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Missing perilymph but leaking blood-endolymph and vestibulocochlear nerve barriers in idiopathic sudden sensorineural hearing loss: A case study

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ABSTRACT

Objectives: To evaluate the pathological changes in the blood-perilymph, blood-endolymph, and blood-nerve barriers of a patient with idiopathic sudden sensorineural hearing loss (SSNHL).
Methods: Potential ossification or fibrosis in the inner ear was evaluated using temporal bone CT and MRI acquired using the 3-dimensional T2-weighted sampling perfection with application-optimized contrasts using a flip angle evolution sequence. Pathological changes in the barriers were analyzed by MRI obtained 4 h after a single-dose intravenous injection of gadolinium chelate using a medium inversion-time inversion recovery imaging with magnitude reconstruction sequence.
Results: The perilymph was absent, while significant enhancements of the vestibulocochlear nerve and the endolymphatic compartments were detected.
Conclusion: Significant injuries in the blood-endolymph and blood-vestibulocochlear nerve barriers and disabled perilymph production may contribute to the development of SSNHL with poor response to treatments.

1. Introduction

The etiology and mechanism of sudden sensorineural hearing loss (SSNHL) are uncertain, and a disrupted blood-labyrinth barrier has been reported in clinical studies (Berrettini et al., 2013; Cadoni et al., 2006; Seltzer et al., 1991; Sugiura et al., 2006; Wu et al., 2014; Yoshida et al., 2008). However, previous reports did not distinguish the pathological changes between the blood-endolymph and blood-perilymph barriers. Here, I report a patient with idiopathic SSNHL displaying missing perilymph but leaking blood-endolymph and vestibulocochlear nerve barriers. To the best of our knowledge, this is the first report of the unique MRI manifestations of missing perilymph in humans. All the protocols followed the Declaration of Helsinki (1964) developed by the World Medical Association and its later amendments.

2. Case presentation

A 52-year-old man suffered from sudden hearing loss, tinnitus and aural fullness in the left ear on 30-8-2019 that were accompanied by transient vertigo, nausea and vomiting for seconds at a time. Audiograms showed an average hearing level of 95 dB nHL at frequencies over 0.5–4 kHz on the left side (Fig. 1). The hearing loss did not recover after systemic administration of methylprednisolone, ginkgo leaf extract and dipyridamole 15 h after onset and delayed infusion of batroxobin 3 d later, in combination with hyperbaric oxygen. After 11 d, the patient visited me seeking further treatment when the hearing became worse (Fig. 1). However, his hearing did not respond to salvage intratympanic dexamethasone therapy. Potential ossification in the inner ear was excluded by temporal bone CT. Uniformly missing signals of both the cochlear and vestibular perilymph on MRI acquired using a 3-dimensional T2-weighted sampling perfection with application-optimized contrasts using a flip angle evolution (SPACE) sequence did not support the existence of either fibrosis or tumor in the inner ear. Significant enhancements of the vestibulocochlear nerve and the endolymphatic compartments (scala media, saccule, utricle, and ampullae of the semicircular canals) but not the perilymphatic

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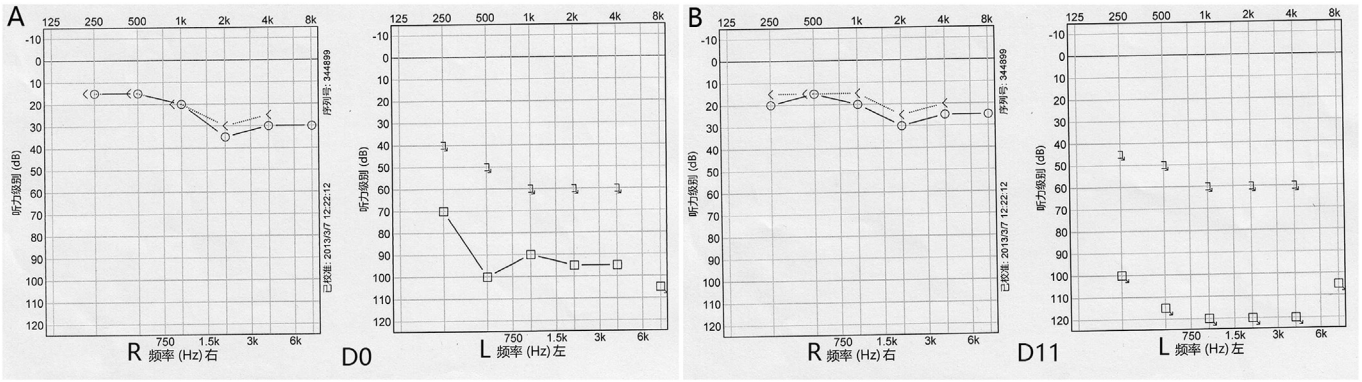

Fig. 1. Pure tone audiograms of a patient with idiopathic sudden sensorineural hearing loss in the left ear. A profound hearing loss on the day of onset (A) progressed to total deafness on the day of MRI measurement (B). D0: the onset day; D11: the 11th day after onset; L; left ear: R: right ear.

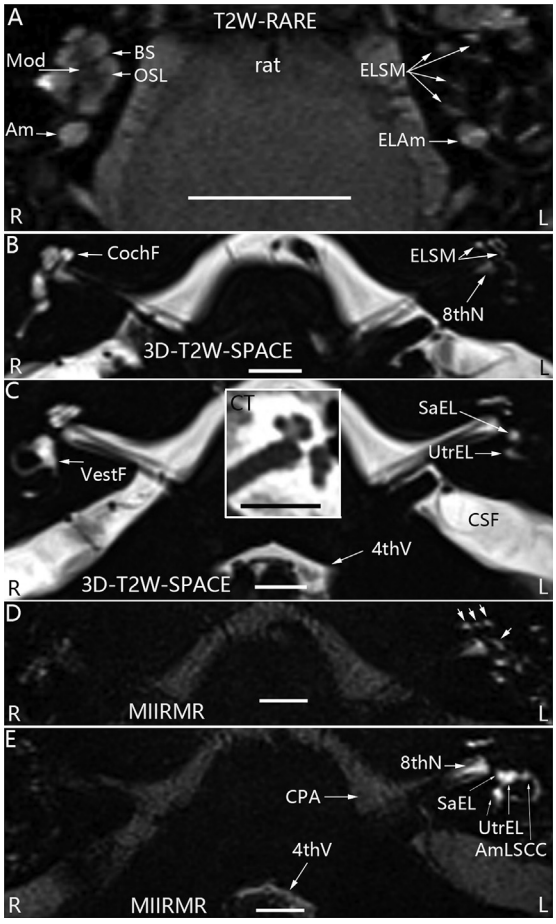

Fig. 2. MRI of a patient with idiopathic sudden sensorineural hearing loss in the left ear 4 h after a single-dose intravenous injection of gadolinium chelate. Positive control (A): A 4.7 T animal MR and bird cage coil were used to image the inner ear of rats that received an injection of superparamagnetic ironic nanoparticles coated with Pluronic® F127 copolymer and oleic acid (Zou et al., 2010). The human inner ear was imaged using a 3 T MR machine and a 20-channel Tim 4G head/neck coil. The inner ear fluids were displayed using the SPACE sequence, which only demonstrated endolymph in the left ear (L) (B, C), and potential ossification in the inner ear was excluded by CT (insert in C). Inner ear enhancement on the left side (L) displaying strong signals in the endolymph of the scala media (arrows in D) and vestibule (E) was detected using medium inversion-time inversion recovery imaging with magnitude reconstruction (MIIRMR); this enhancement was almost negative on the contralateral right side (R). 4thV: cerebrospinal fluid (CSF) in the 4th ventricle; 8thN: vestibulocochlear nerve; Am: ampulla; AmLSCC: Ampulla of the lateral semicircular canal; BS: bony separate between cochlear turns; CochF: cochlear fluids; CPA: CSF in the cerebellopontine angle; ELAm: endolymph in the ampulla; ELSM: endolymph in the scala media; Mod: modiolus; OSL: osseous spiral lamina; SaEL: saccular endolymph; UtrEL: utricular endolymph; VestF: vestibular fluids. Scale bars = 5 mm (A, D, E), 6 mm (B, C), and 8 mm (inserted window in C).

spaces were demonstrated on MRI obtained 4 h after a single-dose intravenous injection of gadolinium chelate (GdC) using a sequence of medium inversion-time inversion recovery imaging with magnitude reconstruction (Zou et al., 2019). The inner ear morphology was similar to that seen on MRI of the rat inner ear, in which the perilymphatic signals were deleted by delivering superparamagnetic iron oxide nanoparticles into the scala tympani (Zou et al., 2010). The images acquired using the SPACE sequence confirmed that the perilymph was missing and that there was no vascular loop in the internal auditory canal (Fig. 2).

3. Discussion

A potential intracochlear schwannoma can be ruled according to its specific segmental enhancement on postcontrast MRI and missing signals of both the endolymph and perilymph in a segmental pattern on T2-weighted MRI (Bittencourt et al., 2014). Again, a potential contribution of endolymphatic hydrops in the left inner ear to the unique MRI manifestation can be definitely excluded because there was no perilymph signal in the left inner ear on MRI acquired using a SPACE sequence. I interpret the present unique MRI manifestation as a result of missing perilymph but leaking blood-endolymph and vestibulocochlear nerve barriers. The strong evidence of missing perilymph on MRI obtained using the SPACE sequence is that both the perilymph and endolymph should display bright signals regardless of whether they are enhanced.

In the normal inner ear, GdC only enters the perilymph but not the endolymph after intravenous injection, even at a high dose (Zou et al., 2003). This is due to the critical role of the blood-endolymph barrier (BEB) in maintaining endolymphatic ionic homeostasis by preventing the entry of deleterious substances from the capillaries of the stria vascularis, which is essential for hair cells to transform vibration into electric activity (Nin et al., 2008). A breakdown in the BEB may cause a failure of hair cell functioning. The blood-perilymph barrier (BPB) is relatively porous, and the perilymph contains proteins five times more concentrated than in the cerebrospinal fluids, including, among others, apolipoproteins D and J (Thalmann et al., 1994). Both electrolytic homeostasis, which is similar to that in the extracellular matrix, and the apolipoproteins are essential for cellular viability and activity in the inner ear. For example, apolipoprotein D performs an anti-neurodegenerative role by preventing oxidative stress and ensuing lipid peroxidation, while apolipoprotein J protects cells to combat cytotoxicity and senescence (Dumont et al., 2002; Navarro et al., 2013). The perilymph is mainly produced in the cochlear glomeruli of Schwalbe in the modiolus, which is a downstream branch of the

labyrinth artery (Zou et al., 2003). That the previous study did not identify occlusion of the labyrinth artery using 7 T MR does not exclude the existence of thrombosis in the downstream vasculatures such as the cochlear glomeruli of Schwalbe in SSNHL (Sato et al., 2014). In the present case, such thrombosis occurred, disabling perilymph production. The delayed infusion of batroxobin failed to resolve the thrombus. The blood-nerve barrier (BNB) is highly restrictive, limiting the entry of substances into the blood and maintaining the homeostasis of nervous tissue. The highly intense signals in the 8th nerve region suggested a breakdown in the blood-nerve barrier that mimics the MRI manifestation of neurosyphilis (Ribeiro et al., 2019). A failure in BNB function may further disturb inner ear activity. Similar to that in other peripheral nerves, endothelial dysfunction in the interface between the blood and the vestibulocochlear nerve may occur through several mechanisms, such as a reduction in nitric oxide and upregulation of pro-inflammatory cytokines (Maiuolo et al., 2019).

This is the only patient among the more than 60 patients with SSNHL imaged using the same protocol (the data will be reported separately).

4. Conclusion

To conclude, the patient in the present case report displayed significant injury in the blood-endolymph and blood-vestibulocochlear nerve barriers as well as disabled perilymph production, contributing to the development of SSNHL with poor response to treatments, and early thrombolytic therapy might be recommended.

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Declaration of competing interest

The author declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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