

Case Report

Auditory neuropathy: A case report and review of literature

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Abstract:

Auditory Neuropathy (AN), also known as Auditory Dyssynchrony(AD) is a very often missed diagnosis in clinical practice. In AN, sound enters the inner ear normally but the transmission of signals from inner ear to brain is impaired. Due to the lack of advanced audiological test facilities in Bangladesh, mostly AN remains undiagnosed.

Here we described a 25 years old male, presented with difficulty in understanding speech for last 10 years. On audiologic evaluation, bilateral moderate sensorineural hearing loss was found on Pure tone audiogram (PTA). Speech discrimination scores were poor and disproportionate to his hearing threshold. Presence of Otoacoustic emission (OAE) revealed the normal function of cochlea. Conversely, absence of Auditory brainstem response (ABR) confirmed the definite dysfunction at auditory neural pathway and hence, the diagnosis of AN was confirmed. Since AN is an underdiagnosed condition in Bangladesh, otolaryngologists should be aware of this clinical entity. All the suspected cases should be referred to specialized centers, where advanced audiological facilities are available.

Key words: Auditory neuropathy, Auditory dyssynchroni, sensorineural, hearing loss

Introduction

Auditory neuropathy(AN) is also known as Auditory Dyssynchrony(AD).¹ These terms are used to classify patients with abnormal auditory neural response in presence of normal cochlear function.² Patients with AN present with varying degrees of hearing loss from mild to profound with extremely poor speech discrimination, regardless of degree of hearing loss. This inconsistency between

speech discrimination and degree of hearing serves as the first indication that a patient may have AN. Although this group of patients were first reported in late 1970's, the term Auditory neuropathy was first introduced at 1996 by Arnold Starr.³

In order to confirm the diagnosis of AN, appropriate auditory tests are those sensitive to cochlear and auditory nerve function, namely – Otoacoustic emission (OAE) and Auditory brainstem response (ABR). In Bangladesh facilities of OAE and ABR is available only in few centers in the capital Dhaka. Hence, AN is an underdiagnosed condition in Bangladesh. Till now, no paper on Auditory neuropathy has been found in Bangladesh on literature search.

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Case Report:

A 25 years old male patient from Rangamati, a hill district of Bangladesh, presented to the

specialized ENT hospital of SAHIC(Society for Assistance to Hearing Impaired Children), Mohakhali, Dhaka, with the complaints of difficulty in understanding speech along with hearing loss for last 10 years. His speech perception was so poor that his history was given by his father. His father stated that the patient was alright until his age of 15 years, then he developed difficulty in speech perception together with hearing loss, which gradually increased over time. His hearing loss was not associated with tinnitus and vertigo.

Further history concluded that, there was no history of aural discharge, trauma to the ear, head injury, ototoxicity, noise exposure or any other medical or surgical illness. Also there was no contributory antenatal, perinatal and post natal history as well as no positive history of hearing loss in family.

On clinical examination, his both Tympanic membrane(TM)s were intact and normal. On tuning fork test – Rinne’s test was positive bilaterally. Weber test was centralized. Absolute bone conduction test was reduced on both sides. Rest of the Ear, Nose, Throat and Head neck examination was normal.

On audiometric evaluation of this patient, pure tone audiogram (PTA) showed sensorineural hearing loss in both ears. (Figure 1) Average loss in right and left ear was 36.8dB and 50dB respectively.

Tympanometry showed normal middle ear pressure with bilateral A type curve. Stapedial reflex threshold (SRT) found absent on both

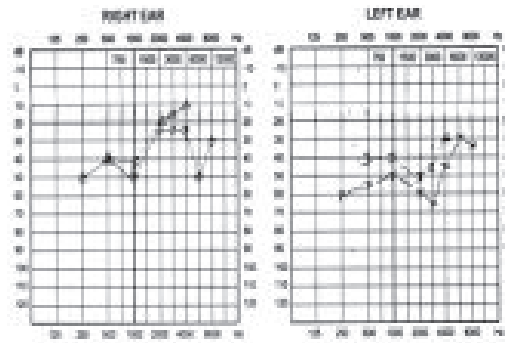


Figure-1: Pure tone audiogram

sides. Speech discrimination test showed very poor score on both sides. Overall discrimination score(ODS) was 30% at 75dB in right ear and 30% at 85 dB in left ear. (Figure 2)

Having these audiological reports, we suspected the case as auditory neuropathy. To confirm the diagnosis, Otoacoustic emission (OAE) and Auditory Brainstem Response

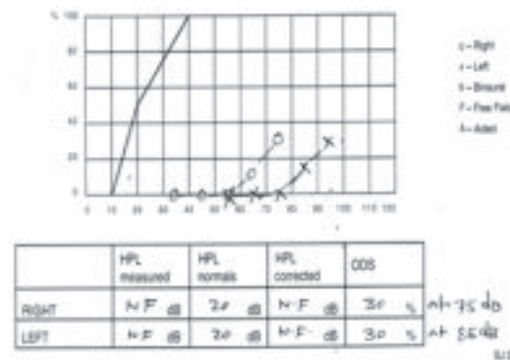


Figure 2: Speech discrimination test

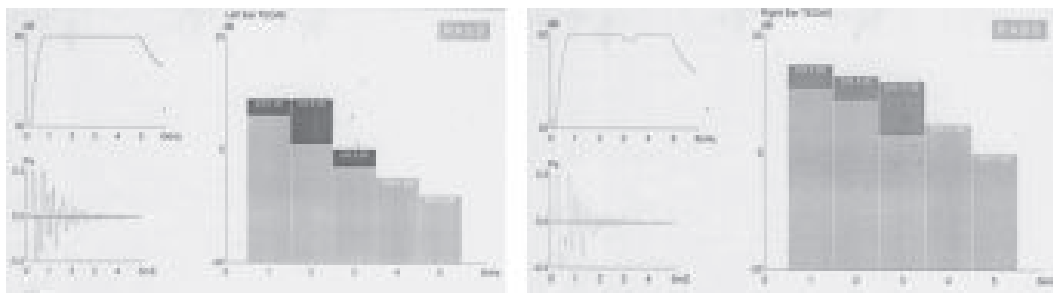


Figure-3: Otoacoustic Emission (OAE)

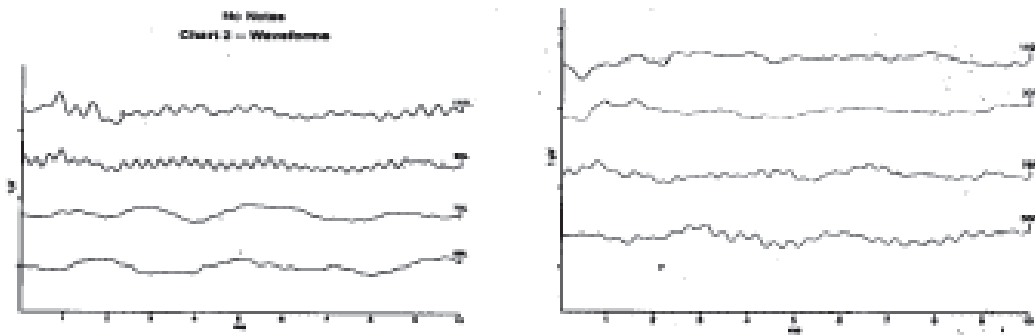


Figure 4: Auditory Brainstem Response (ABR)

(ABR) was done. Transient Evoked Otoacoustic emission (TEOAE) showed pass or presence of otoacoustic emission in both ears which indicates normal outer hair cell function of cochlea. (Figure 3)

On the other hand, ABR shows no definitive Vth wave on either side even after 110 dB auditory click stimuli (Figure 4), which reflects the definite dysfunction at the level of auditory neural pathways at both sides.

Besides the audiological work up, MRI scan of brain was also done and the normal findings excluded any intracranial lesion. Moreover patient was sent to neurologist who excluded any coexisting multi system neuropathies.

The minimum diagnostic criteria of AN are presence of Otoacoustic emission or Cochlear microphonic and absent or severely abnormal Auditory Brainstem response. Here, we found bilateral perceptive hearing loss with disproportionate speech discrimination together with normal OAE and absence of ABR. Hence, the diagnosis of AN was confirmed.

Discussion:

Auditory neuropathy (AN) is a poorly understood disease with unknown etiology which is characterized by abnormal neural function at the level of auditory nerve. Before

the invention of Otoacoustic emission (OAE), a group of patients were presented with absent ABRs who were actually found to have auditory function.^{4, 5} In mid 1980's after the invention of OAE, these group of patients were found to have normal cochlear function. This paradoxical findings between ABR and OAE were first defined as Auditory Neuropathy (AN) in 1996 by Arnold Starr.³

Overall, AN is quite rare. Most authors agreed that from 2-15% of infants with hearing loss may exhibit AN; that is one can expect to identify AN in approximately 1-3 infants per 10000 births.^{6, 7} AN has variable age of onset, with reports from birth to over 60 years of age. Case distribution appears equal among male and female, with no racial bias.⁸

Associated risk factors include neonatal anoxia or hypoxia, genetic hearing disorder, extreme prematurity, hyperbilirubinemia, hereditary sensory motor neuropathies, congenital brain abnormalities, low birth weight, family history of AN.^{2, 9} However, 25% patients have no associated risk factors.⁸ AN has been also reported in association with Charcot-Marrie-Tooth disease,¹⁰ Fredrich's ataxia,¹¹ Refsum's disease.¹²

Usually AN is bilateral but occasionally it can be unilateral too.² Patients always present with varying degree of hearing loss with extremely poor speech discrimination. In

order to confirm the diagnosis, following test battery should be recommended – Air and bone conduction audiometry, stapedial reflex threshold, speech discrimination test, OAE and ABR. Usually PTA result is variable ranging from normal to profound hearing loss. Speech discrimination is poor regardless of hearing threshold. SRT is usually absent. OAE is typically present and ABR is usually absent or severely abnormal in morphology. So, one must present with an abnormal ABR indicating neural dysfunction and presence of OAE indicating normal cochlear function at a minimum for diagnosis of AN.

In addition to the audiological work up, a neurological consult should be included to evaluate the presence of other system neuropathies.¹³ HRCT temporal bone should be done to rule out inner ear malformation in infants. MRI scan can be done to exclude any intracranial lesion.

There may be several potential sites of dysfunction for AN, although none are unanimously agreed upon. It can be an isolated inner hair cell dysfunction, or a synaptic dysfunction between the nerve and hair cell. The peripheral portion of auditory nerve could be demyelinating or can have an axonal neuropathy.^{14, 15}

Regarding the management of AN, a trial should be given with conventional amplification by hearing aid or by assistive listening device like FM (frequency modulation) system. Adequate benefit was reported in some cases but for many patients it was unsuccessful.^{16, 17}

Further research suggested that electrical stimulation can restore neural synchrony in case of demyelination,¹⁸ promote neural survival,¹⁹ and restore temporal coding.²⁰ On the basis of these findings, cochlear implant was introduced for AN in 2001²¹ with good number of positive outcomes in post operative case studies.

However, no tests are currently available to determine whether an individual with AN might benefit from hearing aid or cochlear implant. Most recently, if cochlear implant fails, the option of brainstem implantation has been reported.²²

Special care should be taken for the children, in whom diagnosis is very difficult. AN will not be detected by OAE only newborn screening protocol. So, combined use of OAE and ABR protocol should be considered for high risk newborns and children. In our country hearing screening protocol for new born is currently not available. As a result, it is near to impossible to diagnose AN in children. Rehabilitation programmes with an emphasis on lip reading or visual mode of communication (sign language, cued speech) are required to ensure appropriate development of language and communication skill of these children.

Conclusion:

AN is relatively a new classification of hearing disorder. Proper diagnosis is possible only by careful history and thorough audiological evaluation. Since it is an underdiagnosed condition in Bangladesh, otolaryngologists should be aware of this clinical entity. Suspected cases should be referred to the specialized centers where advanced audiological investigations and rehabilitation facilities are available.

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