A Narrow Internal Auditory Canal with Duplication in a Patient with Congenital Sensorineural Hearing Loss

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A narrow internal auditory canal (IAC) with duplication is a rare anomaly of the temporal bone. It is associated with congenital sensorineural hearing loss. Aplasia or hypoplasia of the vestibulocochlear nerve may cause the hearing loss. We present an unusual case of an isolated narrow IAC with duplication that was detected by a CT scan. In this case, the IAC was divided by a bony septum into an empty stenotic inferoposterior portion and a large anterosuperior portion containing the facial nerve that was clearly delineated on MRI.

Index terms:
Ear, abnormalities
Ear anatomy
Ear, MR
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A narrow internal auditory canal (IAC) with duplication is a rare congenital disorder that is usually associated with other inner ear, middle or external ear abnormalities (1). A narrow IAC can be diagnosed when the diameter of IAC is less than 2 mm as seen on high-resolution CT (HRCT) of the temporal bone (2). This malformation comprises 12% of congenital temporal bone anomalies (3, 4). It has been thought that this anomaly is caused by aplasia or hypoplasia of the vestibulocochlear nerve that results in ipsilateral congenital sensorineural hearing loss (1, 2, 5).

Although HRCT is a common technique that is used to evaluate the inner ear and related structures in patients with hearing loss, it has only been able to demonstrate the presence of bony abnormalities in approximately 20% of patients with congenital sensorineural hearing loss (3, 4). However, technological advances in magnetic resonance (MR) imaging have made it possible to evaluate the fine neural structures along the course of the vestibulocochlear nerve of the IAC in patients with congenital sensorineural hearing loss.

To date, only five cases of narrow IAC with duplication have been reported in the literature (2, 5–8). We report an unusual case of an isolated narrow IAC with duplication with a relatively wide anterosuperior portion containing an intact facial nerve and an inferoposterior empty stenotic portion as evaluated by MRI.

CASE REPORT

A 6-year-old girl presented with unilateral hearing loss. At the physical examination, the otologic status and head and neck status were normal. The facial nerve function was unimpaired and symmetric. The child did not present any vestibular symptoms or tinnitus. No symptoms of ear fullness or pressure were reported. There was no family history of hereditary sensorineural hearing loss. The patient had never had mumps or head trauma. There was no evidence of syndromic deafness stigmas.

Pure tone audiometry and speech testing showed total deafness of the right side, but normal hearing thresholds in the left side. An examination with brain stem electric

response audiometry (BERA) showed no response from the right ear. A reduced right vestibular response was found by a caloric test.

Axial and coronal reformatted HRCT images of the temporal bones were obtained. For further evaluation of the seventh and eighth cranial nerves, high-resolution MR imaging was performed using the 3.0-T Achieva system (Philips Medical Systems, Best, The Netherlands). MR imaging protocols included the following: a three-dimensional driven equilibrium radio frequency pulse (3D DRIVE) sequence (TR/TE 2000/200, 1.2 mm slice thickness, matrix 336 × 336, field of view 200 × 200 mm, number of acquisitions 1) and a T2-weighted fast spin-echo (FSE) sequence (TR/TE 2555/80, 2 mm slice thickness, matrix 400 × 313, field of view 130 × 130, number of acquisitions 2) on axial and parasagittal planes. Parasagittal MR imaging was obtained perpendicular to the course of the acoustic nerve for the IAC.

As seen on temporal bone HRCT, there were two narrow bony canals in the IAC of the right temporal bone. The IAC was divided by a bony septum into a relatively large anterosuperior portion (1.6 mm) and a stenotic inferoposterior portion (0.8 mm) (Figs. 1A–C). The anterosuperior portion ended in a wide connection in the facial canal and a narrow connection to the vestibule. The inferoposterior portion ended in narrow connections to the cochlea and vestibule. The two canaliculi partially joined at the lateral end of the right IAC. The facial nerve canal was intact along its course, except for a slightly widened labyrinthine segment. There were no abnormalities in the

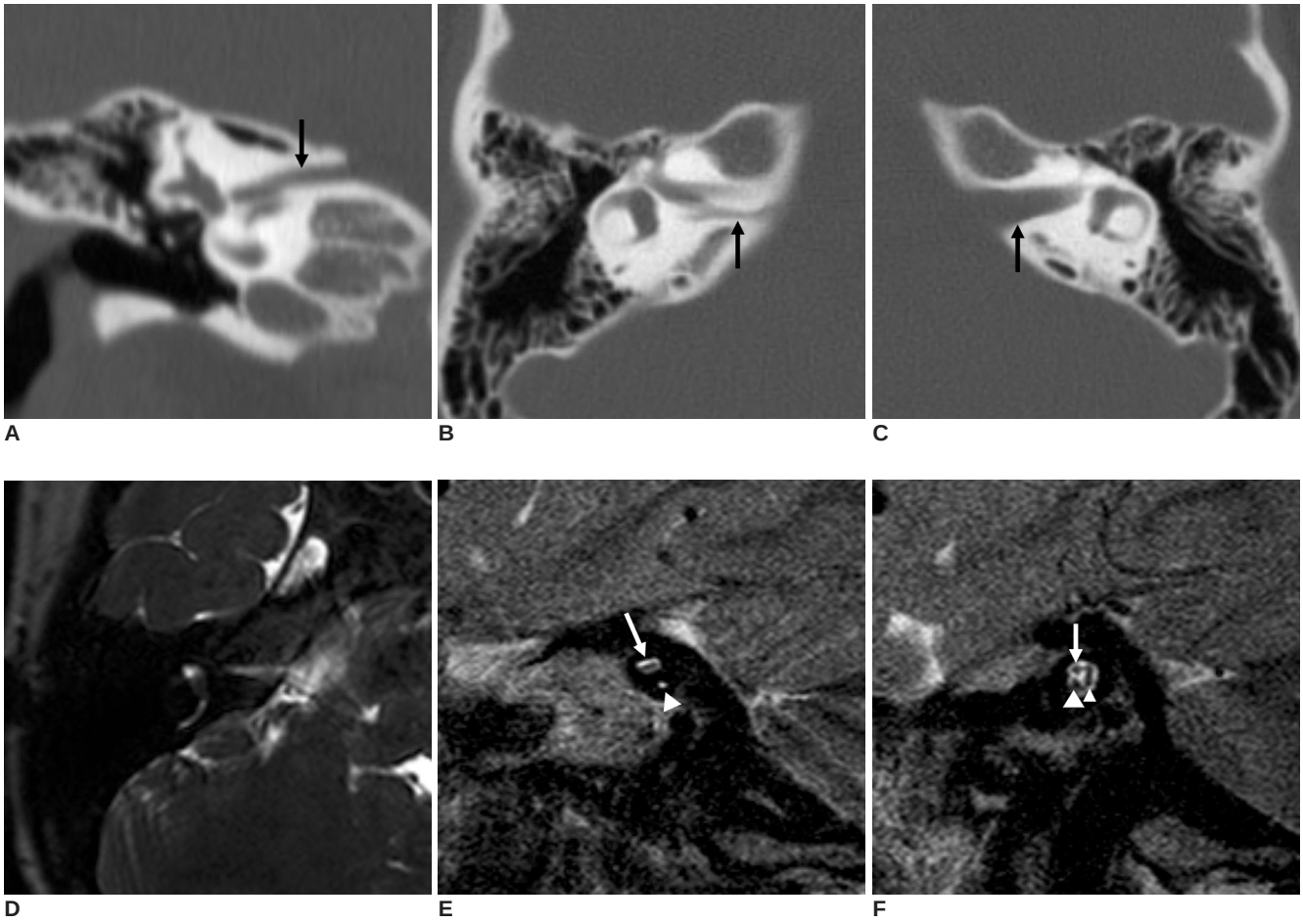

Fig. 1. Narrow internal auditory canal with duplication in 6-year-old girl.
A. Coronal CT image shows narrow internal auditory canal with duplication divided by bony septum into relatively large anterosuperior (arrow) and stenotic inferoposterior portions.
B. Axial CT image shows same findings. Inferoposterior portion (arrow) of internal auditory canal is smaller than anterosuperior portion.
C. Axial CT image shows normal contralateral internal auditory canal (arrow).
D. 3D-DRIVE T2-weighted axial image of right temporal bone shows narrow internal auditory canal with duplication.
E. T2-weighted fast spin echo parasagittal image of right internal auditory canal demonstrates facial nerve that fills upper canal (arrow), but vestibulocochlear nerve is not seen in lower canal (arrowhead).
F. T2-weighted fast spin echo parasagittal image of left internal auditory canal demonstrates normal facial nerve (arrow), common vestibular nerve (small arrowhead), and cochlear nerve (large arrowhead).

cochlea, vestibule, and semicircular canals as seen on HRCT.

As seen on MR imaging, the inner ear structures were normal, and the vestibulocochlear and facial nerves in the IAC were clearly identified on the left side. On the right side, the cochlea, the vestibule, and the semicircular canals were normal. However, there was no identifiable neural structure in the inferoposterior compartment of the right duplicated IAC. There was only one nerve structure, assumed to be the facial nerve, on the anterosuperior compartment of the duplicated IAC (Figs. 1D–F).

DISCUSSION

A narrow IAC is a rare congenital malformation of the temporal bone and it usually exists along with several other abnormalities of the temporal bone and various systemic developmental anomalies, including cardiac, renal, skeletal, and intestinal malformations (1). This malformation comprises only 12% of congenital temporal bone anomalies, and it is usually unilateral (3, 4, 9). Rarely, this malformation may exist in isolation. Congenital isolated narrowing of the IAC implies that there are no associated abnormalities in the inner ear, middle or external ear structures, and the absence of any acquired osseous condition predisposing to stenosis of the IAC (10). In this case, the IAC was unilateral and there was no evidence of other temporal bone or systemic developmental anomalies.

There are two widely accepted hypotheses that explain the association of a narrow IAC with sensorineural hearing loss. One hypothesis is that the embryonic cochlear and vestibule induce the growth of the vestibulocochlear nerve and the bony canal develops around the nerve together with the facial nerve by chondrification and ossification of the mesoderm in the eighth week of gestation. When the vestibulocochlear nerve is aplastic or hypoplastic, the IAC fails to develop and becomes stenotic (1, 4). The other hypothesis is that the primary defect is bony stenosis that inhibits the growth of the vestibulocochlear nerve and causes impaired transmission of an induction signal from the intact cochlea and vestibule. However, given the fact that the facial nerve function is usually preserved and it has a normal gross morphology in most cases of narrow IAC, this hypothesis is less likely (1). The facial nerve develops separately and it is later surrounded as the canal forms around the vestibulocochlear nerve. This may cause the duplication of the IAC in some cases of a narrow IAC with the aplastic or hypoplastic vestibulocochlear nerve. In this case, the right IAC was duplicated with a bony septum that separated the canal into a relatively large anterosuperior portion containing an intact facial nerve and empty stenotic inferoposterior portion.

To date, only five cases of a narrow IAC with duplication have been reported. The first case was reported by Casselman et al. (6) in 1997. These investigators described a case of unilateral narrow IAC with duplication. However, there were no imaging findings of this case in the report. Vilain et al. (8) reported a second case in 1999. These investigators described a case of narrow IAC with duplicated canals that joined at the fundus of the IAC. Cho et al. (5) reported a third case in 2000 and it was a case of narrow IAC with duplication. However, two separate canaliculi of the IAC showed similar diameters and did not join at the fundus of IAC. A fourth case was reported by Ferreira et al. (2) in 2003. These investigators described a similar case of narrow IAC which was associated with bilateral enlargement of the vestibule and the lateral semicircular canal and bilateral dysplastic cochleae. A fifth case was reported by Demir et al. (7) in 2005. This was a case of a narrow IAC that was separated into two narrow bony canals. The contralateral external auditory canal was stenotic and the ossicles were dysplastic. Our case was an isolated unilateral narrow IAC that was not associated with any other labyrinthine abnormality. In this case, the IAC was divided by a bony septum into two bony canals that

Table 1. Reported Cases of Narrow Internal Auditory Canal with Duplication

References	Facial Nerve	Vestibulo-Cochlear Nerve	Vestibular Function	Inner Ear Abnormality	Middle or External Ear Abnormality	MRI Sequences
Casselman et al. 1997	+	–	?	–	+	3D FT-CISS
Vilain et al. 1999	+	+	Abnormal	–	–	3D FT-CISS
Cho et al. 2000	+	–	Abnormal	–	–	3D FSE
Ferreira et al. 2003	+	–	Abnormal	+	–	3D FT-CISS
Demir et al. 2005	+	–	Normal	–	+	3D B-FFE
Present case	+	–	Abnormal	–	–	3D DRIVE

Note.—3D FT-CISS = three dimensional Fourier transformation-constructive interference in steady state, 3D FSE = three dimensional fast spin echo, 3D B-FFE = three dimensional balanced fast field echo, 3D DRIVE = three dimensional driven equilibrium radio frequency pulse, + = presence, – = absence

were joined at the fundus of IAC. These cases are summarized in Table 1.

The normal range of the IAC is 2–8 mm, with an average of 4 mm, and a narrow IAC is defined when a canal is smaller than 2 mm in vertical diameter as seen on HRCT (9). The HRCT scan is the diagnostic modality of choice as it provides excellent bony detail and high sensitivity and specificity in demonstrating the congenital inner ear and temporal bone abnormalities (1). However, a CT scan has a limited role in assessing the neural components of the IAC. Casselman et al. (6) described seven cases with congenital or unexplained sensorineural hearing loss and five of those cases with a normal IAC showed aplasia or hypoplasia of the vestibulocochlear nerve or of only the cochlear branch on MR imaging. In patients with a narrow IAC, assessment of the cochlear nerve is crucial for selecting patients for cochlear implantation (4). Patients with aplastic cochlear nerves may not respond to the electric stimulation of the cochlear implantation. These reports emphasized that an HRCT scan alone is not sufficient to rule out the risk of aplasia or hypoplasia of the vestibulocochlear nerve and that MR imaging should be performed to look for the defect of neural structures in the IAC of patients with sensorineural hearing loss.

MR imaging has become the modality of choice along with HRCT for the assessment of abnormalities in patients with sensorineural hearing loss. Especially, the use of high-resolution gradient-echo imaging provides detailed anatomical images of the vestibulocochlear and facial nerves of an IAC and is essential for cochlear implant candidates (6). There are several newly developed high-resolution gradient-echo MR imaging sequences which include 3D magnetization prepared rapid gradient echo (MP-RAGE), 3D balanced fast field echo (B-FFE), 3D Fourier transformation-constructive interference in the steady state (3D FT-CISS), and 3D DRIVE sequences (5, 7). These sequences are all three-dimensional for data acquisition and submillimetric in spatial resolution, which are optimal to evaluate the neural structures less than 1 mm in diameter, such as the auditory and facial nerves. In this case, we used a 3D DRIVE sequence with a high field 3-Tesla MRI scanner to obtain detailed images for the neural structures of the IAC. Cho et al. (5) reported a case of narrow IAC syndrome with aplasia of the vestibulocochlear nerve and intact facial nerve that were successfully demonstrated by using parasagittal reconstruction MR imaging. In this case, the IAC was narrow and was composed of two separate canaliculi as seen on HRCT. On 3-T MR images using 3D DRIVE and a T2-weighted FSE sequence with axial and parasagittal planes perpendicular to the course of the IAC, there were a relatively larger anterosuperior portion containing the intact facial nerve and a stenotic inferoposterior portion without any neural contents.

In conclusion, an isolated narrow IAC with duplication associated with congenital sensorineural hearing loss and normal facial nerve function is extremely rare, and to the best of our knowledge, this is the sixth case reported for this malformation. An examination of this anomaly should include a past medical history and physical examination, auditometry including auditory brainstem response, a high-resolution CT scan, and high-resolution MR imaging. It is of great importance to make a diagnosis of aplasia or hypoplasia of the vestibulocochlear nerve in the IAC for cochlear implant candidates. For this purpose, high-resolution submillimetric gradient-echo MR images, such as 3D-CISS and 3D DRIVE, or T2-weighted FSE sequences should be obtained in the parasagittal plane perpendicular to the course of the vestibulocochlear and facial nerves.

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