

Multiple Neurofibromatosis Type- 2 (Von Recklinhausen's) With Acoustic Neuroma- A Case Report.

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ABSTRACT

A 62 year old male patient presented with It sided hearing loss since 3 years associated with headache , vertigo and ear ache with nodules allover the body . He gave the history of his father having similar nodules allover the body but was not diagnosed or treated .With the help of MRI , PTA, BERA,OAE (audiological tests) and ENG(vestibular test) it was diagnosed as multiple neurofibromatosis type 2 with unilateral acoustic neuroma (left side).patient was kept on follow up and observation for last 4 years . with no worsening of symptoms Repeat MRI showed no apparent increase in size of tumor This case is reported for its clinical importance and rarity.

Keywords: Multiple Neurofibromatosis, Von Recklinhausen's, Acoustic Neuroma, BERA, MRI, OAE, ENG.

INTRODUCTION

First documented fully by Sandilfort in 1777. It usually arises from the Glial neurilemmal region of the vestibular nerve. Chromosome 22q is responsible for development of both sporadic unilateral and bilateral lesions of Neurofibromatosis. Annual incidence of 1:100,000 in the UK but it is variable from place to place. There is a progressive but slow destruction of the vestibular nerves , this produces pressure effects on the surrounding structures. There is a double layer of arachnoid over the tumor due to invagination- hence there is a good plane of dissection. It may be associated with an Arachnoid cyst. Erosion of the IAM occurs particularly at porus. When the size is more than 2 cm it grows intracranially, first it compresses the Trigeminal nerve as it crosses the petrous apex to enter into the meckels cave.

Pathology Gross – Well encapsulated, firm with nodular surface, well defined plane of separation. Interior of the tumor is much softer.

Histology – The neoplastic cells show two characteristic patterns.

Antoni A – Palisading Pattern

Antoni B – Reticular Pattern

Inner Ear – Degeneration of the cochlea / vestibule, spiral ganglion loss, raised protein levels.

Clinical Features – 6 stages

Asymptomatic

Otological – SNHL, Vertigo, rarely VII palsy.

Trigeminal – Tumor > 2 cm, loss of corneal reflex.

Brain Stem & Cerebellar Compression – Ataxia, lower CN palsy.

Raised intracranial pressure

Terminal stage

Examination & investigations

- Detailed clinical history
- Thorough ENT examination
- CNS examination for corneal reflex, cranial nerves, Cerebellar functions, etc
- PTA
- IMPEDENCE AUDIOMETRY
- OAE
- BERA
- ENG
- CT / MRI SCAN

CASE REPORT

A 62 yr old male Pt presented to OP dept with reduced hearing in the left ear since 3 years Occasional rotatory vertigo lasting for few minutes 6 months Head ache, ear ache on the L side 3 months Presence of multiple swellings all over the body since birth. Family history- father had similar

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nodules all over the body for which he was not diagnosed or treated.



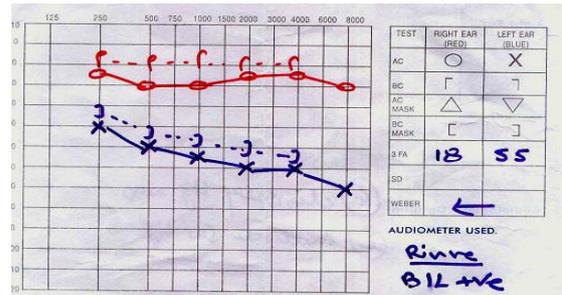
Examination

Multiple swelling of variable size from 1/2 cm to 3 cm in size, firm non tender seen all over the body.
 Left Ear
 Ext ear canal, Tympanic Membrane - Normal
 Tuning Fork Tests-- Lt sensory neural deafness detected.
 Facial nerve - Normal
 CNS – Absent corneal reflex in the left eye, other Cranial nerves are normal.
 Right Ear - normal



Investigations

Pure Tone Audiometry – L Snhl



Impedence audiometry

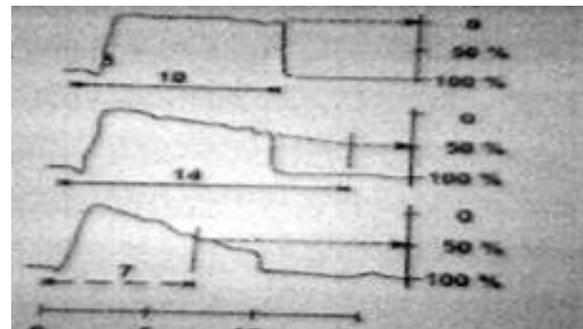
Acoustic reflex decay –

There is a fall in the amplitude of the stapedial reflex to less than 50% within 10 seconds.

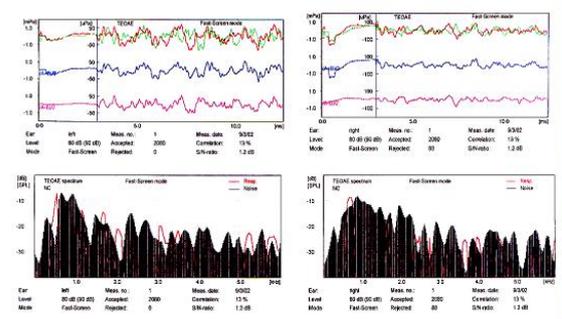
This is indicative of Retrocochlear pathology.

Other Tests That Can Be Used To Detect Retrocochlear Pathology By Impedence

- Metz Recruitment
- Sensitivity Prediction From Acoustic Reflex
- Acoustic Reflex Latency

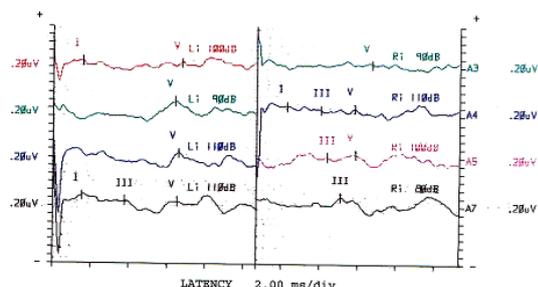


Otoacoustic Emissions – absent in both



Bera

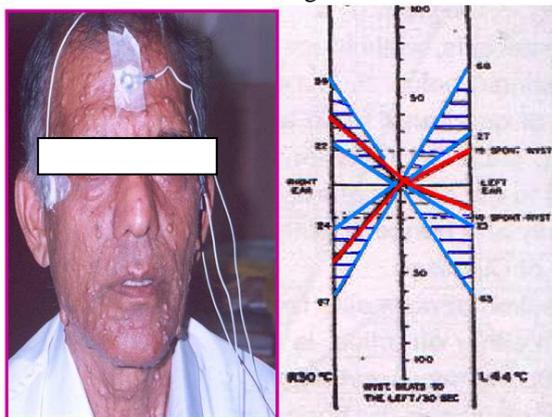
- L Ear - waves I, II, III are present only at higher stimulus, poor morphology of waves I, III, delayed latency of wave V, Prolonged interpeak latencies – suggestive of retrocochlear pathology with moderate SNHL.
- R Ear - waves I, II, III are present at high intensities (110 db), high artefacts noted due to multiple neurofibromatosis.



ENG

Caloric Tests –

ENG - CODE 0011 indicating L Canal Paresis



MRI Scan – Brain

- There is a well defined rounded T1 and T2 isointense lesion in the left CP angle attributing the 7th & 8th nerve complex



Treatment

Patient is kept on follow up and observation for the last 4 years with no worsening of symptoms Repeat MRI showed no apparent increase in size of tumor Case is reported for its rarity

DISCUSSION & CONCLUSION

This is a rare case of Acoustic Neuroma with Multiple Neurofibromatosis, the clinical presentation being giddiness, tinnitus & hearing loss. In a case of unilateral sensory neural hearing loss PTA, Impedence, BERA, OAE, ENG, CT & MRI are useful investigations to rule out retrocochlear pathology. Confirmation of acoustic Neuroma is by a CT scan or MRI , which also gives the extent of

the tumor, thus helping in planning appropriate type of surgery. These tumors can be present bilaterally.

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