting postion are the other disadvantages.

Translabyrinthine approach is ideally suited for small intracanalicular tumours where hearing is already fully compromised.

Middle cranial fossa approach is suitable for tumours with predominant superior extension. Hearing preservation is better with this approach. But it is not suitable for tumours with predominant inferior extension in the posterior fossa.

Chettinad Hospital experience

Out of 15 cases of cerebellopontine angle tumors operated during the last three years twelve were vestibular schwannomas, two were trigeminal schwannomas and one was a cerebellopontine angle meningioma. All were large tumors more than 4cm size. Retrosigmoid suboccipital approach was used in all the cases except one trigeminal schwannoma where both middle and posterior fossa approaches were used. Total excision of the tumor was possible in all the cases. Anatomical preservation of facial nerve was possible in eight cases (Ecg 1,2,3). There was no other major morbidity or mortality in this series. Details of these cases are shown in the table1.

Conclusions

Vestibular schwannomas are the commonest cerebellopontine angle tumors. With the advent of modern microsurgical techniques it is possible to excise these tumors with facial nerve and many times hearing preservation. A previous morbidity and mortality associated with these tumors have reduced drastically. Early diagnosis of these tumors with modern neuro-otological tests and imaging is possible and hence early treatment

References

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2) Ramamurthi R, Mohan SM. Acoustic Schwannomas. In Ramamurthi and Tandon's Textbook of Neurosurgery. Third edition Ed.Tandon PN, Ramamurthi R. Jaypee Brothers 2012 pp 1761-1791.