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LATE- ONSET BILATERAL AUDITORY NEUROPATHY SPECTRUM DISORDER:

A CASE STUDY

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ABSTRACT

Background: *Onset of auditory neuropathy spectrum disorder in early adulthood or middle age is exceptionally unusual. This report describes such an unusual finding of young adult onset, bilateral Auditory Neuropathy Spectrum Disorder.*

Key Words: *Auditory Neuropathy, Otoacoustic Emissions, auditory brainstem response, Acoustic Reflex, Cochlear Microphonics*

INTRODUCTION

Auditory Neuropathy Spectrum Disorder (ANS) is a condition characterized by sensorineural hearing loss with normal cochlear hair cell function (assessed by recordable Otoacoustic Emissions (OAEs) and absent or abnormal auditory brainstem response corroborated with absence of Middle ear reflexes [1]. Typically, auditory neuropathy spectrum disorder patients show a severe speech perception impairment, which appears reduced out of proportion to pure tone threshold, but the clinical presentation of auditory neuropathy spectrum disorder is quite complex. Auditory neuropathy spectrum disorder usually is identified early in life and is mostly bilateral in nature [4-8]. . Prevalence of auditory neuropathy spectrum disorder was reported to be 1.55 percent among 841 hearing-impaired students in the age range, 2 to 20 years [2]. In a group of 183 patients, a prevalence of 0.55 percent was found [3]. However, there is dearth of studies on prevalence

of auditory neuropathy spectrum disorder in adults. Onset of auditory neuropathy spectrum disorder in early adulthood or middle age is exceptionally unusual. 11% (7 of 59) of patients had an onset between the ages, 20 and 60 years [8] with a unilateral presentation. Some individuals who develop peripheral neuropathies in adulthood display auditory neuropathy spectrum disorder with a later onset. There are several studies reporting unilateral auditory neuropathy spectrum disorder in children below 12 years of age [10]. Slinger and Oba [8] reported 4 percent of their 59 cases to be unilateral. Unfortunately, they did not report the age of identification. However, Stuart and Mills observed an unusual finding of late onset unilateral auditory neuropathy spectrum disorder in a 64 year old female based on audiometric and imaging findings [9]. Herein, we describe such an unusual finding of young adult onset bilateral auditory neuropathy spectrum disorder based on reported case history, audiometric and imaging findings. An informed consent was taken from the patient for publication of the report.

CASE REPORT

A 20 year male patient presented with the chief complaint of a progressive bilateral hearing loss and tinnitus that had developed over the past 2 years attended the speech and hearing unit for hearing evaluation. There was no significant past medical history. During hearing evaluation, the patient was diagnosed as having Bilateral Sensorineural Hearing Loss. A subsequent magnetic resonance imaging of the brain and temporal bone was undertaken two weeks after the initial audiologic evaluation to rule out acoustic neuroma. Radiological reports showed no evidence of internal auditory canal mass or lesion on either side. Brain imaging was also negative for infarct, haemorrhage, mass, extra-axial fluid, or hydrocephalus.

During the second auditory evaluation, approximately three weeks later, an extensive behavioral and electrophysiological test battery was undertaken. A double-wall sound-treated audiometric suite, meeting specifications for permissible ambient noise, served as the test environment for behavioral testing. Immittance, Otoacoustic emissions, and evoked potential measures were recorded in a quiet clinical room with a middle ear analyzer, OAE systems, and an evoked potential system, respectively. An insert earphone delivered stimuli for all evoked potentials.

Thresholds for pure-tone and speech stimuli were determined using Madsen Orbiter 922 Clinical Audiometer. Pure tone Audiometry showed Bilateral sensorineural hearing loss that was present upto 1 KHz. Beyond 1 KHz, he showed normal hearing that was less than <25 dB HL (American National Standards Institute, 1996). Tone decay tests were performed with 500, 1000, and 2000 Hz pure-tone stimuli at 20 dB SL. Tone decay was negative bilaterally.

Word recognition was assessed binaurally, under earphones with 50 monosyllabic Phonetically Balanced (PB) Hindi word list in quiet at 30 dB above the spondee recognition threshold. Spondee recognition

thresholds were in agreement with three-frequency pure-tone averages for the left and right ears. Word recognition performance was poorer in both ears and the speech discrimination scores of right and left ears were 64 % and 60 % respectively.

Tympanometry was obtained using a 226 Hz probe tone using LABAT impedance meter. Ipsilateral and contralateral acoustic reflexes were assessed bilaterally with pure tone activator stimuli of 500 Hz, 1 KHz, 2 KHz and 4 KHz. Both ears showed type 'A' tympanogram. Ipsilateral and contralateral acoustic reflexes were absent with left-ear and right ear stimulation of pure-tone stimuli from 500 to 4000 Hz in octave steps. TEOAES were measured using Intelligent Hearing Systems (IHS-2125) and primary tones with an f2/f1 ratio of 1.2 and an L1/L2 of 65/55 dB SPL were used to evoke distortion product otoacoustic emissions (DPOAEs). The f2 frequencies ranged from 1000 to 8000 Hz. DPOAEs were deemed to be present if response amplitude was 3 dB or more above the noise floor at three consecutive frequencies. TEOAE'S were absent in both ears. DPOAEs were observed bilaterally with greater DPOAE amplitudes in both ears.

All evoked potentials were acquired while the patient was resting in quiet. Stimuli were delivered for each ear separately. Auditory brainstem responses were evoked with click stimuli and two channel recordings were obtained using Neuro audio (80 version) digital auditory brainstem response software. Tracings were recorded with different polarities, viz, rarefaction and condensation stimuli to check for cochlear microphonics (CM). CMs were not observed bilaterally and auditory brainstem response showed no waveforms at 100 and 90dBnHL in both ears. As CMs that are diagnostic of Auditory Neuropathy were not evident, after audiological investigations, this patient was referred for neurologic evaluation to rule out any other neuropathologic and/or related conditions. Based on symptoms presented, he was provisionally diagnosed as having (?) Mitochondrial myopathy. For confirmation, the subject underwent Lactic acid test, the results of which were within normal limits. Thus the audiological investigations were supportive and indicative of Auditory Neuropathy in this patient.

DISCUSSION

Although unilateral case studies of auditory neuropathy spectrum disorder have been previously reported in the literature [10] to the best of our knowledge, this is the first reported case of a late adult-onset (i.e..20 years of age) bilateral Auditory Neuropathy Spectrum Disorder. The patient presented with all the classic signs of auditory neuropathy spectrum disorder on both the sides: normal outer hair cell activity as evidenced with observable DPOAEs, absent middle ear acoustic reflexes, abnormal auditory brainstem response, and poor speech recognition scores.

Several key findings were critical to the diagnosis of auditory neuropathy spectrum disorder with this patient. First was the observation of OAEs. DPOAEs on both sides had large amplitudes from 250 to 8000

Hz. The DPOAEs were repeated across a one month period with two different devices to rule out artifact or equipment/ operator error, if any. An additional hallmark of auditory neuropathy spectrum disorder was the absent auditory brainstem response on both sides. One final observation was the patient's poor word-recognition even in quiet situation, which again was expected in a case with ANSD. A classic unusual observation was the absence of Cochlear Microphonics in this case, which is considered a diagnostic feature of Auditory Neuropathy Spectrum Disorder. However, it is observed that CMs diminishes over time in later stages of ANSD and hence should not be considered as diagnostic feature.

Finally, regarding patient's management, considering his poor binaural performance and his reported rigorous listening difficulties, any rehabilitative measures other than modified listening strategies (e.g., preferential seating, reducing or eliminating background noise, etc.) and hearing conservation should be of concern. However, cochlear implant seems to serve best in certain cases with ANSD and this option was not viable in this case because of poor affordability and socio economic status of the client.

CONCLUSIONS

An adult patient with bilateral SN hearing loss showing normal outer hair cell functions, absent auditory brainstem response and poor speech discrimination scores was diagnosed as having Auditory Neuropathy Spectrum Disorder. The key findings of behavioural, speech and physiological testing correlated with the diagnosis of late onset Auditory Neuropathy Spectrum Disorder. Although modified listening strategies are advised for such patients, an audiologist should explore the outcomes of cochlear implantation in individuals with auditory neuropathy spectrum disorder. This is mandatory in all cases of ANSD, irrespective of the age group.

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