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Case Report

Audio-Vestibular Profiling and Rehabilitation Option for Vestibular Schwannomas - A Case Study

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Abstract

Introduction: The balance and hearing nerves that supply the inner ear can develop into benign, slow-growing tumors called vestibular schwannomas (VS), sometimes known as acoustic neuromas, acoustic neurinomas, or acoustic neurilemomas.

Case Report: A 70-year-old male subject reported to the clinic with a complaint of reduced hearing sensitivity in the left ear for 4-5 months in situations where competing stimuli are present (background noise, group discussion, etc.) with vertigo since last 3 month.

Diagnosis: The PTA revealed Mild sensorineural hearing loss (PTA:33.3 dBHL) in the right ear and Moderate tending to moderately severe high-frequency sloping sensorineural hearing loss (PTA:51.6 dBHL) in the left ear.SISI revealed a score of 10% in both the ears at 500 Hz, 1KHz and 2KHz frequencies which suggestive of no cochlear pathology. Tone decay test revealed decay of less than 30 dBHL suggesting no Retro Cochlear Pathology in the right ear while the decay of >30 dBHL at 1K Hz and 2 KHz suggestive of retro cochlear pathology in the left ear. OAE is absent in the left ear. ABR was absent in the left ear at a slow and high stimulus rate. Contralateral ABR was also absent due to mass effect. No peak was obtained in the left ear in OVEMP.

Conclusion: The sensitivity of each test varies from person to person based on the size of tumor and the location of the tumour. Test battery approach is the best way to differentiate the pathology. A combination of radiological and audiovestibular assessment will provide a better picture about vestibular schwannomas.

Keywords: Vestibular Schwannomas; Audio-vestibular Profile; Case Study; ABR

Introduction

The balance and hearing nerves that supply the inner ear can develop into benign, slow-growing tumors called vestibular schwannomas (VS), sometimes known as acoustic neuromas, acoustic neurinomas, or acoustic neurilemomas [1]. The cells known as Schwann cells, which typically wrap around nerve fibers like onion skin to support and insulate nerves, are overproduced in the tumor. As the vestibular schwannoma develops, it affects the nerves that control hearing and balance, typically leading to unilateral (one-sided) or asymmetric hearing loss, tinnitus (ear ringing), and vertigo/loss of balance [2]. The estimated annual incidence of these tumours ranges from approximately 0.6 to 1.9 per 100,000.

According to Mackle., et al. (2007), 3.7% of patients show atypical symptoms [3]. The signs and symptoms of VS include fluctuating hearing loss, tinnitus, ear fullness, vertigo, numbness in the face, facial twitching, etc. Based on the size of tumours the audiological findings vary from person to person [4]. Only pure tone audiometry is not a standard test to detect any tumours in the auditory nerve. There are a lot of tests used to identify or segregate pathology in cochlear or retro-cochlear systems. Nowadays MRI has the highest sensitivity in identifying even small-size tumours [5]. The sign and symptoms of VS varies from person to person. According to that diagnosis and treatment plan should vary. The primary purpose of the study is to profile audio-vestibular findings in a radiologically confirmed vestibular schwannomas case.

Case Report

A 70-year-old male subject reported to the clinic with a complaint of reduced hearing sensitivity in the left ear for 4-5 months in situations where competing stimuli are present (background noise, group discussion, etc.) He had a complaint of dizziness which occurred once in 1.5 months and at that time he felt the sensation of falling which lasted for 3 minutes only. After that incident, he never complained about any other attack of vertigo or dizziness. He can hear the sound but faces a lot of difficulties in understanding speech. After consultation with a physician, a detailed CT scan and MRI was conducted. The MRI findings revealed a Small (0.5*1.4*0.4cm) oblong-shaped altered signal intensity lesion in the left CPA. Lesion extending into the left internal auditory meatus with seventh and eighth nerves likely schwannoma. Consent from the patient was taken to upload his case details.

Audiological evaluation

An audiological evaluation was carried out after obtaining a complete case history of the patient. An otoscopy examination was carried out to evaluate the structure of the Tympanic Membrane and ear canal. After that tuning fork test was carried out followed by Pure tone audiometry (PTA), Speech audiometry, Oto acoustic emission (OAE), Auditory brainstem response (ABR), and vestibular assessment.

A tuning fork test was carried out using a 512 Hz tuning fork and the result indicated that bilateral Rinne positive and Weber lateralized to the Right ear. PTA was done in a sound-treated chamber by using a dual-channel GSI audiometer. The patient was seated in a comfortable chair, proper instruction was given before conducting PTA. The supra-aural headphone was used to estimate the Air-conduction threshold at all octave frequencies from 250 Hz to 8000 Hz in both ears and also inter-octave frequency when needed. Bone-conduction thresholds were estimated by using a B-61 bone vibrator. The PTA revealed Mild sensorineural hearing loss (PTA:33.3 dBHL) in the right ear and Moderate tending to moderately severe high-frequency sloping sensorineural hearing loss (PTA:51.6 dBHL) in the left ear.

Speech audiometry was conducted using Hindi spondee and Phonetically Balanced word list. Speech Recognition Threshold (SRT) in the right was 40 dBHL while on the left it was 80 dBHL. Making was applied to the right ear while doing Word Recognition Score (WRS) in the left ear at 50 dBHL. The WRS was obtained

and revealed 80% in the right ear but 40% in the left ear. The Most Comfortable Level (MCL) and Uncomfortable Level (UCL) for the right ear were 75dBHL and 105dBHL respectively. The same for the left was 85 dBHL and 105 dBHL respectively.

To find out the site of lesion special tests were carried out by using a GSI dual channel audiometer. Short Increment Sensitivity Index (SISI) revealed a score of 10% in both the ears at 500 Hz, 1KHz, and 2KHz frequencies which are suggestive of no cochlear pathology. A tone decay test using the Olsen-Noffsinger modification method was used and it revealed decay of less than 30 dBHL suggesting no Retro Cochlear Pathology in the right ear while the decay of >30 dBHL at 1K Hz and 2 KHz suggestive of retro cochlear pathology in the left ear.

Distortion product OAE (DPOAE) was carried out using a path medical instrument. This suggested normal Outer hair cells (OHC) functioning in the right ear but absent OAE in the left ear suggestive of gross peripheral abnormalities up to the level of OHC.

Auditory Brainstem Response (ABR) was used to evaluate the site of lesion by using slow rate stimulus (11.1/Sec) and High- rate stimulus (77.7/Sec). No peak I, III, or IV was obtained in the left ear at high-intensity stimulus level and both the rate of presentation. This suggests retro cochlear pathology in the left ear. We observed a prolonged absolute latency between III-IV in the right ear at 90 dBnHL intensity.

Vestibular assessments

Cervical VEMP was carried out using a Neurosoft instrument with proper placement of electrodes and instruction to the patient. The latency and amplitude of the peaks p13 and N23 are within normal limit in the right ear and a slight delay of p13 in the left ear. The interaural asymmetry ratio is less than 30% in both 500 Hz and 1KHz which is within the normal limit. Decreased amplitude was observed in the left ear which is more than 50% in comparison to right ear. Hence, the Vestibulo-collic reflex and the saccular function are Normal in right ear, but some abnormalities were reported in the left ear.

Occular- VEMP was carried out and revealed that the latency of the peak P1 and N1 are with in normal limits in the right ears. No peak was obtained in the left ear. The interaural asymmetry ratio could not obtained as there was no peak in left ear. Hence it can be assumed that all the structures are normal in the right ear but not in the left ear that might be due to the tumours in the left side of audio-vestibular nerve.

Provisional diagnosis

- Right ear: Mild sensorineural hearing loss. (Cochlear pathology)
- **Left ear:** Moderate tending to moderately severe high-frequency sloping sensorineural hearing loss.
- **Left ear:** ? Retro Cochlear Pathology: ? Vestibular Schwannoma (According to MRI).

Rehabilitation

Behind the Ear Hearing aid with 12 channels were programmed according to the audiogram. The noise reduction algorithm was activated and the directionality function was kept on. A venting of 1mm was introduced in the ear mould as the patient have better low frequency thresholds in the left ear. Aided WRS was carried out to evaluate the benefit from hearing aid and found to be 60%. Patient was satisfied with the hearing aid.

Discussion

Vestibular schwannomas are known to cause unilateral damage to both auditory and vestibular functions [6]. According to various previous studies, unilateral hearing loss is the initial sign of VS. The degree of hearing loss is not always associated with the anatomical site of the lesion. Speech audiometry, Differential Diagnostic, and special tests have a major role in the differential diagnosis of cochlear and retrocochlear pathology [7-9]. VEMP has a significant role in identifying the location of lesions in the audio-vestibular nerve. In this study the ABR result which indicates no peaks at all has consistence results with the previous study by Saleem., et al. 2019 [10]. According to Marn Joon Park., et al. (2022) in 40% of VS cases, abnormal ABR (No peak) was noted [11]. For tumours measuring <10 mm, the sensitivity of ABR was 66.7%, whereas it increased to 90.3% for tumours measuring >10 mm [11,12]. ABR by itself is not enough to screen for VS since it has the risk of producing false-negative results when looking at intracanalicular tumours that are tiny. But in patients with usable hearing, ABR may be used at a low cost to screen for VS measuring >10 mm, which supports the necessity for more active diagnostic and therapeutic techniques in clinical practice. According to Paul D., et al. sensitivity for ABR detection of vestibular schwannomas was 93.4% (95% CI 92.6-94.3, P = 0.0000) [12]. For tumors less than 1cm (8 studies, 176 patients) sensitivity was 85.8% (95% CI 80.6-90.1, P = 0.0116). Prolonged latency was also observed in the contralateral ear that's may be due to the mass effect of the lesion side which rotate the brainstem towards other side and cause a compression of later peak in contralateral ear. Deepa Aniket Valame., et al. in their study documented that in VS patient who have small tumours (<2 CM) has delayed latency and reduced amplitude in C-VEMP which is consistence with our findings [13]. Niels West., et al. 2018 in their study found out O-VEMP to be more sensitive in identifying VS [14]. In 100% of the participants, no O-VEMP was obtained in the affected side which was also observed in our case, indicating superior vestibular nerve (SVN) affection on the lesioned side. These findings will be help full in efficiently diagnosing VS patients. So many other tests like caloric test, VHIT test could be conducted to estimate the vestibular nerve function in this case, which is a limitation in this study. Comparisons of tumour size and ABR response, VEMP response could be correlated.

Conclusion

The sensitivity of each test varies from person to person based on the size of tumor and the location of the tumour. Test battery approach is the best way to differentiate the pathology. A combination of radiological and audiovestibular assessment will provide a better picture about vestibular schwannomas.

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