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Clinical Report

A Challenge for Cochlear Implantation: Duplicated Internal Auditory Canal

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Duplication of the internal auditory canal is an uncommon, congenital malformation that can be associated with sensorineural hearing loss owing to aplasia/hypoplasia of the vestibulocochlear nerve. Only 14 such cases have been reported to date. We report the case of a 13-month-old girl with bilateral, congenital, sensorineural hearing loss caused by narrow, duplicated internal auditory canals and discuss the challenges encountered in the diagnosis and treatment of this condition.

KEYWORDS: Congenital hearing loss, sensorineural hearing loss, internal auditory canal duplication, temporal bone malformation

INTRODUCTION

Anomalies of the internal auditory canal (IAC) are infrequently observed in clinical practice [1]. These anomalies are usually unilateral and are associated with other defects of the inner, middle, or external ear [1, 2]. A narrow, duplicated IAC is a very rare defect characterized by the division of IAC into two by a complete or incomplete bony septum [1, 3]. To date, only 14 cases of a narrow, duplicated IAC have been reported, including a case of IAC triplication. A duplicated IAC is often associated with other systemic developmental anomalies such as malformations of the heart, kidneys, skeletal system, and intestinal tract [4]. IAC anomalies can also present as a component of disorders such as Michel anomaly, Mondini malformation, Bing–Siebenmann dysplasia, Scheibe dysplasia, Klippel–Feil syndrome, and CHARGE syndrome [4].

A narrow, duplicated IAC is associated with ipsilateral, congenital, sensorineural hearing loss (SNHL) caused by aplasia or hypoplasia of the vestibulocochlear nerve or its cochlear branch. Although IAC anomalies are relative contraindications to cochlear implantation, Casselman et al. [5] and Maxwell et al. [6] have shown moderate speech perception test results after cochlear implantation.

CASE PRESENTATION

An otherwise healthy 13-month-old girl was referred to our department for the investigation of hearing loss after failing to clear screening tests for hearing in both ears. She had a family history of SNHL (third-degree relatives). An examination of the head and neck in the patient was unremarkable, and the facial nerve function was normal and symmetrical. Her mother had had an uneventful gestation. The patient had no history of otalgia, otorrhea, or ototoxic medications and showed no evidence of perinatal sepsis, meningitis, or recurrent otitis media. Brainstem-evoked response audiometry testing revealed bilateral and profound SNHL. Computed tomography (CT) of the temporal bone demonstrated that both IACs were partially divided into two by a horizontal bony septum. The diameter of the right and left IACs were 2.3 and 1.8 mm, respectively. Both divided canals were narrow. The maximum calibers of the superior and inferior canals of the right IAC were 1.1 and 1.0 mm and of the left IAC were 0.8 and 0.9 mm, respectively. On both sides, the superior canal was continuous with the labyrinthine segment of the facial canal. The inferior canal of the right IAC was continuous with the cochlear aperture. The left IAC was not connected with the left cochlea (Figure 1). Magnetic resonance imaging (MRI) revealed that the right cochlear nerve was hypoplastic and that the left cochlear nerve was absent (Figure 2). Three-dimensional MRI confirmed that the right IAC was continuous with the cochlear aperture and a tiny canal and that the left IAC and left cochlea were not connected (Figure 3a, b). Promontory stimulatory testing detected V waves on the right but not on the left, indicating that the cochlear nerve was present only on the right. Considering the findings of the radiological and audiological tests, right cochlear implantation was performed, despite that fact that a narrow, duplicated IAC is a relative contraindication for cochlear implantation. The operation was performed after obtaining informed consent from the patient's parents. During the operation, no gusher was noted, and no other intraoperative and postoperative complications occurred. X-ray films in the Stenvers and transorbital views were used to confirm that the device had been accurately placed in the cochlea (Figure 4a, b). Two-year follow-up evaluations showed that the cochlear implantation improved speech perception in our patient. After her first year, she started speaking with one words such as "anne," "baba," and "dede." She started to identify easy commands and react to her name quietly, but she did not have any speech perception in noise. In her second year, her vocabulary expanded and she started using two-word phrases. She could repeat speech perception test words presented quietly and in several different signal-to-noise ratios.

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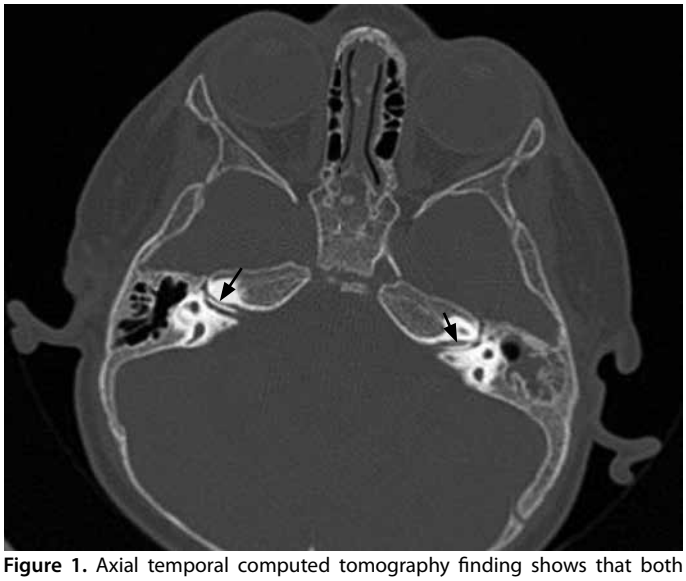

Figure 1. Axial temporal computed tomography finding shows that both internal auditory canals are partially divided into two by a horizontal bony septum (arrows).

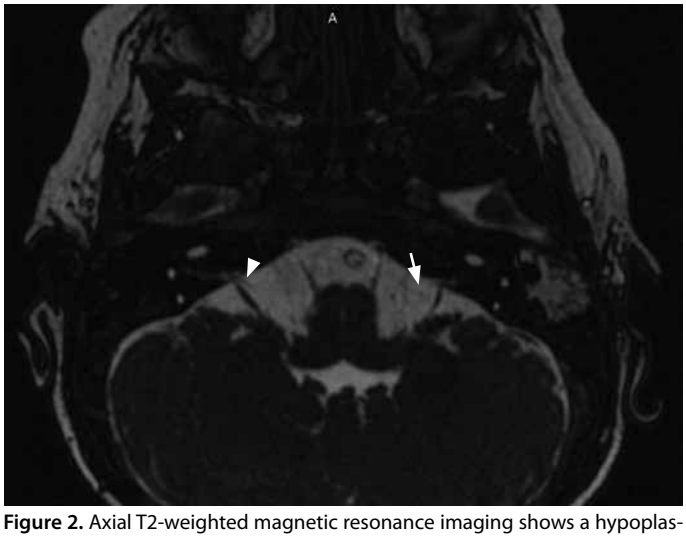

Figure 2. Axial T2-weighted magnetic resonance imaging shows a hypoplastic cochlear nerve on the right side (arrowhead) and no cochlear nerve on the left side (arrow).

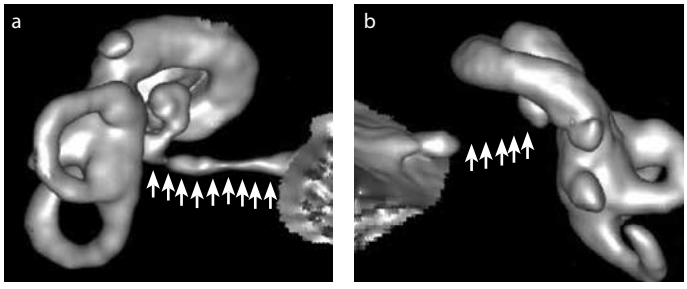

Figure 3. a, b. Three-dimensional magnetic resonance image shows that the internal auditory canal (IAC) is continuous with the cochlear aperture and a tiny canal on the right side (a, arrow) and that there is no connection between the left IAC and the left cochlea (b, arrow).

DISCUSSION

Only 20% of patients with congenital SNHL are found to have visible, bony abnormalities of the inner ear on CT [7]. Anomalies of IAC, such as atresia, stenosis, aplasia and hypoplasia, comprise 12% of congenital temporal bone anomalies [7, 8]. The diameter of a normal IAC is 2–8 mm (mean, 4 mm); IAC with a diameter of <2 mm is considered narrow or stenotic. IAC is almost perfectly symmetrical in healthy individuals, and the difference between the left and right IACs is <1 mm in 99% of individuals and 1–2 mm in 1% of patients [9].

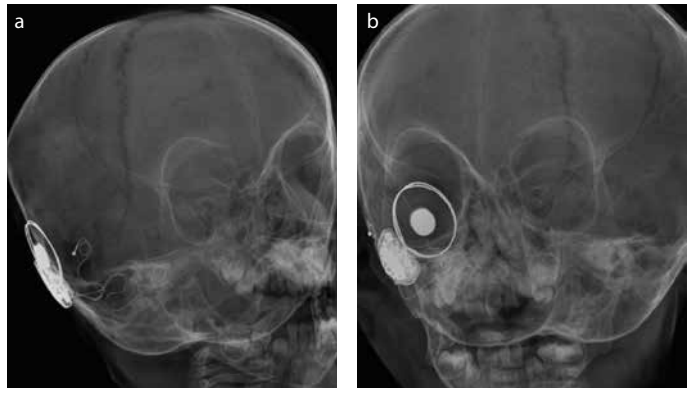

Figure 4. a, b. Stenvers X-ray (a) and transorbital X-ray (b) films show electrodes in place.

Two hypotheses have been proposed to explain the mechanism underlying IAC stenosis. The first and more widely accepted hypothesis states that the embryonic cochlea induces the growth of the vestibulocochlear nerve (eighth cranial nerve) and that the bony canal develops around the eighth as well as the seventh cranial nerves via mesoderm chondrification and ossification in the eighth gestational week. When the eighth cranial nerve is hypoplastic or aplastic, IAC does not properly develop. The other hypothesis states that the bony defect inhibits the growth of the eighth cranial nerve via mechanical narrowing. However, the latter hypothesis does not explain why facial nerve (seventh cranial nerve) function is preserved in most patients with IAC stenosis [4]. High-resolution CT shows bony structures in detail and is highly sensitive and specific for demonstrating congenital abnormalities of the inner ear and temporal bone. However, it has a limited role in IAC assessments because it cannot display neural structures within IAC in sufficient detail [4, 5]. In contrast, MRI is valuable for evaluating the neural components of IAC in patients who have SNHL. Casselman et al. [5] have described cases of congenital SNHL associated with a normal IAC on temporal CT and eighth nerve aplasia or hypoplasia on MRI. High-resolution gradient echo imaging can provide detailed images of the vestibulocochlear and facial nerves in IAC and is an essential investigation in the preoperative work-up of patients who are candidates for cochlear implantation [5, 7, 10, 11].

Internal auditory canal stenosis is a clinically relevant anomaly that affects some patients who would benefit from cochlear implantation. This anomaly is a relative contraindication to this procedure because a part of the auditory pathway is missing in patients with IAC anomalies [7]. Several cases of cochlear implant failures have been reported in patients with narrow IACs [4]. However, a few cases of improved hearing after cochlear implantation have been described in patients with hypoplasia of the cochlear branch, as in the case of our patient [5]. Promontory stimulatory test results are crucial to determine whether the auditory pathway is intact, which correlates with better speech perception results after cochlear implantation [12, 13]. Negative promontory stimulatory test results indicate that the auditory pathway is

not intact. However, the nerve can still be functional but not enough electrical activity to produce positive results [13]. Based on this, some patients with negative promontory stimulatory test results can benefit from cochlear implantation.

The work-up for IAC duplication/stenosis should include taking the medical history and performing physical examinations, audiometry (promontory stimulatory test), high-resolution CT, and high-resolution MRI. Candidates for cochlear implantation must undergo tests to detect aplasia or hypoplasia of the vestibulocochlear nerve within IAC so that an appropriate treatment can be planned.

Informed Consent: Written informed consent was obtained from patients' parents who participated in this study.

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