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The Advantages of Having Tone Burst Auditory Brainstem Response in Finding Hearing in Patients with Cerebellopontine Angle Tumor- A Case Study

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ABSTRACT

Acoustic neuroma is a rare non cancerous tumor. It grows slowly from on over production of Schwann cells and also called a vestibular schwannoma. The tumor then presses on the hearing and balance nerves in the inner ear. Schwann cells normally wrap around and support nerve fibers. A large tumor can press on the facial nerve or brain structures. The early signs of Acoustic Neuroma are Unilateral or Asymmetric hearing loss and/or tinnitus, loss of balance from which Auditory symptoms are usually the first indication of a developing Acoustic neuroma. Unfortunately early detection of tumor is sometimes difficult because of the symptoms may be subtle and may not appear in beginning stage of the growth. Once these symptoms appear, the following examination (PTA, Immittance, OAE, ABR, HAT) are essential for proper diagnosis.
CT (Computerized tomography) and MRI (Magnetic resonance imaging) scans help to determine the location and size of the tumour. Early diagnosis offers the best opportunity for successful treatment.

Key words: Acoustic neuroma, Hearing loss, tone burst ABR.

INTRODUCTION

Acoustic neuroma or vestibular schwannoma is a benign tumour of the Schwann cell most commonly arises from the vestibular nerve. [1] They occur predominantly on the vestibular division of the VIIIth nerve of the oligodendroglial /Schwann cell interface, at or within the internal auditory meatus. [2]

Vestibular schwannoma predominantly affects adults in their fifth and sixth decades and are indeed rarer in children, in which are mainly due to neurofibromatosis type 2.It accounts 5-10% of intracranial tumours and most common neoplastic lesion in the cerebellopontine angle. In the year 2004-2010 the incidence of vestibular schwannoma was 17.4 in 1 million inhabitants. In U.S the overall incidence was 1.09 per 100,000 populations. Incidence increased with age to a peak of 2.93 p100, 000 in the 65-74 year old age group. [3] Approximately 10 tumours are diagnosed per million individuals each year. VS are slow growing tumour, growing at approximately 1-2mm per year.

The cause of an acoustic neuroma is generally unkown but can be related to von Recklingausen neurofibromatosis. [4] Acoustic neuroma is allied to neurofibromatosis type 2 (defect on chromosome 22) bilateral disease. It had a causative predisposition mutation. The continuous noise exposure may also cause NF2. Radiation exposure may predispose a patient to the development of that condition as well. [7]

As it is a slow growing tumour, it takes some time before signs and symptoms present. It is a sensorineural deafness and can also present as vertigo, nausea, altered balance, Instability while moving the head and nystagmus and the majority patients will have tinnitus. The tumors may cause

dysfunction of the vestibular cochlear nerve, including unilateral hyeracusis. Hearing loss, tinnitus and vertigo. Diplopia may occur due to abducens palsy. If the trigeminal nerve involved, facial parasthesia, facial pain, absent corneal reflex and mastication disorder can occur. Matthies et al provide a series of 1,000 patients and explain that 95% of patients undergoing surgery have hearing loss prior to their operation and 63% were suffering from tinnitus. [5,6]

Due to the compression of the facial nerve, the symptoms may include paralysis, increased lacrimation and facial weakness, disturbance in the trigeminal nerve cause tingling in the tongue impairment in the corneal reflex and glossopharyngeal and vagus nerves make palatal paresis, hoarseness of voice and dysphagia. Large tumor may cause gait ataxia, in coordination of the upper limb, and rarely dysarthria. [7]

CASE REPORT

A 78 years old male, came to the Department of Audiology with the concern of hearing loss in Both ears for the past 40 years with an acquired onset (has an history of an injury to the left ear before 40 years and it was not treated) and progressive in nature, the patient also had a family history of hearing loss (the patient's sister), he had localization difficulties. His workplace is filled with continuous machinery noise and he was working for almost 18 years in that environment and it was on daily basis, he experiences noisy tinnitus occasionally in both ears and had vertigo for 2-3 months before reporting to the Audiological Department. No history of vomiting and headache. His medical history reveals that he was a Diabetic patient and he was under medication for the same for the past 7 years. The patient undergone MRI and it reveals that Right Acoustic Schwannoma in the right Cerebellopontine angle extending along right Internal Acoustic Meatus (IAM)- 6mm of Fundal cap of CSF present, another MRI was done after 8 months and it reveals that there is no significant internal change when compared to the previous investigation. The patient undergone Audiological test battery which includes: otoscopic examination which helps in visualizing and examining the condition of the ear canal and ear drum; Tuning fork tests values particularly to screen types of hearing loss prior to Puretone audiometry; Pure Tone Audiometry (PTA) is the first step in Audiological evaluation which test in both air and bone conduction modalities usually the patient's with acoustic neuroma will have a configuration that shows High frequency hearing loss; Speech Audiometry is the test that assess the patient's ability to hear and to understand speech, understanding of speech is usually impaired in the retrocochlear lesion; Immittance Audiometry helps in determining the middle pathology; Otoacoustic Emission (OAE) is necessary to rule out cochlear status, specifically hair cell function, OAEs does not show any significant changes in both tumour and non-tumour condition; Auditory Brainstem Response (ABR) is the reference screening technique for acoustic neuroma and Hearing Aid Trial (HAT) is done to determine the management options that are available for acoustic neuroma.

The test batteries are as follows:

- The otoscopic examination reveals that the right side has retracted and dull tympanic membrane, whereas the left side was intact.
- The Tuning fork test was done. It reveals Weber lateralized to Right side
- PTA reveals that Right ear was Moderately Severe Mixed Hearing Loss and Left ear was Profound Hearing Loss (refer figure 1).

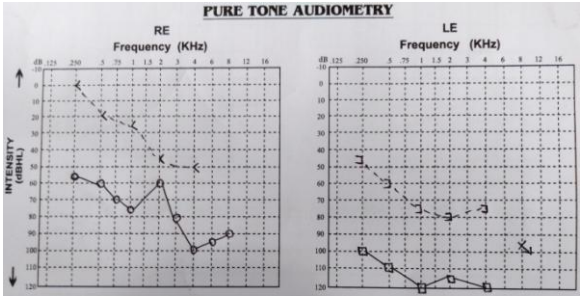

FIGURE 1: PURE TONE AUDIOMETRY

- Speech Audiometry was performed only to the right ear and could not be tracked in the left ear as the PTA average was beyond the limit, the right ear reveals that the Speech Recognition Scores(SRT) were 75 dBHL and Speech Identification Scores (SIS) were 95% , the Speech Awareness Level (SAL) was 100dBHL in left ear.
- Immittance reveals that Both ears was 'A' type tympanogram (refer figure 2), Multicomponent tympanometry was done and it reveals that the Right ear was indicating Mass dominant pathology and the Left ear showed Normal resonant frequency (refer table 1).

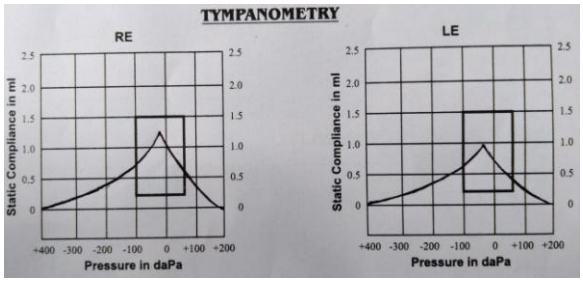

FIGURE 2: IMMITTANCE AUDIOMETRY

FREQUENCY	B & G PATTERNS	
	RIGHT EAR	LEFT EAR
226 Hz	1B1G	1B1G
678 Hz	3B1G	1B1G
1000 Hz	5B3G	3B1G

OAE- DPOAEs were performed which reveals that DPOAEs are absent in both ears with reduced SNR (refer figure 3).

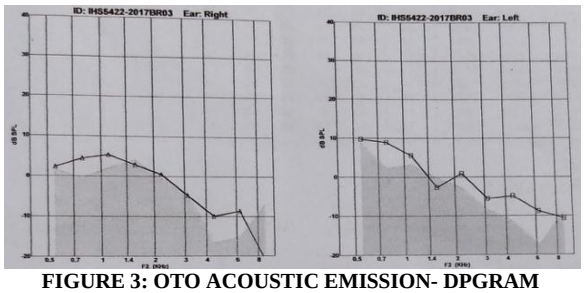

FIGURE 3: OTO ACOUSTIC EMISSION- DPGRAM

ABR- ABR was performed using click stimulus for which no replicable peaks could be obtained even at 90dBnHL in rarefaction and alternating polarities at the rate of 11.1/s, however, we could obtain some replicable 5th peak for 500 Hz tone burst stimuli in right ear and for left ear we couldn't obtain any replicable peaks for both clicks and tone burst stimuli at the rate of 11.1/s in rarefaction and alternating polarities (refer figure 4).

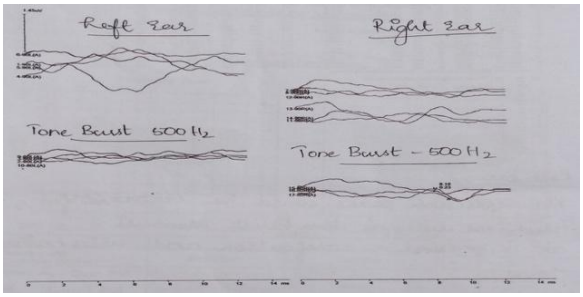

FIGURE 4: AUDITORY BRAINSTEM RESPONSE

Finally the patient undergone HAT with two set of hearing aids, one was his own hearing aids that was Siemens nitro BTE in both ears and trial hearing aid that was Hansaton Sound HD RIC in right ear and Hansaton flow 675 UP in left ear, the trial reveals that aided responses are within the speech spectrum in right ear and aided responses are out the speech spectrum with poor speech perception scores (refer figure5)

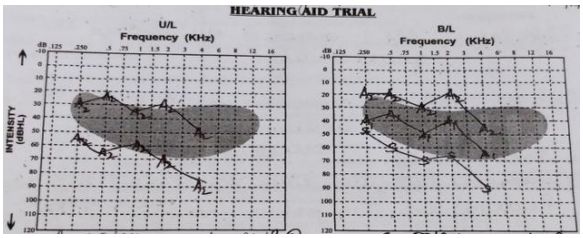

FIGURE 5: HEARING AID TRIAL

We recommended the client to use same or similar gain hearing aid with a RIC power receiver in right ear which he was benefitted from the hearing aid trial, whereas in left ear he can use hearing aid only for awareness purposes and to have a review with an otolaryngologist.

CONCLUSION

Acoustic neuroma is a condition which affects speech perception where symptoms are exhibited later. Hence that has to be noted earlier to avoid speech perception problem. In order to find the acoustic neuroma at an early stage tone burst ABR is a best solution.

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