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Case Report
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Cochlear Apalsia and Cochlear Hypoplasia Incidental CT Finding A Case Report

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Abstract

The etiology of profound hearing loss in children is complex. Congenital inner ear malformation is an important cause of SNHL in children. It consists of labrynthine aplasia, cochlear aplasia, common cavity deformity, cystic cochleovestibular anomaly, cochlear hypoplasia and Modini's deformity. We report one patient who was born deaf and was being evaluated in our service for possible cochlear implantation. CT scan evaluation shows cochlear aplasia on one side and hypoplasia.

Keywords: Choclear Apalsia; Cochlear Hypoplasia; CT scan

Case Report

A 2 years old baby boy presented to our clinic for speech and hearing assessment. The baby is a product of full term spontaneous vaginal delivery with unremarkable pre, ante and post natal history, no family history of deafness nor concequanity, hi is the first child for a health 28and 30 years old parent. Mother started to notice his low response to sounds at age of 9 months but didn't seek medical advice until age of 17 months. No history of recurrent ear dishrags, snoring or mouth breathing, upon examination patient was continuously crying, hyperactive and running, no obvious dismorphic features, ear, nose and throat examination were normal . Neurological examinations were normal. Tympanogram bilaterally type A, Play audiometry shows no response, ABR also shows no response bilaterally. An I.Q testing show a score of 75 which is border line, also seen by psychologist and was diagnosed as DLD.

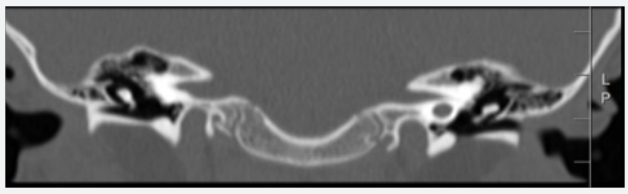

Figure 1.

Radiological examination computerized tomography (CT) (Figure 1) scan of temporal bone shows absent cochlea on the right side with large vestibule, with tiny rudimentary cochlea on the left side less than one fluid filled cochlear turn with normal appearing basal turn could not be found, and normal vestibule, lateral and posterior semicircular canals. With high jugular bulb in right side with no bony dehiscence, on both sides the internal acuastic met us was well developed and middle ear structures was normal (Figure 2). Magnetic resonance imaging (MRI) confirmed CT scan finding and also showed normal facial and auditory nerve bilaterally.

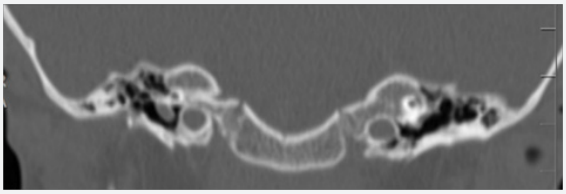

Figure 2.

Discussion

Temporal bone CT is a well-established technique for evaluation of pediatric hearing loss, generally best suited for evaluation of trauma and no acute congenital dysplasias of the otic capsule. For patients under consideration for cochlear implantation, the surgeon not only needs to identify congenital abnormalities (such as coexisting middle ear abnormalities, type of cochlear abnormality, pattern of cochlear ossification, and vestibular aqueduct abnormality) but also a variety of anatomic information, such as grade of mastoid bone aeration and internal auditory canal size and shape. The information helps in assessing the feasibility of cochlear implantation, side of implantation, modification of implantation technique, and anticipation of complications

To provide this information to the clinician, a high-resolution CT protocol needs to be applied. We perform direct axial-plane imaging through the temporal bone parallel to the hard palate using a bone algorithm, 512 × 512 matrix, and contiguous 0.625-mm sections. At our institution, direct coronal imaging is omitted for patients with sensor neural hearing loss to save radiation exposure. Instead, coronal reformatted images are generated. Congenital sensor neural hearing loss can be classified based on its aetiology. Broadly, causes can be divided into: non-genetic (including infective, metabolic, traumatic, and teratogenic) and genetic. Malformations of the outer and middle ear are predominantly unilateral (ca. 70-90%) and mostly involve the right ear. Inner ear malformations can be unilateral or bilateral. The incidence of ear malformations is approximately 1 in 3800 newborns in 15-20% of the cases there were bilateral malformations. According to Swartz and Faerber [1-8] the classification system of congenital cochlear malformations suggested by Jackler et al and Sennaroglu and Saatci which entails a correlation with the early stages of bony labyrinth embryogenesis is practical from the radiologist's perspective.

The classes include: labyrinthine aplasia, cochlear aplasia, common cavity deformity, cystic cochleovestibular anomaly (incomplete partition type I), cochlear hypoplasia and Mondini deformity (incomplete partition type II). Moreover, they depend on the timing of the insult, with respect to fetal development. The earlier it takes place, the more severe the hearing impairment. Cochlear aplasia results from the arrest of otic development at the late 3rd gestational week. It is characterised by absence of the cochlea, and variable development of vestibule and semicircular canals. On CT, the anterior bony labyrinth is hypoplastic, with absence of cochlear development or normal convexity of the cochlear promontory. The posterior labyrinthine structures are usually dysplastic, and the IAC may be normal, hypoplastic or dilated associated absence of the cochlear nerve is common. It differs from labyrinthine ossificans, which is an acquired condition in which the cochlea has been formed but the lateral wall of anterior labyrinth is flat (instead of having a convex in configuration). As for labyrinthine aplasia, cochlear aplasia is an absolute contraindication for cochlear implantation [9-12].

Cochlear hypoplasia is due to arrest during the sixth week results in a hypoplastic cochlea consisting of a single turn or less. This deformity comprises approximately 15% of all cochlear anomalies. Radio graphically; a small bud of variable length (usually 1 to 3 mm) protrudes from the vestibule. The vestibule is frequently enlarged with accompanying semicircular malformations in about half of faces. Small cochlear lacking a modiolus or other internal architecture has been described histologically. Hearing is variable in these ears and may be remarkably good considering the minute size of the cochlea. The variability of hearing presumably is accounted for by degree of membranous labyrinthine development within the truncated cochlear lumen.

Conclusion

Cholcear aplasia is an absolute contraindication for cochlear implant. While cochlear hypoplasia, although it limits full insertion of the implant electrodes, still will yield benefit to the patient because of the residual auditory cells present. Precise radiologic information also affects surgical planning. Surprisingly in our patient ct scan shows right cochlear aplasia and left cochlear hypoplasia which was found during pre operative evaluation by radiology for cochlear implant, and decision was made no to implant due to absolute contraindication in the right side as absent cochlea and a relative contraindication with high risk of failure in the left ear due to small rudimentary cochlea.

Teaching Point

High-resolution CT plays a vital role in the diagnosis of congenital sensor neural hearing loss and helps with surgical decision making and planning whether cochlear implant is indicated or not and estimating the successful rate.

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