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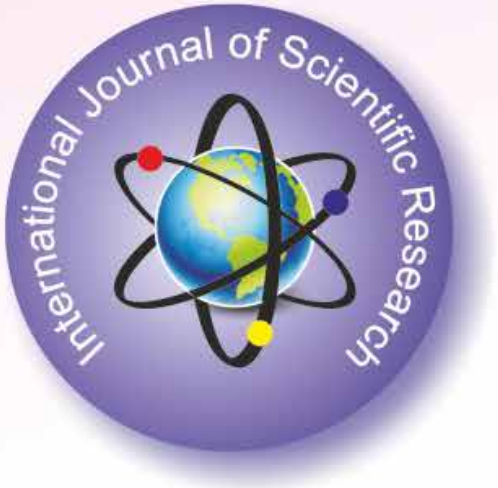

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Research Paper

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Can hormonal influence be a cause of auditory neuropathy

Medical Science

KEYWORDS : Jugular foramen,schwannoma,acoustic neuroma,

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ABSTRACT

Auditory neuropathy (AN) is described as a hearing disorder characterized by abnormal auditory nerve functioning in the presence of normal cochlear receptor hair cell activity. Common causes of AN include hyperbilirubinemia, low birth weight, anoxia in children. There is a sub group of AN who are reported to have normal hearing sensitivity till a particular age and following which they develop hearing loss which is not associated with any disease. In adults exact causes are not reported. Hormonal imbalance can be the one of the major cause in adults. Our clinical experience in individuals with AN also suggests that, most clients have been reported to acquire AN immediately, prior to or after their puberty.

Introduction

A hormone is a chemical substance secreted by the endocrine glands directly into the blood stream and regulates the function of a distant organ. Hormones can be divided into protein, polypeptide and steroid. Estrogen is a steroid hormone. The female sex hormone estrogen is a multi-tasker, contributing to roles as diverse as the differentiation and function of the reproductive track, memory storage and bone growth. Estrogen receptor (ER) is of two types, which are present in the inner ear of mice, rats and humans. In human 17 ER acts as an inhibitory agent and is a preservatory agent (Hultcrantz, Simonoska, & Stenberg 2006). Helfer(2001) mentioned that poor hearing seen in females more than men might be due to the hormonal differences. Turner's syndrome is an example to indicate the relation of estrogen and hearing which is characterised by low or absent estrogen production with hearing loss (Hederstierna ,Hultcrantz&Rosenhall 2009). Canlon and Frisina (2009) mentioned that the influence of sex hormones on auditory system starts in the peripheral and then moves to the central.

Auditory Neuropathy is a broad terminology which includes disorders that are characterized by grossly abnormal or absent Auditory Brainstem Response (ABR). The exact site of lesion and its pathophysiology is not yet completely understood. These individuals typically show reduced speech perception ability, especially in presence of background noise. Prevalence of this disorder in Indian context was retrospectively analysed by Kumar and Jayaram, 2006 this also reports that out of 11,205 individuals with Sensory Neural Hearing Loss, 61 were found to be associated with AN. This yields a prevalence of 0.54% or 1 in every 183 participants.Common causes of AN include hyperbilirubinemia, low birth weight, anoxia (Rance, Beer, Shepherd Dowell , King , Rickards&, Clark . 1999) and Otoferin mutation (Varga , Kelley , Keats 2006). There is a sub group of AN who are reported to have normal hearing sensitivity till a particular age and following which they develop hearing loss which is not associated with any disease condition. Kumar and Jayaram (2006) reported that 49 out of 61 individuals with AN self-reported on the onset of the disease between 11 years to 31 years of age. Two of the female participants also reported to have developed these symptoms following their first delivery. This disorder is twice the time in females compared to males (Kumar and Jayaram, 2006).Canlon et al (2009) reported that the hormonal changes can have an effect on the central nervous system (CNS). The interpeak interval and amplitude of postmenopausal females were longer, which indicate that hormonal changes may account for differences in neural conduction time. Animal study by CarasBrenowitz&Rubel.(2010) showed increased ABR thresholds and prolonged wave I latency in breeding conditioned white crowned sparrows which suggests that hormones act on auditory processing components presynaptic to the auditory nerves. Our clinical experience in individuals with AN also suggests that, most clients have been reported to acquire AN immediately, prior to or after their puberty.

Method

Three cases participated in the present study. The details of these cases were as follows.

Case 1:

A 15 year old female presented as an outpatient to the Kasturba hospital, Manipal came to the Department of ENT with the complaint of difficulty in hearing bilaterally. Otological examination revealed bilateral tympanic membrane intact and retracted. Patient reported to have presence of tinnitus bilaterally of continuous type since 5 months. The client was then referred to the Department of speech and hearing for detailed audiological evaluation. Detailed case history revealed the presenting complaint as difficulty in hearing bilaterally and difficulty in understanding speech since 5 months with onset as sudden followed by a roaring kind of tinnitus. She reported to have difficulty in understanding speech in various hearing situations such as while using telephone, while watching television and in noisy environment and also reported to have difficulty in localization. No significant medical history was reported by the client.

Case 2:

A 21 year old female presented as an outpatient to the Kasturba hospital, Manipal came to the Department of ENT with the complaint of difficulty in hearing since 2-3 years. Otological examination revealed bilateral tympanic membrane intact and normal. Patient reported to have tinnitus occasionally. The client was then referred to the department of speech and hearing for audiological evaluation. Detailed history revealed the presenting complaint as difficulty in hearing bilaterally and difficulty in understanding speech since 2-3 years. She reported to have difficulty in various hearing situation; while using telephone, while watching television and in noisy environment and also reported to have difficulty in following classroom instructions with no difficulty in localization. No significant medical history was reported by the client.

Case 3:

A 19 year old female presented as an outpatient to the Kasturba hospital, Manipal came to the Department of ENT with the complaint of difficulty. Otological examination revealed bilateral

tympanic membrane intact. Patient reported to have tinnitus of ringing type. The client was then referred to the department of speech and hearing for audiological evaluation. Detailed case history revealed presenting complaint to be difficulty in hearing bilaterally since childhood and difficulty in understanding speech. She reported to have difficulty in hearing in noisy situations. She was able to hear soft sounds but was not able to understand. She reported to have no difficulty in localization. No significant medical history was reported by the client.All three cases reported to have irregular menstrual cycle.

Procedure:
Detailed audiological evaluation was carried out for all the three cases. Initially routine audiological evaluation was done which included Pure Tone Audiometry, Speech audiometry and immitance. Later these patients were subjected for various eletrophysiological tests and Otoacosutic Emission (OAE)

Results:
All the patients gave highly inconsistent responses for PTA. . All the patients were not able to understand the instruction that was given for speech audiometry. SRT-PTA correlation couldn't be obtained. 0% score was obtained for SDT. Speech-in-Noise also gave 0% score in negative 10 dB SNR, 0 dB SNR, positive 10 dB SNR. Tympanometry results revealed normal middle ear functioning. Electrophysiological test including ABR and LLR was done. This shows absence of ABR peaks bilaterally at 90 dBnHL with no replicability and poor morphology. LLR was present for case 1 and case 2 and absence in case 3. Presence of OAE in all the three cases revealed normal outer hair cell functioning. (Table 1) .On the basis of all audiological evaluation the final impression given to all the three cases was Auditory Neuropathy. All the three cases were counseled regarding the hearing status and recommended for hearing aid trial.

None of the cases were found to be benefitting with the hearing aid therefore they were recommended for various communication strategies such as auditory visual mode, lip reading and gestural communication.(Table 2)

Discussion:
A hormone is a chemical substance secreted by the endocrine glands directly into the bloodstream.Estrogen is the main female sex hormone and plays an important role in hearing function. The influence of estrogen starts from peripheral and then moves on to the central. Animal studies indicate that estrogen can have an influence on auditory system. Increased ABR latency in postmenopausal stage is an indicative of neural conduction delay. Auditory Neuropathy is a blanket term used to describe a range of disorders affecting afferent neural activity in the peripheral and central auditory pathways as reported by Berlin et al in 2001 and Starr et al in 1996. Hederstierna et al in 2009 and Hultrantz et al in 2003 reported thatEstrogen influence in the auditory system leads to cochlear pathology. Also Canlon in 2009 reported that influence of sex hormones on auditory system starts in the peripheral and then moves to the central. The present case series report tries to correlate the hormonal effects on auditory neuropathy. The onset of the condition was reported to be 14 to 15 years of age by all the patients and they also reported of irregular menstrual cycle which was an indicative of hormonal imbalance. The absence of ABR and presence of

OAE in the present case series indicates auditory neuropathy. This shows that influence f hormonal imbalance can have a nerve conduction delay leading to the poor synchronous firing of the auditory nerve. Wharton et al in 1990 reported that increased latency in females is observed during the postmenopausal stage. Weiss et al in 2011 reported that hormonal changes have an effect on central nervous system during postmenopausal stage, therefore it can be assumed that it can have an efeect on perimenopausal stage too.

Conclusion:
From the current series of case report it can be postulated that hormonal imbalance can have a strong influence on auditory neuropathy. It highlights the importance of regular audiological evaluation in females, especially before and after puberty and delivery. Since there is no histopathological evidence to prove the estrogen level difference in these patients, further research to confirm the hypothesis put forward is warranted.

Table 1: Results of various Audiological evaluations

	CASE 1	CASE 2	CASE 3
	Rt Lt	Rt Lt	Rt Lt
PTA(dBHL)	26.6 36.6	41.6 48.3	56.6 50
SRT, SDS, SPIN			
Tympanometry	'A' 'A'	'A' 'A'	'A' 'A'
Reflexes	B/L absent	B/l absent	B/l absent
TEOAE	B/L present	B/L present	B/L present
ABR	B/L absent	B/L absent	B/L absent
LLR	B/L present	B/L present	B/L absent

Table II: Hearing aid trial was done and following were the responses.

	CASE 1	CASE 2	CASE 3
Type of hearing aid (B/L)	Siemens Infiniti Pro	Bernafone Win 102 Siemens Infiniti Pro Widex B2	Bernafone Win 102
Responses for questions	Unaided Aided 2/5 0/5	Unaided Aided 1/5 2/5	Unaided Aided 1/5 2/5
Response for spondees	Unaided Aided 2/5 2/5	Unaided Aided 1/5 2/5	Unaided Aided 1/5 2/5

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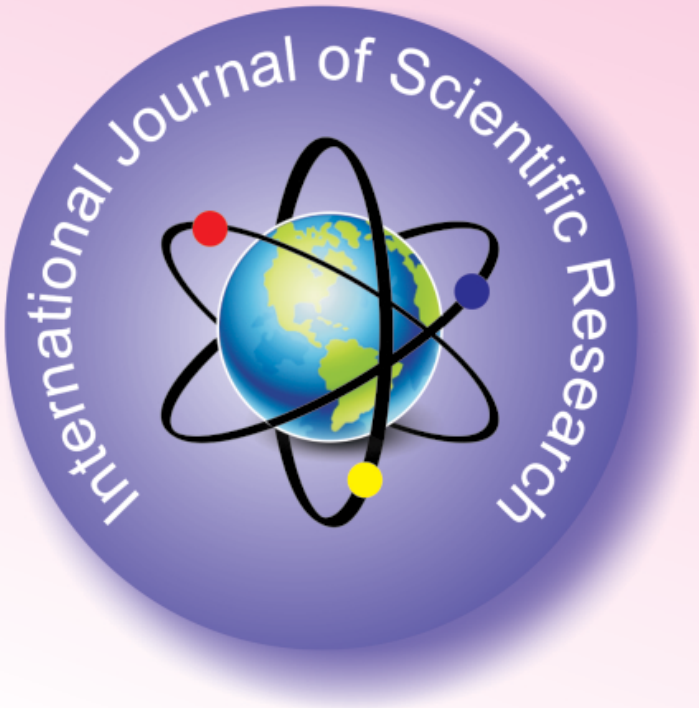

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