Inner Ear Anomalies Causing Congenital Sensorineural Hearing Loss: CT and MR Imaging Findings

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Many congenital dysplasias of the osseous labyrinth have been identified, and the differential diagnosis of these dysplasias is essential for delivering proper patient management. We retrospectively reviewed the computed tomography (CT) and magnetic resonance (MR) imaging findings of 20 children who had congenital sensorineural hearing loss. The children included cases of enlarged vestibular aqueduct and endolymphatic sac \( n = 8 \), aplasia of the semicircular canal \( n = 4 \), lateral semicircular canal-vestibule dysplasia \( n = 3 \), common cavity malformations with a large vestibule \( n = 1 \), cochlear hypoplasia \( n = 1 \), Mondini’s dysplasia with large vestibular aqueduct \( n = 1 \), Mondini’s dysplasia with a large vestibule \( n = 1 \), and small internal auditory canal \( n = 1 \). Six cases were unilateral. Nine cases had combined deformities, and nine cases had cochlear implants. CT was performed with a 1.0-mm thickness in the direct coronal and axial sections with using bone algorithms. MR was performed with a temporal 3D T2 FSE 10-mm scan and with routine brain images. We describe here the imaging features for the anomalies of the inner ear in patients suffering from congenital sensorineural hearing loss.

Index words: Ear abnormalities
Computed tomography (CT)
Magnetic resonance imaging (MR)

Congenital malformations of the inner ear can affect any portion of the membranous or the bony labyrinth. A primary or secondary deficiency of the cochlear nerve may also present as a congenital anomaly. CT and MRI allow for a detailed analysis of each lesion and they can provide guidance for the proper treatment of each patient. Both CT and MR imaging can be used to detect inner ear malformations, yet the two techniques are often complementary. CT is preferred in those cases where associated middle or external ear malformations must be excluded. MRI is preferred in the case of evaluating subtle changes in the membranous labyrinth or where there are abnormalities of the nerves in the internal auditory canal [1- 5].

We describe here the CT and MRI findings for a variety of deformities of the osseous labyrinth involving the cochlea, vestibule, semicircular canal, internal auditory canal, and vestibular aqueduct in patients suffering from congenital sensorineural hearing loss.

Embryology

Most temporal bone anomalies are caused by the pre-
mature arrest of development or by the complete failure of formation [1, 6]. Congenital malformations tend to involve either the inner ear or the middle and external ear. In some craniofacial syndromes or chromosomal disorders, anomalies of both regions do occasionally coincide. Embryopathies that are due to toxin exposure may also result in combined anomalies [1, 2, 4, 7-9].

The inner ear develops between the 4th week and 8th week of gestation, it grows between the 8th and 16th weeks and it ossifies between 16th and 24th weeks. Malformation of the inner ear results from arrest during the various stages of embryogenesis. The cochlear duct develops first and cochlear malformations range from complete aplasia, which generally develops at the 3rd week, to incomplete partition or a cochlea with incomplete or no interscalar septa, and this generally develops at the 7th week. Cochlear development is complete at the 8th week. The saccule, endolymphatic duct and utricle are completed at 11 weeks and the semicircular canals (SCC) develop between the 19th and 22nd weeks. The lateral SCC or duct is the last to form and thus, it is most frequently affected [6-9].

**Michel’s Anomaly**

Michel’s anomaly, complete labyrinthine aplasia, is caused by early arrest of differentiation of the otic placode during the 3rd gestational week [6]. The diagnosis can be made when the inner ear structures are absent.

Labyrinthitis ossificans [Fig. 1A, B] may have a similar appearance, but for this acquired disorder, the general shape of the formed labyrinth is preserved, and the medial wall of the middle ear bulges over the lateral SCC and the cochlear promontory. In labyrinthine aplasia, the middle ear medial wall is flat and featureless.

**Common Cavity Malformation**

Developmental arrest between the 4th and 5th gestational weeks results in a single labyrinthine structure that is called a common cavity. This is seen as a single fluid-filled space in the bony otic capsule, and it is without the internal cochlear or vestibular differentiation (Fig. 2A-C). The SCC or vestibule can be normal or malformed. In these patients, the internal auditory canal (IAC) can be recognized, unlike the patients with complete labyrinthine aplasia. One fourth of all cochlear malformations are common cavity malformations.

**Cochlear Aplasia or Hypoplasia**

Arrest of cochlear development during the 5-6th weeks results in cochlear aplasia and hypoplasia [6]. In cochlear aplasia, the cochlea is seen as a single cavity without separation of the different turns and the cochlea is also without distinguishable scala vestibuli/scala tympani. The vestibule and SCC can be normal, although they are most often malformed or only their remnants are seen. One turn or less is generally noted in cochlear hypoplasia [Fig. 3A]. Cochlear hypoplasia represents 15% of all cochlear malformations. These malformations are often found in patients with brachio-oto-renal syndrome. Cochlear aplasia and hypoplasia display as a dysmorphic cochlear cavity with no discrete internal cochlear architecture, and there are often coexisting vestibular anomalies [Fig. 3B]. Dysplastic changes of the vestibule may be seen as a bulbous enlargement.
Incomplete partition of the cochlea (Mondini malformation)

Incomplete partition of the cochlea (Mondini malformation) occurs when cochlear development is arrested during the 7th week of gestation. This malformation accounts for 55% of all cochlear malformations. In these patients, the cochlea has only 1.5 turns, and often there is preservation of the basilar turn while the interscalar septum and osseous spiral lamina are absent (Fig. 4A, B). Other features of mild dysplasia include a deficient modiolus and an incomplete interscalar septum along with the asymmetric or undivided scalar chambers (4).

The interscalar septal defect and the absence of the osseous spiral lamina of the middle ear and the apical turns can best be demonstrated on heavily T2-weighted gradient-echo images or on the fast spin echo images. Incomplete partition is often associated with a large endolymphatic duct (ELD) and the sac and vestibular aqueduct.

Overall, the vestibule, SCC and ELD/sac malformations are found in 20% of these patients (Fig. 5A, B). Due to the variable development of the sensory neuroepithelium, there are various degrees of sensorineural hearing loss noted for these patients.

Vestibular Dysplasia

The saccule and utricle are completely formed at the 11th week of gestation. Isolated malformations of the vestibule are rare and these malformations are most often associated with other inner ear malformations such as SCC anomalies or cochlear anomalies (Fig. 2). The SCC can be partially or completely assimilated in the vestibule, and the vestibule can be enlarged and irregularly shaped. There appears to be a reciprocal relationship between the size of the SCC, particularly the lateral

Fig. 2. An 8-year-old boy with a left common cavity malformation.
A. (axial scan) and B (coronal CT scan). A single cavity (arrows) is found in the inner ear, and this represents the cochlea and the vestibule. This cavity is called a “common cavity”. The IAC is visible.
C. Axial 10-mm-thick 3 D FSE T2-weighted image at the level of the cochlea (arrows).

Fig. 3. A 6-year-old boy with right cochlear hypoplasia. The axial 10-mm-thick T2-weighted 3 D FSE image shows only a small basal turn (arrow) of the right cochlea.
B. A short, broad lateral semicircular canal is confluent with the vestibule, and together, they form a single fluid-filled cavity (arrows).
canal, and the size of the vestibule. An enlarged vestibule is associated with an underdeveloped lateral SCC.

This malformation is the second most commonly observed isolated deformity (Fig. 6A, B). In its mildest form, the lateral SCC, as a whole, is smaller than normal and the vestibule is slightly larger than normal. In its more severe forms, the vestibule extends further into the lateral and superior aspects of the otic capsule. The internal diameter of the vestibule is measured at the level of the lateral SCC and the width and length measurements are made at the mid-portion. The vestibule is considered abnormally enlarged if the width is >3 mm and length is >6 mm (4).

**Semicircular canals/ducts**

A malformation of a SCC that’s associated with a normal cochlea is most likely to be due to a problem occurring between the 7th and 22nd week of gestation. An open pouch persists in the most common SCC anomaly, and lesser degrees of these SCC anomalies are represented by segmental dilatation on parts of the canal. The lateral SCC develops last and is the most likely to be abnormal, and the superior SCC is the least likely to be malformed. Dilatation of the lateral SCC is often accompanied by dilatation of the vestibule (Fig. 6), and this is occasionally detected as an incidental finding. Isolate aplasia of the posterior SCC has been described in patients with Waardenburg and Alagille’s syndromes (10). The diagnosis of ‘CHARGE’ syndrome (abnormalities of the coloboma and heart, choanal atresia, retarded growth, and abnormalities of the genitals and ear) is nearly cer-
tain when aplasia of all the SCCs/ducts is found (Fig. 7A, B).

**A large endolymphatic duct and sac (LEDS)**

A large endolymphatic duct and sac (LEDS) constitutes enlargement of the endolymphatic sac and duct and there is corresponding enlargement of the bony vestibular aqueduct (VA) (Fig. 8) (8, 12). The VA or endolymphatic duct are considered enlarged when their diameters exceed that of the posterior SCC or when they are more than 1.5 mm in diameter, as measured halfway between the common crus and the external aperture of the VA (7).

The diameter of the VA in normal temporal bones is 0.4-1.0 mm. LEDS is the most commonly identified radiographic anomaly of the inner ear (6, 13, 14). It has been found that 1-1.5% of the cases referred for tomography of the inner ear structures have an abnormally large VA. Patients typically present with a progressive, severe sensorineural hearing loss in childhood or early adulthood, and this condition is often exacerbated by minor trauma (13, 15). The majority of patients have a mixed sensorineural/conductive hearing loss. Patients with a LEDS have an associated cochlear malformation in 76% of cases (Fig. 5) and there is vestibular malformation in 40% of cases (5, 11). Vertigo is present in 30% of cases. Any correlation between the severity of the radiological abnormality and the associated clinical symptoms is unclear. The mechanism by which LEDS causes progressive SNHL is still unknown.

Several possible mechanisms of cochlear damage have been proposed, and these include injury caused by pressure effects, physiologic endolymphatic dysfunction and susceptibility to minor trauma (5, 13). The high prevalence of coexisting cochlear anomalies is probably a manifestation of a common underlying embryologic insult.

**Small Internal Auditory Canal**

The internal auditory canal (IAC) is usually normal, but it may be large or small in association with the inner ear anomalies. Aplasia or severe narrowing of the IAC is associated with sensorineural hearing loss (Fig. 9). An enlarged IAC, due to dural ectasia, has been described in neurofibromatosis.

Congenital stenosis of the IAC may exist as an isolated condition or it is noted along with a number of other osseous anomalies of the temporal bone. Isolated narrowing of the IAC implies that there is an otherwise radiographically normal otic capsule and the absence of any osseous condition predisposing the patient towards acquired stenosis (5). A history of significant head trauma...
should also be excluded. The development of the IAC is separate from that of the labyrinth.

The normal range of IAC diameters has long been quoted as being 2–8 mm with an average of 4 mm, and these numbers are based on Valvassori’s series. An IAC smaller than 2 mm is considered stenotic. MRI is potentially sensitive enough to demonstrate the 4 distinct nerve bundles in the IAC (5). Aplasia of the cochlear nerve is manifested as a loss of the normal round filling defect in the antero-inferior IAC. Due to the concomitant embryologic development of the IAC and the nerves, congenital aplasia is also accompanied by a stenotic IAC. A stenotic IAC is often associated with the absence of the vestibulocochlear nerve, and this is a contraindication for cochlear implantation (5, 9, 16).

Three types of aplasia or hypoplasia can be distinguished (5). A stenotic IAC with the absence of the VIIIth cranial nerve is a ‘type 1’ malformation. In ‘type 2’ malformations, a common vestibulocochlear nerve is found with hypoplasia or aplasia of its cochlear branch. When this is associated with other inner ear malformations, it is called a ‘type 2A’ malformation; it is called a ‘type 2B’ malformation when the inner ear is normal. In these patients, the facial nerve often has an aberrant course.

Conclusion

Inner ear anomalies are a rare, but they are important because they result in sensorineural hearing loss. The inner ear dysplasias are best evaluated with a combination of thin section CT to observe the bony labyrinth, and heavily T2-weighted gradient-echo or fast spin-echo 3D T2-weighted MRI are the best choices to observe the membranous labyrinth or any abnormalities of the nerves in the internal auditory canal.

References

CT와 MR의 비교: CT와 MR의 비교

1. 공통 캐비티
2. CT와 MR의 비교

공통 캐비티의 크기와 모양을 비교하고, CT와 MR의 각 특성을 분석한 결과, CT와 MR의 응용 범위와 결과는 유사하였다. CT의 경우, 공통 캐비티의 크기와 모양을 정확하게 분석할 수 있었고, MR의 경우, 공통 캐비티의 모양과 심리적 특성을 분석할 수 있었다. 이 결과는 공통 캐비티의 특성과 CT 및 MR의 응용 범위를 비교함에 있어 유용한 결과를 제공하였다.