Clinical Features for the Patients with Unilateral Cochlear Nerve Canal Stenosis

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Abstract

Objective: The clinical symptoms, audiological and imaging features of unilateral cochlear nerve canal stenosis were reported. Method: The clinical data of 12 patients with unilateral cochlear nerve canal stenosis diagnosed in Beijing Friendship Hospital from January 2018 to December 2019 were analyzed retrospectively, including age of onset, clinical symptoms, signs, audiology, HRCT and MRI of temporal bone and so on. Results: Twelve patients (6 males and 6 females) were identified with unilateral cochlear nerve canal stenosis. All patients presented with hearing loss and no other complaints. Among them, 2 patients were accompanied with accessory ears and the rest had no positive signs. 11 patients had severe sensorineural hearing loss, 1 patient had normal low-frequency hearing and high-frequency hearing loss. Imaging showed that 9 patients were suspected atresia of the cochlear nerve canal, 3 patients were narrow. Conclusion: For patients with unilateral stenosis of cochlear canal, early diagnosis and intervention should be carried out as early as possible, and the healthy ears should be protected.

1. Introduction

Congenital sensorineural hearing loss (SNHL) is a common congenital malformation in the world, with an estimated prevalence of 1-2 newborns per 1000, although estimates vary and may be higher(Morton and Nance, 2006). The osseous cochlear nerve canal is the space between the cochlear modiolus base and the internal auditory canal base which carry the cochlear nerve fibers. The definition of cochlear nerve canal stenosis (CNCS) is the hypoplasia or absence of the cochlear nerve, including the congenital cochlear nerve deficiency and the acquired cochlear nerve deficiency(Glastonbury et al., 2002). The prevalence of CNCS in children with unilateral sensorineural hearing loss (USNHL) has been estimated around 57%(Yi et al., 2013). As we all know, cochlear nerve atrophy sometimes leads to an unsatisfactory outcome after cochlear implantation(CI). Therefore, it is very important to judge whether there is CNCS before surgery. High-resolution computed tomography(HRCT) can accurately display the characteristics of various types of skeletal cochlear malformations, and is the preferred method for examination of congenital sensorineural hearing loss. CT findings of the internal auditory canal(IAC) stenosis suggest cochlear nerve aplasia(Jackler et al., 1987,Shelton et al., 1989). The diameter of BCNC less than 1.5mm was defined as hypoplasia by Liu(刘伟 et al., 2013), similar to the value established by Komatsubara(Komatsubara et al., 2007) and Miyasaka(Miyasaka et al., 2010). Herein we report 12 unilateral cochlear nerve canal stenosis cases.

Materials and methods

We retrospectively analyzed 12 unilateral CNCS patients (6 males and 6 females) aged 5-32 years (mean age 14.50±7.68 years) whose chief complaint was hearing loss at [removed for blind peer review] between January 2018 to December 2019. Exclusion conditions were inadequate CT or MRI examination and a history of potential diseases (preterm birth, hypoxia, and hyperbilirubinemia,trauma, meningitis). All patients underwent pure tone audiometry(PTA), acoustic immittance, HRCT and Magnetic resonance imaging (MRI) of temporal bone.

3. Results

3.1 Clinical manifestations and signs

The clinical manifestations of the 12 patients were hearing loss on the affected ear and normal hearing on the healthy ear. No patients experienced tinnitus or vertigo. Among the 12 CNCS patients, 2 cases had accessory auricle (Figures 1). The average time from the early presentation of symptoms to diagnosis of unilateral CNCS was 6.68±5.07 years. Nine of the 12 unilateral CNCS were found on the left ear, and 3 were found on the right ear. Otoscope examination revealed an intact tympanic membrane in all patients.

3.2 Audiologic evaluations

Patients with USNHL confirmed by a pure tone audiogram were included in this study. Cases 1 to 11 had almost no residual hearing in the affected ears, and case 12 had normal hearing at low frequencies. Audiogram shapes of case 12 were high frequency sloping. In addition to the PTA, acoustic immittance and distortion product otoacoustic emission (DPOAE) were performed. Acoustic immittance test showed that all of the cases were type A. In 12 patients, the amplitude of DPOAE decreased in 6 patients, and the other 6 patients did not lead out (Figures 2).

3.3 Imaging Study

3.3.1 HRCT

The diameter of cochlear nerve canal was measured by HRCT. Each patient underwent a multidetector-row CT scan of the temporal bone HRCT. CT images were obtained in the direct axial plane using a slice thickness of 1mm. The diameter of cochlear nerve canal was evaluated by measuring the line of the inner edge of the cochlear nerve canal at the internal auditory canal fundus perpendicular to the modiolus. The criterion for CNCS is that the axial HRCT shows a cochlear nerve canal diameter of less than 1.5mm.

In addition to CNCS, we also analyzed abnormalities in other sites, such as cochlea, IAC, vestibule, semicircular canal and vestibular aqueduct. All patients had unilateral CNCS. Patients with bilateral dysplasia were excluded, as were cases with other malformations suggestive of more complex syndromes. CT showed 9 cases with suspected atresia of the cochlear canal, and the average width of the other 3 cases was 1.17±0.09mm(Figure 3). Two of these patients had stenotic internal auditory canal(Figure 4).There were no abnormalities found in the external auditory meatus or the middle ear on either side.

3.3.2 MRI

MRI was obtained on a 3.0 Tesla system using a eight-channel head and neck unite coil. The MRI scan included 3-D T2 space dark fluid sequence in the IAC axial and oblique sagittal images with a slice thickness of 0.5mm.The imaging parameters for the three-dimensional driven equilibrium (3D-DRIVE) sequences were as follows: echo train length=70; matrix=384×324; repetition time/echo time=1400/162ms; field of view=15cm; section thickness=0.5mm; overlap=0.75mm; number of acquisitions =2; acquisition time=3.5min. Axial MRI can show the facial nerve and the cochlear nerve(Figure 5). Then, images visualizing four nerves (the facial nerve, the superior vestibular nerve, the inferior vestibular nerve, and the cochlear nerve) were generated by reconstructing three-dimensional constructive interference in the steady state (CISS) image through IAC in a vertical direction. The MRI showed that the cochlear nerve was not detected in 9 cases and thin in 3 cases (Figure 6).

4.Discussion

Congenital sensorineural deafness affects 11% of children and is one of the most common health problems(Erenberg et al., 1999). USNHL is usually found in primary schools, with about 0.1-3 percent of children affected(Bamiou et al., 1999; Bess et al., 1998). In this study, the average age of diagnosis was 14.50±7.68 years, and the average time from symptoms to diagnosis was 6.68±5.07 years. The full-scale and performance intelligence quotient(IQ) score of children with unilateral hearing loss were lower than those of normal hearing children in a meta-analysis of children with unilateral deafness(Purcell et al., 2016).

Congenital unilateral deafness is caused by genetic factors, inner ear deformity, perinatal infection and other factors(Fatterpekar et al., 2000). According to imaging examination, the most common cause of USNHL in children is cochlear nerve abnormality(Lee et al., 2018). It is reported that the cochlear nerve deficiency occurs in up to 18% of congenital sensorineural deafness(McClay et al., 2008). The rate of CNCS in children with unidentified unilateral SNHL was 52.4%(Lim et al., 2018), and it is higher than that in children with bilateral deafness(Nakano et al., 2013). The mechanism of CNCS remains speculative. CNCS may be caused by partial or complete hypoplasia of nerve, or as a result of post-develop-mental degeneration. In SNHL patients, both congenital cochlear nerve defect and acquired cochlear nerve degeneration can be seen. In this study, all patients had no identifiable etiology and no clear family history for their unilateral CNCS.

Previously, it was believed that CNCS was associated with severe sensorineural deafness(Buchman et al., 2006a). However, in this report, there was no profound hearing loss in case 12. When this happens, it is thought that the affected ear has a very tiny cochlear nerve that conserves part of its function(Miyanohara et al., 2011). A small amount of residual cochlear nerve fibers transmitting voice information may be sufficient without raising the threshold(Valero et al., 2012).

CNCS can be manifested as auditory neuropathy(AN)(O'Leary and Gibson, 1999). In some children, auditory neuropathy may be caused by or related to CNCS(Buchman et al., 2006b). In this study, DPOAE was detected in all 12 patients; none of them had normal DPOAE amplitude, indicating normal function of outer hair cells. In this study, DPOAE was detected in 6 of 12 cases of unilateral CNCS(50%). The inducing rate of DPOAE has nothing to do with hearing level. The cause of DPOAE not elicited is considered to be congenital CNCS related inner ear microstructural malformation(Taiji et al., 2012).

CT is a very effective method for USNHL children. HRCT is commonly used to study the cause of hearing loss because of its wonderful resolution of temporal bone structure and low cost(Pagarkar et al., 2011). HRCT can evaluate the inner ear structure in detail, such as large vestibular aqueduct syndrome(LVAS),CNCS and stenosis of internal auditory canal. The most common abnormality in USNHL was CNCS, accounting for 69.7%(Masuda and Usui, 2019). CNCS is always related to cochlear abnormality(Miyasaka et al., 2010) or various coexisting syndromes(Buchman et al., 2006b). None of the patients studied in this report had known syndromes. But in this study, 2 of 12 ears of CNCS were accompanied by internal auditory canal stenosis. In USNHL, MRI is the gold standard imaging because cochlear nerve deficiency is frequently observed(Paul et al., 2017). Miyasaka(Miyasaka et al., 2010) reported that 8 of 42 ears (19%) had CNCS on MRI, among which 4 had inner ear deformity. In this study, MRI imaging showed that the cochlear nerve was not detected in nine ears, and the cochlear nerve was very thin in three ears.

In addition, the risk of progression of ipsilateral ear disease may increase in children with CNCS(Purcell et al., 2017). The contraindications of CNCS for CI in bilateral deafness patients are controversial. It is reported that the presence of absent cochlear nerve seemed to be associated with poorer outcomes(Tay et al., 2019). However, it has also been reported that cochlear implantation has some benefits in children with cochlear nerve defects(Birman et al., 2016). Although unilateral hearing loss is usually not treated by CI, future research can explore the relationship between CNCS and auditory rehabilitation.

Conflict of Interest Statement

None of the authors have potential conflicts of interest to be disclose.

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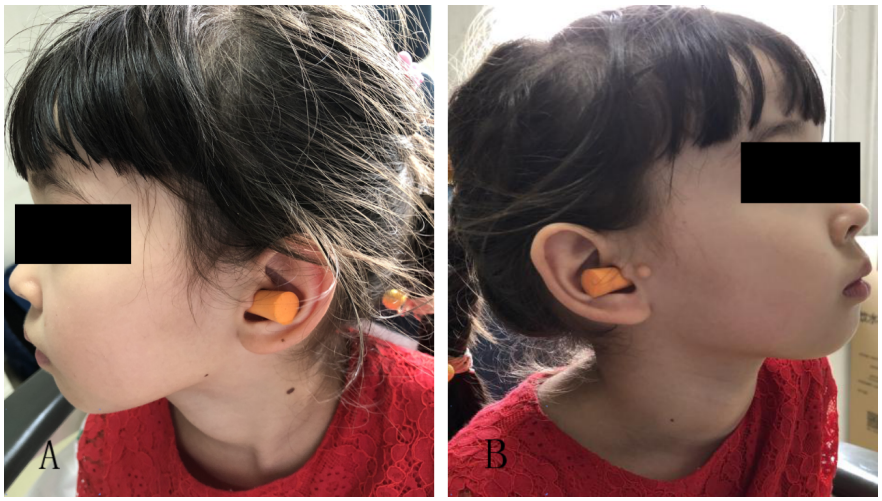

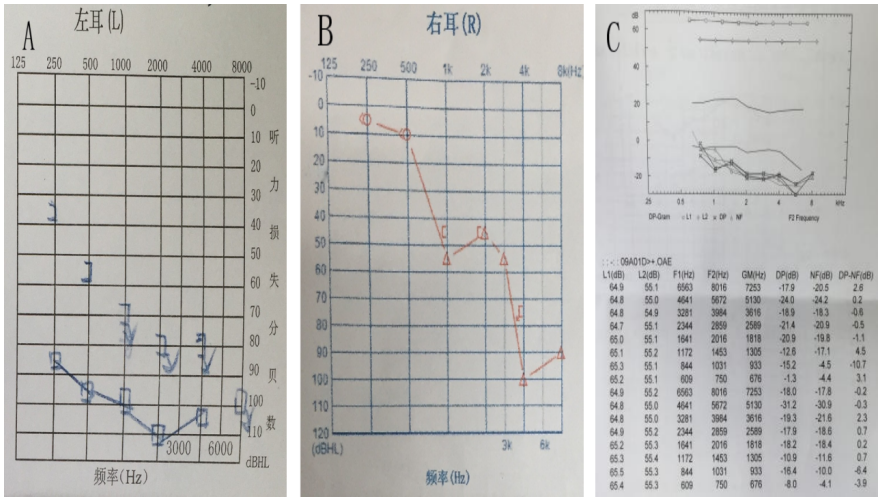

L1(dB)	L2(dB)	F1(Hz)	F2(Hz)	GM(Hz)	DP(dB)	NF(dB)	DP-NF(dB)
64.9	55.1	6563	8016	7253	-17.9	-20.5	2.6
64.8	55.0	4641	5672	5130	-24.0	-24.2	0.2
64.8	54.9	3281	3984	3616	-18.9	-18.3	-0.6
64.7	55.1	2344	2859	2589	-21.4	-20.9	-0.5
65.0	55.1	1641	2016	1818	-20.9	-19.8	-1.1
65.1	55.2	1172	1453	1305	-12.6	-17.1	4.5
65.3	55.1	844	1031	933	-15.2	-4.5	-10.7
65.2	55.1	609	750	676	-1.3	-4.4	3.1
64.9	55.2	6563	8016	7253	-18.0	-17.8	-0.2
64.8	55.0	4641	5672	5130	-31.2	-30.9	-0.3
64.8	55.0	3281	3984	3616	-19.3	-21.6	2.3
64.9	55.2	2344	2859	2589	-17.9	-18.6	0.7
65.2	55.3	1641	2016	1818	-18.2	-18.4	0.2
65.3	55.4	1172	1453	1305	-10.9	-11.6	0.7
65.5	55.3	844	1031	933	-18.4	-10.0	-8.4
65.4	55.3	609	750	676	-8.0	-4.1	-3.9