CASE REPORT

Endolymphatic Hydrops of the Labyrinth Visualized on Noncontrast MR Imaging: A Case Report

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We admitted an 11-year-old girl with enlarged endolymphatic duct and sac syndrome to our hospital with severe nausea and vertigo. Three-dimensional constructive interference in steady state (3D-CISS) and fluid-attenuated inversion recovery (3D-FLAIR) images revealed a reflux of proteinous or hemorrhagic fluid into enlarged endolymphatic space in the labyrinth. This is the first imaging report to show endolymphatic hydrops visualized by noncontrast-enhanced MR imaging in a living human patient.

Keywords: *endolymphatic hydrops, magnetic resonance imaging, temporal bone disease, 3D imaging*

Introduction

Recent studies have reported the visualization of endolymphatic hydrops on magnetic resonance (MR) imaging after intratympanic administration of Gd-DTPA¹⁻³ or possibly after intravenous administration of high-dose Gd-DTPA⁴ in patients with Ménière's disease. However, such visualization by nonenhanced MR imaging has not been reported in living humans.

Case Report

We admitted an 11-year-old girl with hearing deterioration in her right ear and attacks of vertigo to our hospital for severe nausea and vomiting. Since age 2, she had been followed for enlarged endolymphatic duct and sac syndrome (large vestibular aqueduct syndrome). From time to time, she had hearing loss and vertigo attacks and received steroid drip infusion and hyperbaric oxygen therapy. She also had a goiter and was diagnosed with Pendred syndrome based on genetic analysis. On this admission, hearing in her right ear was unmeasurable on the scale and in the left ear was 86.3 dB (average of 500, 1000, and 2000 Hz).

MR imaging obtained one day after admission (Fig. 1A-D) revealed an enlarged endolymphatic duct and sac that showed high signal on 3-dimen-

sional fluid attenuated inversion recovery (3D-FLAIR) and mixed signal on 3D constructive interference in the steady state (3D-CISS) images. On 3D-FLAIR images, the endolymphatic space in the cochlea and vestibule also showed high signal. Conversely, on 3D-CISS images, the signal in the labyrinth was low. These areas were thought to correspond to the reflux of high proteinous or hemorrhagic fluid from the endolymphatic duct and sac. Reference to images from other patients with Ménière's disease obtained after intratympanic injection of Gd-DTPA suggested that these areas were enlarged endolymphatic space (Fig. 1E).

Her nausea and vomiting improved during her 7-day stay in the hospital. On the day she was released, she fell and hit her head on the floor. She then complained of worsening hearing loss in her left ear and worsened nausea and vomiting and returned to the hospital. Six days after this head trauma, she underwent a second MR examination, which revealed increased signal in the basal turn of the left cochlea on 3D-FLAIR that seemed to extend into the perilymphatic space (Fig. 2).

All MR images were obtained on a 3-tesla scanner (Magnetom TRIO, Siemens Medical Solutions, Erlangen, Germany). We performed 3D-CISS imaging (voxel size $0.5 \times 0.5 \times 0.4$ mm) to obtain reference images of the anatomy of the labyrinthine fluid-space. 3D-FLAIR imaging (voxel size $0.7 \times 0.7 \times 0.8$ mm) was performed using a variable flip-angle echo train with an average flip angle of 120 degrees and an echo-train length of 119. The fea-

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Fig. 1. An 11-year-old girl with bilateral enlarged endolymphatic duct and sac syndrome. First magnetic resonance (MR) imaging series obtained 1 day after admission. **(A)** Posterior part of the enlarged endolymphatic duct and sac of the right ear shows lower signal on the 3-dimensional constructive interference in steady state (3D-CISS) image (arrow; EES). Low signal areas corresponding to endolymphatic space are seen in the cochlea (scala media, SM), vestibule (V), and posterior ampulla (A). Conversely, these same areas show high signal on 3D fluid-attenuated inversion recovery (3D-FLAIR) **(B)**. Note that no area corresponding to endolymphatic space can be recognized in the left labyrinth on 3D-CISS **(C)** and 3D-FLAIR **(D)**. As an anatomical reference for enlarged endolymphatic space, an image from a different patient who had Ménière's disease (a 60-year-old woman) is shown **(E)**. This is a 3D real inversion recovery (IR) image acquired after intratympanic injection of Gd-DTPA. This patient does not have an enlarged endolymphatic duct and sac. On 3D real-IR after intratympanic injection of Gd-DTPA, endolymphatic space shows negative signal (black), perilymphatic space shows positive signal (white), and surrounding bone shows zero signal (gray).²

Fig. 2. Images of the left ear. **(A)** A 3-dimensional fluid-attenuated inversion recovery (3D-FLAIR) image from the first magnetic resonance (MR) examination. The enlarged endolymphatic duct and sac show high signal (EES); however, no signal can be detected in the cochlea and vestibule. **(B)** A 3D-FLAIR image from the second MR examination performed 11 days after the first examination and 6 days after minor head trauma after which she experienced worsened left-side hearing loss. The enlarged endolymphatic duct and sac (EES) can be seen as in the previous study, and a slight signal increase can be seen in the cochlea (short arrows). High signal also extended into perilymphatic space. This finding is consistent with the reflux of proteinous or hemorrhagic fluid from the endolymphatic sac into the cochlea and with the contamination of the endo- and perilymph. **(C)** A 3-dimensional constructive interference in steady state (3D-CISS) image from the second MR examination is shown for anatomical reference. On 3D-CISS, the signal of the enlarged endolymphatic sac (EES) is low. That of cochlear fluid (short arrows) is high, suggesting only a small amount of reflux. Comparing **(B)** and **(C)**, the high signal in the cochlea in **(B)** can also be seen to extend into the scala tympani (perilymphatic space, ST, C).

tures of this variable flip-angle sequence have been reported elsewhere.⁵ This sequence allows the use of very long echo-train lengths without severe blurring while maintaining contrast similar to that of 3D-FLAIR obtained with a conventional turbo spin echo sequence, even with a long effective echo time.

Discussion

Enlarged endolymphatic duct and sac syndrome is the leading cause of progressive sensorineural

hearing loss in children and young adults. Though the syndrome is congenital, in most cases, the hearing loss is acquired.^{6,7} Because hearing loss increases after minor head trauma, as in our case, patients are usually advised to avoid contact sports.

The cause of hearing loss in this syndrome remains unknown,⁸ but 2 major theories exist.⁹⁻¹² The first is that cerebrospinal fluid (CSF) pressure in the internal auditory canal is transmitted through an insufficiently developed cochlear modiolus to inner-ear hair cells, but reports of normal-

sized cochlear modioli in some cases of this syndrome⁹ seem to refute this theory. The other theory is that the hyperosmolar fluid in the endolymphatic duct and sac refluxes into the labyrinth and damages hair cells.⁸ Proteinous or hemorrhagic fluid have been reported in the enlarged endolymphatic duct and sac on MR images.¹⁰ Recent literature supports the latter theory,^{9–11} especially a case report of a reflux of proteinous or hemorrhagic fluid from the duct and sac into the labyrinth visualized in 3D-FLAIR images.¹¹ However, no separate visualization between endo- and perilymph fluid was possible, probably as a result of contamination of the fluid by rupture of the Reissner's membrane.

In the present case, separation between endo- and perilymph was apparent in the right ear, and the endolymphatic space seemed to have enlarged according to the image criteria reported previously.¹³ In the criteria, the size of the endolymphatic space in the cochlear basal turn was scored subjectively from 0 to 3: 3, the cochlear duct is larger than the perilymphatic space of the scala vestibule; 2, the Reissner's membrane is bulging toward the scala vestibule but smaller than the perilymphatic space of the scala vestibule; 1, the Reissner's membrane is not bulging; and 0, the Reissner's membrane is bulging toward the scala media or there is no visualization of endolymphatic space. In our patient, the score of the right cochlea was 3. In the left ear, in contrast, contamination of the endo- and perilymph seemed likely at the time of the second MR examination.

This case report provides the first images that show endolymphatic hydrops visualized by non-contrast-enhanced MR imaging in a human patient. It is also the first imaging report to suggest fluid reflux into the cochlea just after head trauma in this syndrome. The knowledge obtained from the present case may improve understanding of the pathophysiology of enlarged endolymphatic duct and sac syndrome.

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