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Enlarged Vestibular Aqueduct- A Rare Case Report and Review of Literature

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Abstract

Background: Enlarged vestibular aqueduct is a one of the developmental abnormalities of inner ear and it is encountered in 1-12% of hearing impaired population. **Methods**: 15 year male presented to ENT OPD with decreased hearing in both ear since 6 years. Clinical examination, audiological and radiological investigations were done and diagnosis of bilateral enlarged vestibular aqueduct was made. **Conclusion:**The present case reports emphasizes importance of detailed radiological evaluation especially with CT scan and MRI in young patients presenting with sensorineural hearing loss on audiological evaluation.Patient was counselled and treated conservatively with hearing aids and also counselled about the requirement of Cochlear Implant in future.

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Keywords: Enlarged Vestibular Aqueduct, Sensorineural Hearing loss, Childhood

INTRODUCTION

The vestibular aqueduct, is a connection between the vestibule and the posterior cranial fossa, lies in temporal bone. It contains the endolymphatic duct, which terminates at the endolymphatic sac within the bony operculum. Large Vestibular Aqueduct (LVA) is an inner ear anomaly which is diagnosed with the temporal bone computed tomography (CT scan) and inner ear Magnetic Resonance Imaging (MRI). Non-syndromic sensorineural hearing loss (NSSHL) associated with LVA is of significant importance because the hearing loss associated with this condition may be fluctuating and sometime mixed loss, which normally starts in childhood and may also be associated with vestibular symptoms later.

Both auditory and vestibular symptoms may arise due to enlargement of the vestibular aqueduct. In 1971 Mondini first discovered enlarged Vestibular aqueduct while dissecting a temporal bone.[1] Meniere's like symptoms in enlarged vestibular aqueduct patient first by Valvasori in 1969.[2]Valvasori and Clemis[3] in 1978 found clinical relationship between enlarged vestibular aqueducts and hearing loss after study of enlarged vestibular aqueduct in 50 out of 3,700 tomograms (1.4%). Out of 50 cases, most had congenital hearing loss and many had vestibular symptoms. The size of enlarged vestibular aqueducts was in range of 1.5–8.0 mm in diameter, in their study. They considered a cut off of greater than 1.5 mm in anterior–posterior diameter as abnormally enlarged vestibular Aqueduct. The enlarged vestibular aqueduct remains the most common inner ear anomaly found on radiographic evaluation of children with hearing loss.[4] Enlarged vestibular aqueducts have also been associated with various other disease likeWaardenburg'ssyndrome,[5]branchio-oto-renal syndrome,[6]otofaciocervical syndrome,[7] and Noonan's syndrome.[8]

In children with sensorineural loss, various differential diagnosis should be kept in mind including congenital infectious diseases (cytomegalovirus, syphilis, intrauterine infections, and less frequently, rubella and toxoplasmosis) or acquired diseases (bacterial meningitis), ototoxicity, autoimmune diseases, traumatic, vascular and hereditary diseases. Bento et al,[9] in 2001 proposed the following protocol in the investigation of possible etiopathogenic factors which included: complete blood count, hemosedimentation rate, glycemic curve, and 5-hour insulin levels, serological tests for syphilis, toxoplasmosis, cytomegalovirus, rubella, measles, autoimmune disease factors (rheumatoid factor, C3, C4, reactive C protein, LE cell, antinucleus factor, ASLO, native anti-DNA, alpha 1 glucoprotein, and IgE dosage). Even after detailed investigations the etiology remains unknown till date.

CASE HISTORY

15 year old boy presented to ENT OPD with acute worsening of decreased hearing bilaterally since 1month, which was associated with headache, giddiness, vomiting and tinnitus. The child had a past history of decreased hearing bilaterally,

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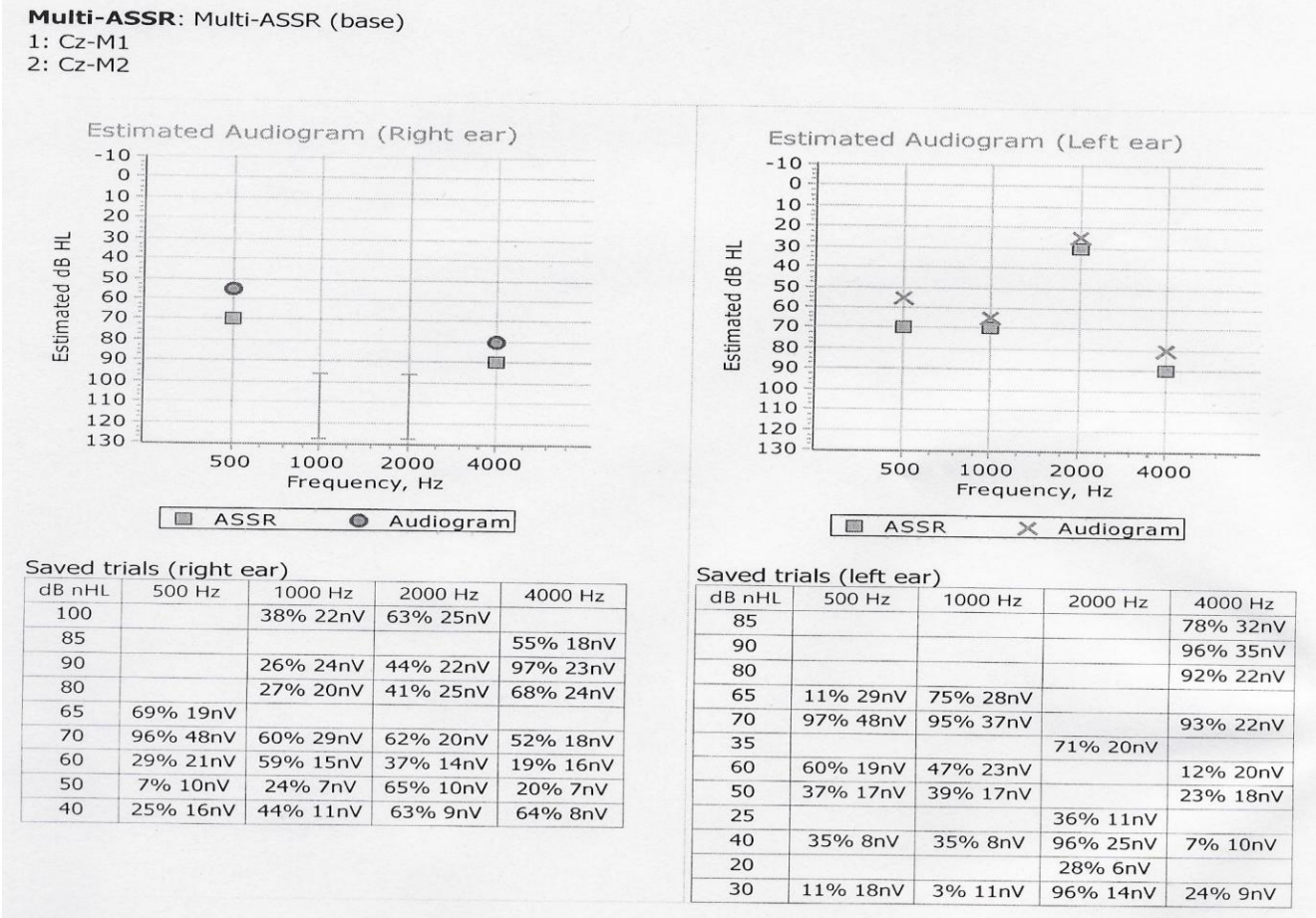
following a fall, since then he was on hearing aid. No recent history of head trauma, exposure to loud noise, ear discharge, fever, seizure, blurring of vision, prolonged use of ototoxic drugs. No history of similar complaint in the family. His mother had history of intermittent fever during the last trimester of antenatal period which was later diagnosed as pulmonary Tuberculosis in the postpartum period, for that she had taken Anti-tubercular therapy for 9 months and child was not breastfed till 9 months. There was no history suggestive of TORCH infection and Hypothyroidism in antenatal period. In the Postnatal period, child cried soon after birth, no NICU admission and received all the vaccination timely as per the National immunization schedule. Growth and development of the child was adequate for the age. On examination of ear bilateral tympanic membrane was found intact. Rinne's test showed bilateral sensorineural hearing loss (SNHL) and Weber's test lateralised to left and rest of the ear examination was normal. On examination of neck no sinus, fistula or any abnormal swelling. Complete blood count, liver and renal function test were normal. Pure tone audiometry showed Severe to profound SNHL in right ear and moderately severe SNHL in left ear. In Auditory steady state response test (ASSR), average hearing threshold of right ear was 83.75 dB and left ear was 56.25 dB (Fig 1).



Saved trials (right ear)

dB nHL	500 Hz	1000 Hz	2000 Hz	4000 Hz
100		38% 22nV	63% 25nV	
85				55% 18nV
90		26% 24nV	44% 22nV	97% 23nV
80		27% 20nV	41% 25nV	68% 24nV
65	69% 19nV			
70	96% 48nV	60% 29nV	62% 20nV	52% 18nV
60	29% 21nV	59% 15nV	37% 14nV	19% 16nV
50	7% 10nV	24% 7nV	65% 10nV	20% 7nV
40	25% 16nV	44% 11nV	63% 9nV	64% 8nV

Saved trials (left ear)

dB nHL	500 Hz	1000 Hz	2000 Hz	4000 Hz
85				78% 32nV
90				96% 35nV
80				92% 22nV
65	11% 29nV	75% 28nV		
70	97% 48nV	95% 37nV		93% 22nV
35			71% 20nV	
60	60% 19nV	47% 23nV		12% 20nV
50	37% 17nV	39% 17nV		23% 18nV
25			36% 11nV	
40	35% 8nV	35% 8nV	96% 25nV	7% 10nV
20			28% 6nV	
30	11% 18nV	3% 11nV	96% 14nV	24% 9nV

Fig 1: Showing Auditory steady state response (ASSR) of patient

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High resolution computed tomography (HRCT) temporal bone scan revealed bilateral dilated vestibular aqueduct measuring 3mm on right side and 3.7mm on left side with non-visualized cochlear modiolus and rest of the CT findings were normal. Thus the cause of patient's sensorineural hearing loss was found to be due to enlarged vestibular aqueduct which was clearly evident on HRCT temporal bone (Figure 2).

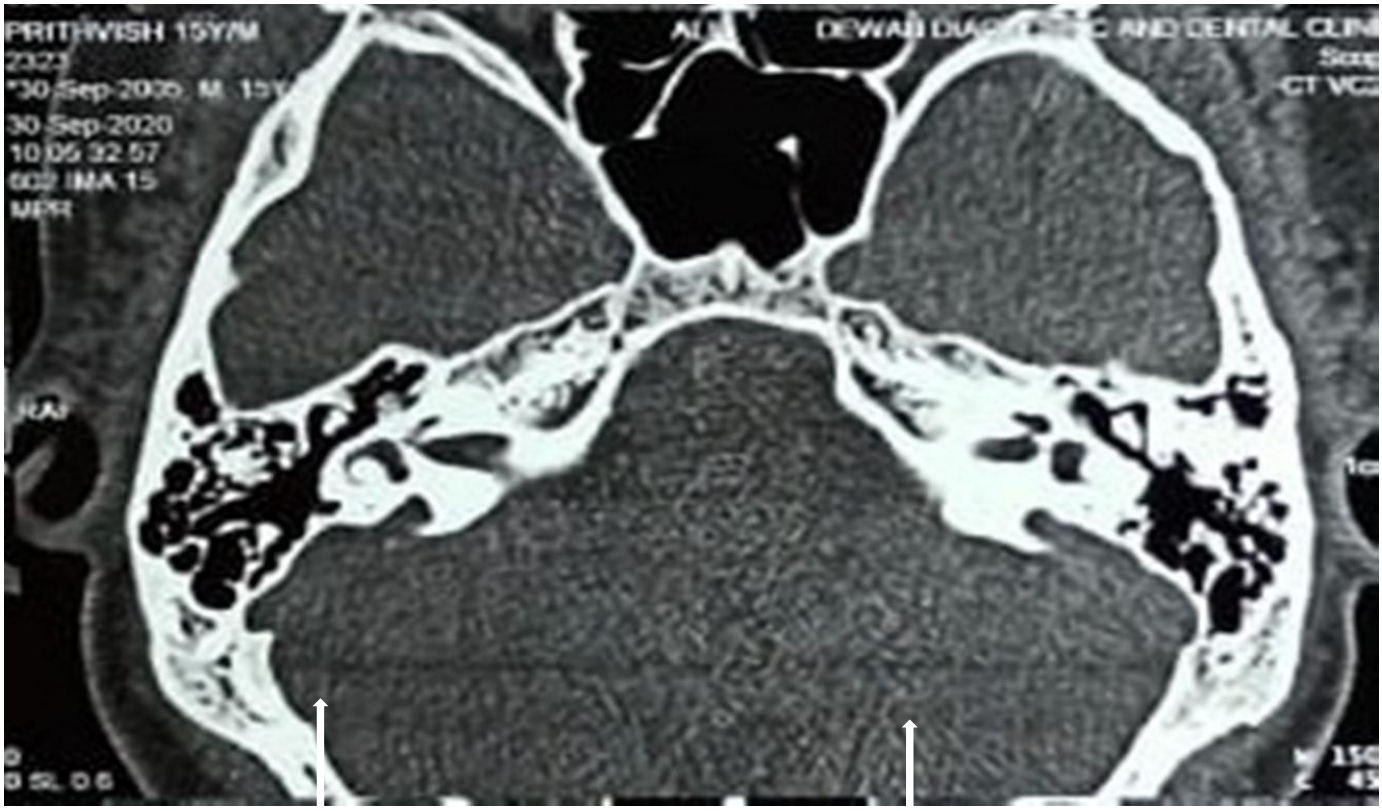

Fig 2: HRCT temporal bone showing bilateral Enlarged Vestibular Ducts

DISCUSSION

Vestibular aqueduct (VA) is a bony canal and it makes a communication between posterior-medial portion of the epidural space vestibule with the medium posterior fossa (Zazal, 1995).[10] Endolymphatic duct lies inside the vestibular aqueduct which connects the endolymphatic sac with the vestibule labyrinth, and through ductus reuniensit gets together with the cochlear duct. Vestibular aqueduct and endolymphatic sac are immature and small at the time of birth while other inner ear structures are well developed. As the child grows and posterior cranial fossa is expanded, Vestibular Aqueduct and endolymphatic sac also increase in size and reach maturity at about the age of 4 years.

Vestibular Aqueduct of size less than 1.5 mm and when not more than the diameter of the adjacent semicircular canal is, is considered normal. Enlarged vestibular aqueduct is characterized by enlargement associated with sensorineural hearing loss which may also be mixed with no defined cause. One of the many hypotheses considered are affection to homeostasis in endolymphatic circulation as a result of ductal enlargement, with

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consequent damage to cochlear neuroepithelium.[11] Another hypothesis is whether enlargement of Vestibular Aqueduct and endolymphatic sac would not be an isolated anomaly, but rather a continuity of the cochleovestibular malformation. Lemmerling and Antonelli[12] reported an association of defects in the modiolus of patients with Large Vestibular Aqueduct.

Antonelli classified VA enlargement into 5 grades. This measure is made from the intermediate point between the external opening and the common crura region.
GRADE I: lumen of the aqueduct is only visualized in the temporal bone cortex.
GRADE II: lumen of the aqueduct is visible close to common crura.
GRADE III: lumen of the aqueduct is larger than the common crura, but it is not visible in the topography of the vestibule output.
GRADE IV: the internal portion of the aqueduct is visible and its diameter in the output topography is smaller or equal to the common crura.
GRADE V: the internal portion of the aqueduct is visible and the diameter of the output topography is larger than the diameter of the common crura.

Children with LVAS may have moderate or severe hearing loss in childhood, but residual hearing with the help of conventional hearing aids allows the child to get adapted to some situations like developing spoken language and going to school.[13] Many of these children need support and rehabilitation in form of special schools with emphasis on sign language for communication.

Clinical presentation of large Vestibular Aqueduct is variable, normally its onset is at childhood and there is moderate to severe fluctuating or progressive loss; sometimes, it may also be sudden. Vertigo may be present and it is more common than it is believed. Although Abe et al[14] reported that one of the main characteristics of LVAS is fluctuating NSSHL, especially inhigh frequencies, there is no characteristic audiometric finding of LVAS and is a reason why we should be attentive in all cases of NSSHL and mixed loss in childhood. All such cases require detailed audiological and radiological evaluation.

In the present case report, patient presented with hearing loss since childhood noticed at about age of 9yrs, without any significant family history. Patient is using hearing aid since age of 9 years and recently noticed worsening of hearing even with hearing aid. Patient has developed a good language. Parents were also advised for a possibility of cochlear implant in future, if not benefited with hearing aid. Special instructions were given regarding prevention of head trauma so as to reduce the risk of further worsening of hearing loss by trauma.

CONCLUSION

LVAS is a clinical entity that should be kept in mind as differential diagnosis of sensorineural or mixed hearing loss on audiometry in all children presenting to an Otolaryngologist. Early diagnosis of large Vestibular Aqueduct or any other cause of hearing loss is essential because it allows more favourable rehabilitation treatment such as cochlear implant, in terms of speech and language development, and thus a better productive life.

Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent

for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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