CASE REPORT

Enlarged Vestibular Aqueduct Syndrome (Evas) - A Rare Disorder In Children

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Abstract

EVAS is considered to be rare disorder and its true prevalence is not known. MRI is considered to be the most appropriate diagnostic test. We report 5yr and 6yr old sisters who came to our OPD with complaints of not able to hear and not able to speak by birth.

Key Words

Enlarged vestibular aqueduct syndrome(EVAS)

Introduction

EVAS is a rare identity in children. Male to female ratio is found to be 1:1.5 (1). The term enlarged vestibular aqueduct is used when a vestibular aqueduct is larger than normal. Some studies suggest that the vestibular aqueduct is enlarged if it is more than 1.5mm in diameter while other define EVAS as being more than 4mm in size (2). It is a MRI finding, so MRI is considered to be the most appropriate test (2,3).

Case Report:

Two sisters at the age of 6yr and 5yr brought with chief complaints of not able to hear and speak by birth. There is no any other complaints of vertigo, unsteadiness, decreased vision. No any dysmorphic features and no history of trauma present. On general examination both the girls are active normal in appearance ,no any dysmorphic features are present. Systemic examination are within normal limits. BERA was done which shows moderate to severe SNHL present. From the clinical standpoint, SNHL is the unique presentation in EVAS among children (4) OAE-Absent in both ears.

MRI pictures of both sisters shows enlarged bilateral vestibular aqueduct and endolymphatic sac. Younger sister

Fig 1. MRI pictures shows enlarged bilateral vestibular aqueduct and endolymphatic sac. Younger sister 5yr having AP diameter of aqueduct is 3.3 mm on right side and 2.9 mm on left side

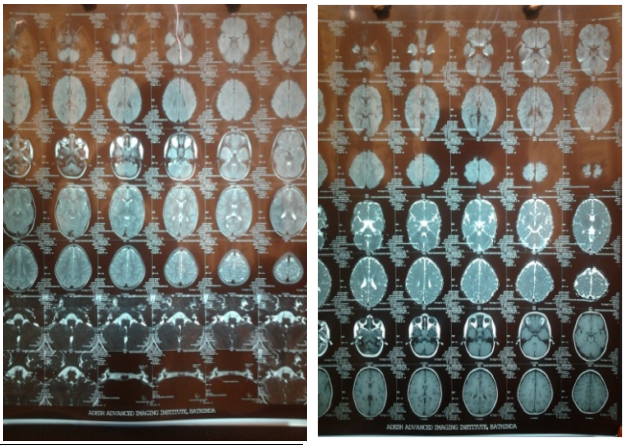

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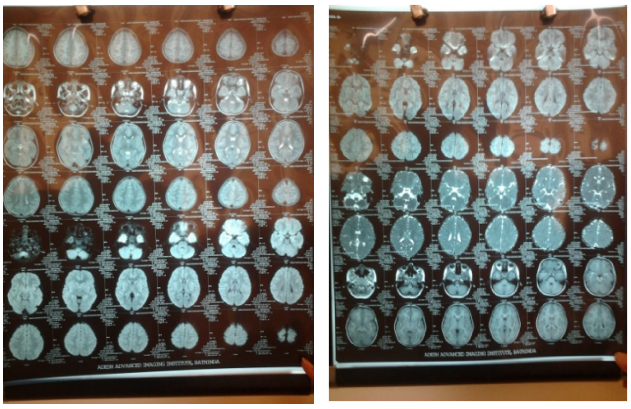
Fig 2. MRI pictures of both shows enlarged bilateral vestibular aqueduct and endolymphatic sac. Older sister 6yr having AP diameter of aqueduct 2.9 mm on right side and 4.6 mm on left side



5yr having AP diameter of aqueduct is 3.3 mm on right side and 2.9 mm on left side(*Fig 1*) Older sister 6yr having AP diameter of aqueduct 2.9 mm on right side and 4.6 mm on left side.(*Fig 2*)

Discusion

The present case is relevant because of the rarity of this disease in the children. It is a hearing loss that usually brings EVAS to the attention of a pediatrician. Such loss can be sensorineural, conductive or both. Clinical signs vary from hearing loss to vestibular symptoms like spinning vertigo, unsteadiness, feeling of vague instability.

In our case both sisters are not able to hear and speak by birth otherwise all general and systemic examination is within normal limits.

In addition to complete medical history and physical examination, to diagnose a case of EVAS, requires hearing testing and radiological evaluation. In children with SNHL incidence of radiological abnormalities has been reported to be as high as 39%. (5).

On Audiological testing bilateral sensorineural hearing loss was present.

There was no nystagmus and no other vestibular symptoms present. So vestibular system is considered to be normal.

For radiological assessment MRI was done which shows enlarged bilateral vestibular aqueduct and endolymphatic sac in both the sisters.

In such cases cochlear implant has significantly improved hearing (6).

References

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