

An Unusual Case of Unilateral Cochlear Nerve Agenesis with Bilateral Lateral Semicircular Canals Aplasia

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Abstract

Imaging of the inner ear structures is becoming essential nowadays as the increasing number of cases of cochlear implants being performed globally. We report here a case of a 19-year-old boy who had congenital hearing loss and was being evaluated in our service for possible cochlear implantation. Audiometry showed profound mixed sensorineural hearing loss of the right side and mild to moderate conductive hearing loss in the left side. The magnetic resonance imaging revealed only two nerves in the narrowed right inner auditory canal: one in the anterior superior quadrant, identified as the facial nerve, and one on the posterior quadrants, representing the incomplete separation of the superior and inferior vestibular nerves which is an anatomical variant. Bilateral lateral semicircular canals were not visualized and there is dysplastic vestibules morphology. The diagnosis for the patient was right cochlear nerve agenesis with bilateral lateral semicircular canals aplasia.

Keywords: Cochlea Nerve Agenesis; Cochlea Implant; Lateral Semicircular Canal Aplasia; Congenital Sensorineural Hearing Loss

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Introduction

The incidence of congenital sensorineural hearing loss is approximately 1–3 cases per 1,000 live births [1]. Since its commencement almost half a century ago, cochlear implantation has grown to become an accepted and well-recognized treatment option [2]. For this, the presence of the spiral ganglion cells and

the cochlear nerve is essential for signal transmission from a cochlear implant to the brain. Thus, the preoperative analysis of cochlear anatomy is obligatory because cochlear implantation is contraindicated in patients with cochlear nerve agenesis and inner ear malformations [3].

The purpose of imaging of the cochlea and Internal Auditory Canal (IAC) is to establish cochlear duct permeability, the presence of cochlear nerve in the IAC and to choose implantation side for cochlear implant candidate [4]. Both High-Resolution Computerized Tomography (HRCT) and Magnetic Resonance Imaging (MRI) gives information on the IAC thickness, the presence of the cochlear, vestibular, and facial nerves, and inner ear malformations [4]. The purpose of this paper is to present an unusual case of a 19-year-old girl congenitally deaf who was found to have unilateral agenesis of the cochlear nerve in narrowed IAC and bilateral lateral semicircular canals aplasia during preoperative workup for cochlear implantation.

Case Report

A 19-year-old girl presented with progressive bilateral hearing loss since childhood, more prominent in the right ear compared to left. The patient was free of vertigo and tinnitus. No symptoms of ear fullness or discharge were reported.

At the physical examination, otoscopy findings were normal with intact tympanic membranes and normal external auditory canals. Nasal endoscopy showed clear bilateral fossa of Rosenmuller with hypertrophy of bilateral inferior turbinate. No pus or nasal polyps seen. The facial nerve function was unimpaired and symmetrical.

She was born per vaginal delivery at term with normal birth weight. Antenatal history revealed mother had gestational diabetes mellitus on diet control. There was no history of ear infection or trauma. There was no family history of hereditary sensorineural hearing loss.

Pure tone audiometry and speech testing revealed profound mixed sensorineural hearing loss of the right side and mild to moderate conductive hearing loss in the left side. Acoustic reflex was absent bilaterally. Tympanometry showed Type A tympanogram bilaterally indicates normal middle ear function.

Axial and coronal reformatted HRCT images of the temporal bones were acquired. For evaluation of the seventh and eighth cranial nerves, 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), T1-weighted, T2-weighted, FLAIR and T2-weighted Fast Spin-Echo (FSE) on axial and sagittal oblique planes.

In temporal bone HRCT, the right IAC was relatively narrowed compared to the left side, measures 0.7mm on the right side and 2.8mm on the left side (Figure 1). There was also aplasia of the bilateral lateral semicircular canal with dysplastic vestibules (Figure 2). Bilateral inferior and superior semicircular canals were normal. The facial nerve canal was intact along its course. There were no abnormalities in the cochlea, ossicles, and tympanic membranes.

Figure 1: Coronal HRCT temporal shows right IAC was relatively narrowed compared to the left side (arrow).

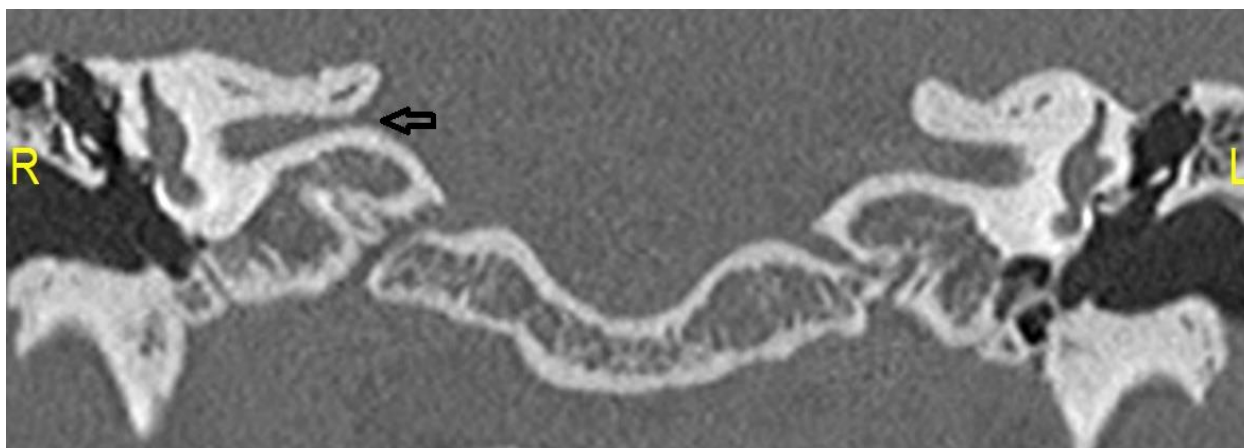
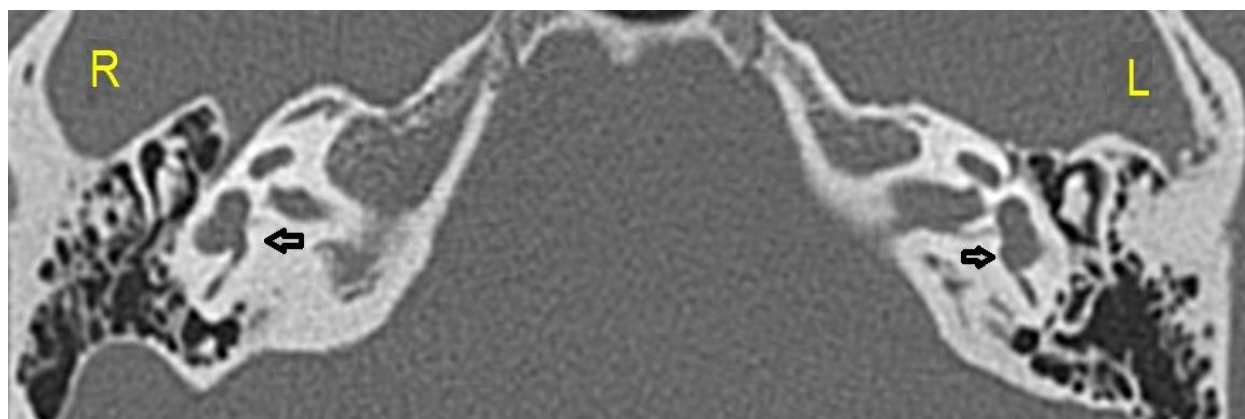
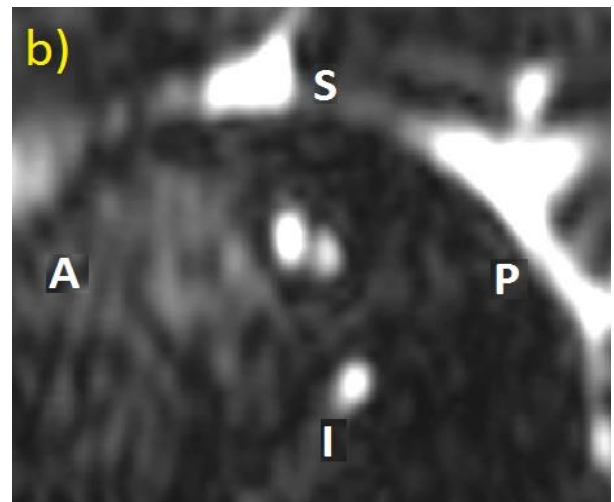
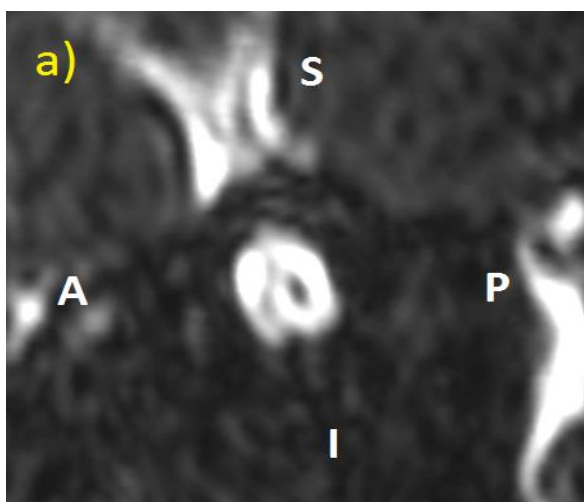


Figure 2: Axial HRCT temporal shows aplasia of the bilateral lateral semicircular canal with dysplastic vestibules (arrows).



Additional to CT findings, in MR imaging, the facial and cochlear nerves identified in the anterior part of IAC on the left side. In the posterior side of left IAC, only one nerve identified, likely represent the incomplete separation of the superior and inferior vestibular nerves, an anatomical variant. (Figure 3a). On the right side, only two nerves are identified on the left side, one in the anterior superior quadrant, also represent the incomplete separation of the superior and inferior vestibular nerves, an anatomical variant. There was no identifiable neural structure in the anterior inferior quadrant, in keeping with agenesis of cochlear nerve (Figure 3b).

Figure 3: Sagittal oblique 3D DRIVE MR imaging was obtained perpendicular to the course of the acoustic nerve in the IAC. The anterior (A), posterior (P), superior (S), and inferior (I) aspects of the canals are labeled for ease of orientation. a) The facial and cochlear were clearly identified on the left side. b) Agenesis of cochlear nerve in the right side. Facial nerve in the anterior superior quadrant. Only one vestibular nerve is seen in the posterior quadrant of bilateral sides, showing incomplete separation of the superior and inferior vestibular nerves (anatomical variant).



Discussion

Cochlear nerve anomalies are very rare in the populations. Mc Clay and et al., reported that cochleovestibular nerve anomalies were seen in 18% of patients with the sensorineural hearing loss [5]. Since the diagnosis is based solely on radiologic findings, neuroimaging of all patients should be analyzed carefully and all the audiological tests including otoacoustic emissions, brainstem response should be performed to the patients with sensorineural hearing loss [6].

On HRCT temporal, a bony cochlear nerve canal of <1.3mm or IAC of <3mm is suggestive of cochlear nerve anomaly. A closed bony cochlear nerve canal confirms the diagnosis. [2] However, in one study, 38% of cochlear nerve anomaly cases on MRI were actually found to have a normal sized IAC and bony cochlear nerve canal on HRCT. In order to avoid a missed diagnosis, MRI has been suggested as the first line imaging modality over HRCT [7].

MRI cross-sectional images done in the sagittal oblique plane are the best for the detection of cochlear nerve aplasia,

hypoplasia, or agenesis because they allow the complete visualization of the facial, cochlear, and vestibular nerves. Aplasia of the cochlear nerve is the absence of the nerve in the anterior inferior quadrant in the IAC. The vestibular nerves may be identified separately or as one only nerve. The latter case is considered an anatomic variant as seen in our case [8].

The drawback of MRI is in the case of a narrowed IAC where resolution may be insufficient for identifying separate nerves within the canal [7]. A very thin cochlear nerve also cannot be seen on MRI and it can intermingle with facial or vestibular nerve so may be reported as cochlear agenesis. A few cochlear nerve branches could still deliver some acoustic information to the auditory center [9].

Govaerts et al., have suggested a radiological classification of the hypoplasia and aplasia of the vestibulocochlear nerve in 2003 based on the affected branch and related labyrinth dysplasia. Type I: the vestibulocochlear nerve is affected, with or without dysplastic labyrinth and stenotic IAC. Type IIa: the cochlear branch is affected with labyrinth dysplasia.

Type IIb: the cochlear branch is affected with normal labyrinth. Type III: vestibular nerve is affected [10]. However, the isolated hypoplasia or aplasia of the vestibular branch without the presence of the branch to the cochlea has not been described up to now [11]. The present patient was a Type IIb with lateral semicircular canals aplasia and vestibular dysplasia.

The proposed pathogenesis of cochlear nerve anomaly includes failure in development of the nerve or as a result of degeneration. Although the underlying mechanisms remain unclear, postulated theories include vascular injury, apoptotic nerve remodeling, neurotrophic infections, metabolic and neurologic disorders [12].

In conclusion, congenital cochlear nerve agenesis is very uncommon. The imaging studies are the most important tools in the diagnosis of this entity. CT and MRI can evaluate possible abnormality of the inner ear including the labyrinth and vestibulocochlear nerve. Patients with cochlear nerve agenesis usually may not benefit from cochlear implantation. Brain stem implantation may be a better therapeutic option than cochlear implantation in these patients [13, 14].

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