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Case Report

Bilateral duplication of the internal auditory canal: A case with successful cochlear implantation

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ABSTRACT

We report a case of bilateral duplication of the internal auditory canal (IAC). An 11-month-old girl with congenital profound hearing loss was referred to our hospital. Imaging evaluations revealed bilateral IAC duplication, which contained the facial and cochleovestibular nerves in one canal, but no nerves in the other. She underwent cochlear implantation. At 5 months after surgery her hearing thresholds with the cochlear implant are 40 and 45 dB at 2000 and 4000 Hz, respectively. Bilateral duplicated IAC is extremely rare, with seven cases reported in the literature. This case represents a previously unreported type of IAC duplication, and is the first case in which cochlear implantation was successfully performed.

1. Introduction

Inner ear malformations are detectable on computed tomography (CT) in about 20% of patients with congenital sensorineural hearing loss (SNHL) [1]. In contrast, malformations of the internal auditory canal (IAC) are rare and are seen in association with a cochlear malformation, such as Michel deformity, common cavity deformity, cochlear aplasia, cochleovestibular hypoplasia, and incomplete partition types I or II [2]. They may include absence of the IAC, a bony defect at the lateral end, entire stenosis (hypoplasia), stenosis of the bony canal for the cochlear nerve, dilatation, and a narrow, duplicated IAC [3]. Duplicated IAC is a rare malformation, with only 20 cases having been reported in the literature. Bilateral cases of duplicated IACs are even rarer, with only seven cases reported. We present a case of bilateral SNHL with duplicated IACs bilaterally. The current case represents a previously unreported type of IAC duplication, in which cochlear implantation was performed successfully.

2. Case presentation

An 11-month-old girl was referred to our hospital for cochlear implantation. Her hearing impairment was detected during a newborn hearing screen, and she started wearing hearing aids at 6 months of age. At the initial visit to our facility, her hearing threshold on conditioned orientation reflex audiometry (COR) was equal to or higher than 95 dB at 250, 500, 1000, 2000, and 4000 Hz. Her aided hearing levels ranged from 65 to 80 dB on COR. She does not have family history of hearing impairment. She underwent surgery for aproctia and hyperdactylia at 8 and 10 months of age, respectively. She had mild mental retardation and also had vesicoureteral regurgitation that was followed up without treatment.

Temporal bone computed tomography (CT) revealed that the external and middle ears appeared normal, but the bilateral inner ears were malformed. Although the basal turn of the cochlea appeared normal (Fig. 1a), the modiolus was deficient, and the middle and apical turns appeared bulbous due to the absence of the normal interscalar septa (Fig. 1b). The vestibule appeared normal bilaterally, but the semicircular canals were malformed (Fig. 1c). The posterior semicircular canals were enlarged. The lateral semicircular canals were present, but their posterior ends were blind-ended bilaterally (Fig. 1c). The left anterior semicircular canal appeared normal, but the right one was cystic (not shown in the figure). The internal auditory canal (IAC) was duplicated (Fig. 1c). Connections to the labyrinthine portion of the facial nerve and the superior vestibular nerve were identified at the lateral end of the anterior IAC. The posterior IAC exhibited wide communication with the vestibule. These findings were the same for both ears. The diameters of the right anterior and posterior IACs were 2.3 and

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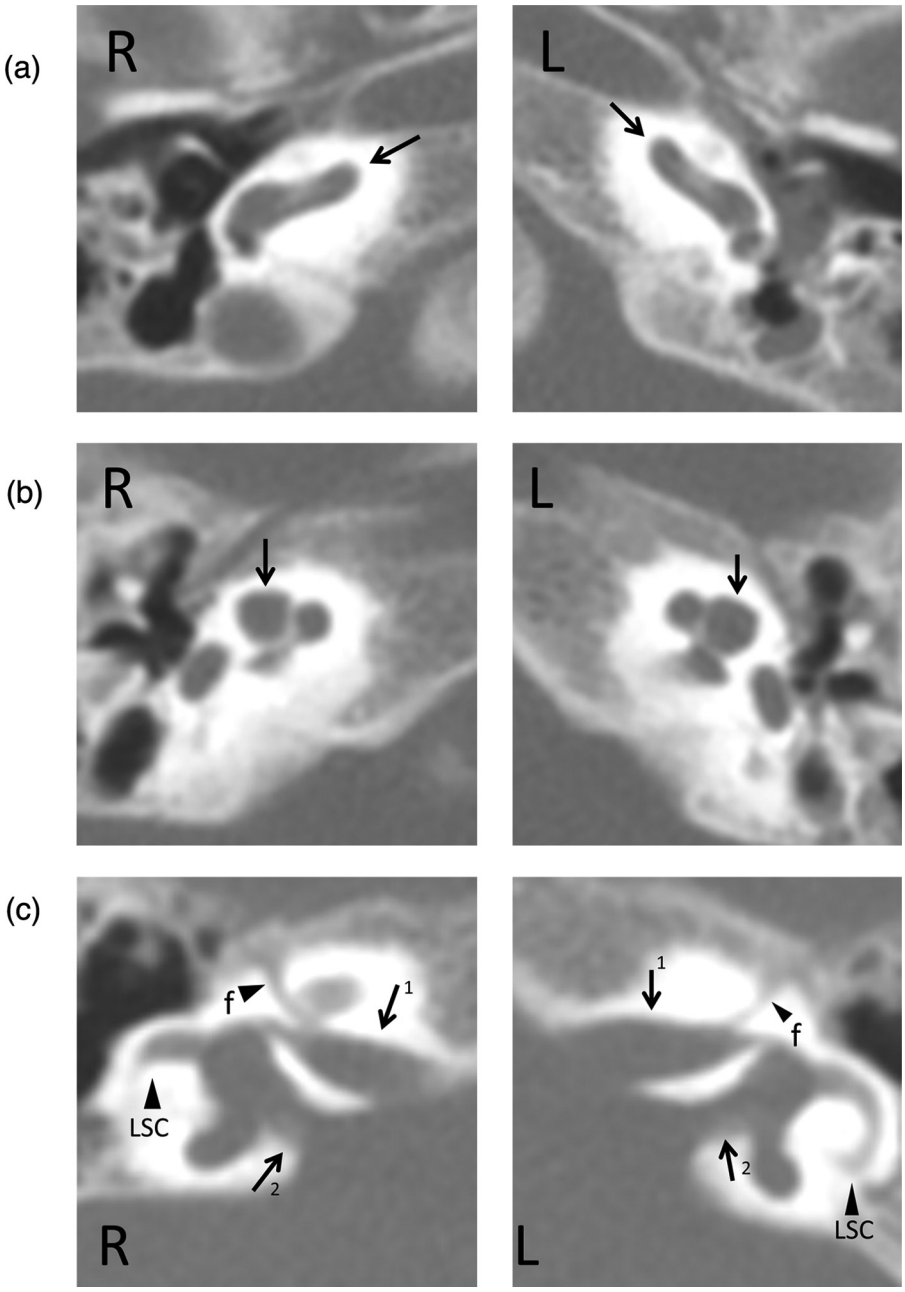

Fig. 1. Axial CT images of the inner ear. (a) The basal turn appeared normal bilaterally (arrows). R: right ear, L: left ear. (b) The modiolus was deficient, and the middle and apical turns appeared bulbous due to the absence of the normal interscalar septa (arrows). (c) The anterior internal auditory canal (IAC) (arrow 1) was connected to the labyrinthine portion of the facial nerve and the superior vestibular nerve. The posterior IAC (arrow 2) was connected to the vestibule. The posterior end of the lateral semicircular canal was blind-ended bilaterally. f: the labyrinthine portion of the facial nerve, LSC: lateral semicircular canal.

3.7 mm, respectively, and those of the left anterior and posterior IACs were 1.7 and 3.4 mm, respectively. T2-weighted magnetic resonance imaging (MRI) using the constructive interference in steady state (CISS) sequence clearly identified the cochlear nerve in the anterior IAC bilaterally (Fig. 2a). On parasagittal MRI, the cochlear nerve, vestibular nerve, and facial nerve were identified in the anterior IAC bilaterally (Fig. 2b). The posterior IACs appeared uniformly hyperintense on T2-weighted imaging, suggesting that cerebrospinal fluid (CSF) filled the space (Fig. 2a and c, both panels).

She was diagnosed as having bilateral profound hearing loss due to inner ear malformation. Cochlear implantation was performed at 1 year 7 months of age. The right ear was chosen for implantation because the history of otitis media was less frequent in the ear than in the left ear. The mastoid portion of the facial nerve was bifurcated, which was not clearly observable in preoperative CT images, with an additional branch identified anterior to the normally positioned facial nerve. However, posterior tympanotomy was possible without injuring the nerves, and cochleostomy was performed anteroinferior to the round window. Mild CSF gusher was encountered when cochleostomy was performed. An electrode array (Cochlear™ Nucleus® CI24 RE Contour Advance; Cochlear Ltd.) was inserted into the scala tympani of the cochlea. The results of intraoperative neural response telemetry and intraoperative electrically evoked auditory brain stem response were both normal. She is now in a rehabilitation program. At 5 months after the surgery, her hearing thresholds with the cochlear implant are 40 and 45 dB at 2000 and 4000 Hz, respectively.