Dr. Sivakumar. A, MBBS, DLO, DNB, Department of ENT participated and presented a paper during the 9th South Zone & 16th Tamilnadu State Annual Conference (ROCKENT2013). He also chaired the session on 4th October, 2013.
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AUDITORY NEUROPATHY WITH BILATERAL HAT EARS – A RARE CASE REPORT


ABSTRACT: Auditory neuropathy is a hearing disorder in which sound enters into the inner ear normally but the transmission of signals from the inner ear to auditory cortex is impaired due to the poor brain development. We report a case of 7 year male child presented with hard of hearing, deformed both external ears and inability to speak since birth. On examination bilateral hat ears, asymmetry auricles, hyperactivity, hyperactive attention deficit and from auditory brainstem response and otoacoustic emissions report it is diagnosed as auditory neuropathy and associated with various forms which is not included in ENT guidelines till date.

KEYWORDS: Auditory neuropathy, Bilateral hat ears, Hyperactive attention deficit.

INTRODUCTION: The finding of normal auditory function accompanied with abnormal brainstem response was defined as auditory neuropathy (AN) by Starr, et al. (1996) in 1996. Hearing disorder in which the transmission of signals from the inner ear to the brain is impaired. It may have normal hearing, or hearing loss ranging from mild to severe. The main cause is due to damage to the inner hair cells. The hallmark of auditory neuropathy is a negligible or very abnormal ABR reading together with a normal OAE reading. The treatment in Occupational Therapy, Hearing Aids, Frequency Modulation Systems, Lip Reading, and Auditory—verbal Therapy. We report a case of auditory neuropathy with associated features like bilateral hat ears, hyperactive attention deficit.

MATERIALS AND METHODS:

CASE REPORT: Patient Dhai was 7 year male case to the ENT outpatient department with the history of hard of hearing, deformed both ears, inability to speak since birth. Perinatal history was prolonged labour, neonatal hypoxic episode after delivery with prolonged assisted ventilation.

On examination bilateral hat ears, asymmetry auricles (Fig 2, 3), hyperactivity (Fig 4), hyperactive attention deficit. Plab audiometry shows bilateral sensorineural hearing loss (Fig 5). Brain stem evoked response shows both ears – no reproducible peaks could be obtained for click stimuli at 40 dbHL, with rate of 4/1. Cochlear microphonics was observed which recovers with change in polarity of stimulus suggesting of auditory neuropathy (Fig 6). Otoacoustic emission we recorded which was present in both ears (Fig 8, 9).
Computerized tomography shows normal study (Fig 10 to 15). Treatment occupational therapy for increasing attention, complete inside canal (CC) hearing communication, speech stimulation and speech education for language development, and auditory verbal therapy lip reading.

RESULTS: A child with mild hearing and inability to speak with negligible or very abnormal baseline evoked response audiogram with normal otoacoustic emission audiogram suggest auditory neuropathy. A normal otoacoustic emission audiogram in a sign that the outer hair cells are working normally (Fig 10).

DISCUSSION: Auditory neuropathy is a hearing disorder in which sound enters the inner ear normally but the transmission of signals from the inner ear to the brain is impaired. It can affect people of all ages, from infancy through adulthood. People with auditory neuropathy may have normal hearing, or hearing loss ranging from mild to severe; they rarely have poor speech perception ability, meaning they have trouble understanding speech clearly. Often, speech perception is worse than what would be predicted by the degree of hearing loss. The cause is mostly damage to the inner hair cells specialized sensory cells in the inner ear that transmit information about sounds through the auditory system to the brain. Other hair cells help amplify sound vibrations entering the inner ear from the middle ear. When hearing is working normally, the inner hair cells convert these vibrations into electrical signals that travel an nerve impulses to the brain, where the impulses are interpreted as sound. Some children who have been diagnosed with auditory neuropathy experienced certain health problems as newborns, during or shortly before birth. These problems include jaundice, premature birth, low birth weight, and an inadequate supply of oxygen to the unborn baby. The hallmarks of auditory neuropathy is a negligible or very abnormal ABR reading together with a normal OAE reading. A normal OAE reading is a sign that the outer hair cells are working normally.

For threshold testing an ABR test monitors brain wave activity in response to sound using electrodes that are placed on the person’s head and ears. An OAE test uses a small, very sensitive microphone inserted into the ear canal to measure the faint sounds produced by the outer hair cells in response to stimulation by a series of clicks. ABR and OAE testing is painless and can be used for newborn babies and infants as well as older children and adults. Treatment includes use occupational therapy for increasing attention, complete inside canal (CC) hearing aid trial for communication, speech stimulation and speech education for language development, sign language, auditory verbal therapy lip reading, frequency modulation (FM) systems are helpful. Cochlear implants and auditory brain implants are not useful effectively.

CASE REPORT

CONCLUSION: The child presenting with deaf mute and perinatal insult along with abnormal brainstem evoked response and normal otoacoustic emission suspect auditory neuropathy. The features of auditory neuropathy along with bilateral but exasperate attention deficits, and hyperactivity are recent clinical findings which are not reported in any ESN syndrome till date.

REFERENCES:

CASE REPORT

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