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Case Report

Inner Ear Deformities: Case Report of Mondini's Type 2 Ear Deformity

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Abstract

This is a case report of two patient who reported to ENT OPD with parents complaining of difficulty in speaking and hearing, the patients were evaluated and diagnosed with Mondini's Type 2 ear deformity.

Keywords: Inner Ear; Cochlea; Mondini's; Deformity; Congenital Deafness; Hearing Loss; Audiology

Abbreviations

ENT: Ear Nose Throat; OPD: Outpatient Department; ICU: Neonatal Intensive Care Unit; BERA: Brainstem Evoked Response Audiometry; OAE: Otoacoustic Emission; ASSR: Auditory Steady State Response; HRCT: Hight Resolution Computed Tomography; MRI: Magnetic Resonance Imaging; CISS: Constructive Interference in Steady State

Introduction

Introduction should reflect the background, purpose and significant of the study that is carried out.

Congenital malformations of the inner ear usually occur in association with other anomalies in well-known hereditary syndromes, the affected individuals are often severely deaf from birth. The syndrome occur in consistent patterns of morphological deficit which can be classified according to the underlying anatomical, embryological or metabolic alteration. It is unusual to find congenital malformation of the inner ear of individuals who had no obvious hearing loss at birth [1]. However, it becomes equally important to identify them in children below 5 years of age when most of their developmental milestones are achieved. Especially the ones, having speech impairment as their parents give history of.

We wish to describe the anatomy of congenital ear anomalies in 4 ear of two different patients who had progressive bilateral sensorineural hearing loss acquired after birth and comment on anatomical and functional significance of these changes.

Materials and Methods

Two patients presented to ENT OPD at our hospital with parents providing history of inability to speak and listen with pre-lingual deafness. Of them was a 6-year-old female child who communicated to her family in sign language since birth with no history of intubation or NICU admission post birth, and the other was a 2-year-old male child who developed insidious loss of hearing followed by inability to speak 2 years after birth with no history of NICU admission.

Both the children were investigated where audiological reports suggested physiological dysfunction of the ear from moderate to severe hearing level using BERA, OAE, ASSR, Impedance audiometry. Both the patients were neurologically and clinically assessed by the pediatric neurology department to rule out any central or psychological cause of deafness and speech disability.

Radiologically, Mondini's inner ear deformity was identified by using HRCT Bilateral Temporal bone and MRI Brain with Inner ear with CISS protocol. The parents and relatives were counselled regarding the disease and for further management with Cochlear implant surgery proceeding pre-operative vaccination and fitness.

Conclusion: Identifying Inner ear deformity on an OPD basis with efficient history taking and sufficient investigations are evident to say that the prevalence of this condition might be under-

looked and needs more experience training to make sure the patient is treated on time, reducing the burden of hearing and speech disabilities and loss of functionality in society. The following investigations were used apart from detailed clinical history of deafness:

- BERA
- OAE
- ASSR
- HRCT Bilateral Temporal bone
- MRI Brain with inner ear with CISS protocol and oblique saggital cut of Internal Auditory canal.
- WHO classification for deafness

Results and Discussion

We found out that the patients were diagnosed with Mondini's type 2 congenital inner ear deformity leading to speech impairment.

Having diagnosed these deformities, it becomes a challenge to plan further management and counsel the parents of the children for hearing aid trials and cochlear implant surgery.

As surgeon's point of view, its challenging to insert the implant and assessing the placement of the electrode.

Conclusion

Based upon this report, it becomes more important to have an efficient history system for parents presenting with their children having speech disability and unable to hear and make sure they are timely investigated and managed without further delay ensuring better functionality in society and reducing the burden of having a morbidity with deafness.

Audiological assessment followed by radiological evaluation of inner ear anatomy should be done before considering for surgical intervention by cochlear implant and it must be thoroughly studied and planned accordingly.

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Conflict of Interest

No financial interest or any conflict of interest exists.

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