



The missing perilymph sign on MRI indicates a perilymphatic fistula: A case report

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ABSTRACT

A perilymphatic fistula (PLF) is a rare condition in which perilymph leaks from the cochlea or vestibule into the surrounding cavities, most commonly through round and oval windows, and causes cochlear and vestibular symptoms. However, vague symptoms and the lack of a clear diagnostic test have made the existence of PLF a controversial subject for decades. Here, we report a case of definite PLF confirmed by surgery in a patient who underwent mastoidectomy 20 years prior, revealing a specific sign of missing perilymph on MRI. T2-weighted MRI revealed a nodular bright signal in the cochlea and vestibule and a large area of bright signal in the middle ear cavity with a long tail running toward the vestibule in the left ear. MRI via T2-sampling perfection with application-optimized contrasts by using a flip angle evolution sequence revealed a bright signal in the cochlear endolymph but not in the perilymph. The specific sign of a missing perilymph on MRI has a diagnostic role for PLF.

KEYWORDS

perilymphatic fistula, imaging, diagnosis.

1 Background

A perilymphatic fistula (PLF) is a rare condition in which perilymph leaks from the cochlea or vestibule into the surrounding cavities, most commonly through round and oval windows, and causes cochlear and vestibular symptoms. PLF is important because it is among the few potential causes of hearing loss and vestibular disturbance that can be treated surgically. The history of PLF can be traced back to as early as 1962, when Farrior first reported that perilymph escaped through the stapes prosthesis that had entered the inner ear. In 1968, Fee reported 3 cases of dizziness, which were diagnosed as PLF through the oval window following mild head injury or a series of head injuries without concussion. Two years later, Stroud and Calcettera introduced the term “spontaneous PLF” on the basis of 4 patients, namely, a 7-year-old girl whose symptoms occurred 1 year after modified radical mastoidectomy and 3 adults with no surgical history (Stroud et al., 1970). However, vague symptoms and the lack of a clear diagnostic test have made the existence of PLF a controversial subject for decades. Cochlin-tomoprotein was detected in perilymph leakage but may also exist in the blood and cerebral spinal fluid (Ikezono et al., 2018). This test is complicated and has not yet been applied in the clinic.

In 2010, Zou et al. first reported a specific MRI manifestation mimicking perilymphatic loss in an animal study by selectively suppressing the perilymphatic signal on T2-weighted MR images via novel superparamagnetic iron oxide nanoparticles that were injected into the scala tympani of rats (Zou et al., 2010). Later, Zou

reported signs of missing perilymph on MRI in a patient with idiopathic sudden sensorineural hearing loss (Zou, 2020). Here, we report a case of definite PLF confirmed by surgery, demonstrating a specific sign of missing perilymph on MRI.

2 Case report

A 34-year-old female who visited author ZJ at the outpatient department in February 2023 with complaints of periodic headache for 20 years and persistent postural-perceptual dizziness for more than 10 years following mastoidectomy in 2004 at another hospital underwent surgery to remove the cholesteatoma in the left ear and otopyorrhea and hearing loss in the right ear (present for 3 years). She became deaf in the left ear immediately after surgery. A CT scan revealed that all the ossicular chain and semicircular canals were missing, a small vestibular cavity with a possible pneumolabyrinth appeared, the surgical cavity was occupied with soft tissue in the left ear, and the middle ear cavity and antrum were occupied with soft tissue in the right ear (Figure 1A–D). MRI via a readout-segmented echo-planar (RSEP) imaging-based technique revealed a large positive signal for cholesteatoma in the left ear and a spotted positive signal in the right ear. T2-weighted MRI revealed a nodular bright signal in the cochlea and vestibule and a large area of bright signal in the middle ear cavity, with a long tail running toward the vestibule in the left ear (Figure 2A, B). MRI via T2-sampling perfection with application-optimized contrasts by using a flip angle evolution (SPACE) sequence revealed a bright signal in the cochlear endolymph but

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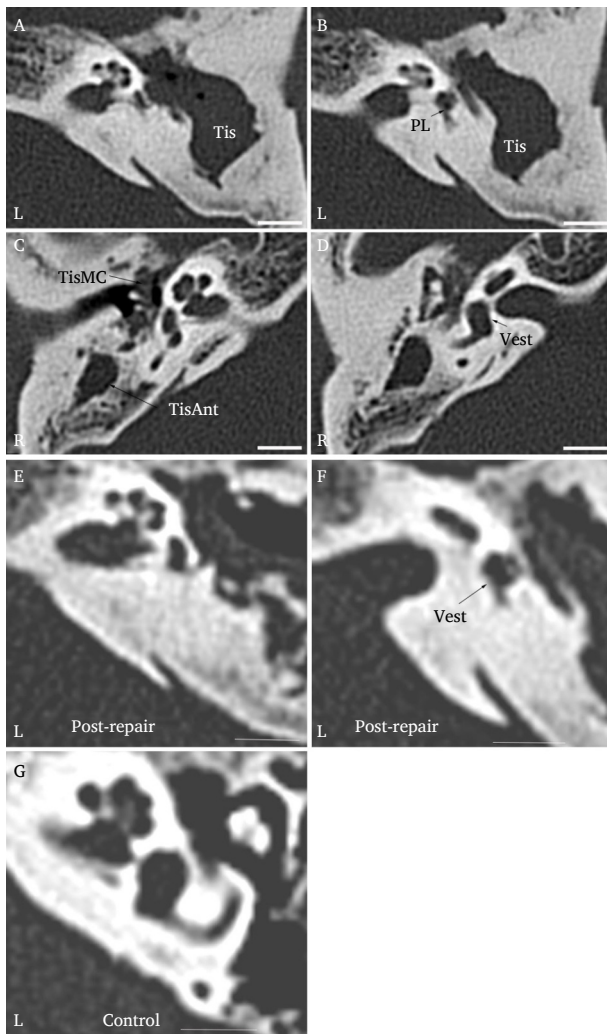


Figure 1 CT images of the temporal bone before surgery in the right ear in 2023. A possible pneumolabyrinth (PL) was detected in the vestibule (B) compared with the right side (D); no pathological changes were found in the cochlea (A and C). Seventy-six days postsurgery, the suspected PL disappeared in the left vestibule (F); no changes were detected in the left cochlea (E). A control image of a normal ear is shown (G). Tis: soft tissue; TisAnt: soft tissue in the antrum; TisMC: soft tissue in the middle ear cavity. Scale bar = 6.0 mm.

not in the perilymph, but the signal in the vestibule was faint in the left ear (Figure 2C-F).

A modified canal wall-up mastoidectomy plus tympanoplasty and meatoplasty aimed at removing the cholesteatoma in the right ear was performed through an endaural approach on June 7, 2023, with significant hearing improvement achieved in the right ear (Figure 3). However, the headache and dizziness did not resolve. A revised mastoidectomy and meatoplasty were performed in the left ear on March 20, 2024. During surgery, an obvious perforation filled with mucoid fluid was identified on the round window membrane that was repaired. Both headache and dizziness resolved after surgery, although the hearing did not recover, possibly because of delayed treatment. MRI at 4 d after round window repair revealed slight refilling of the perilymph in the left cochlea and vestibule (Figure 4). A CT scan performed at 76 d postsurgery demonstrated that the suspected pneumolabyrinth disappeared in the left vestibule (Figure 1B and F), and no changes were found in the cochlea (Figure 1A and E).

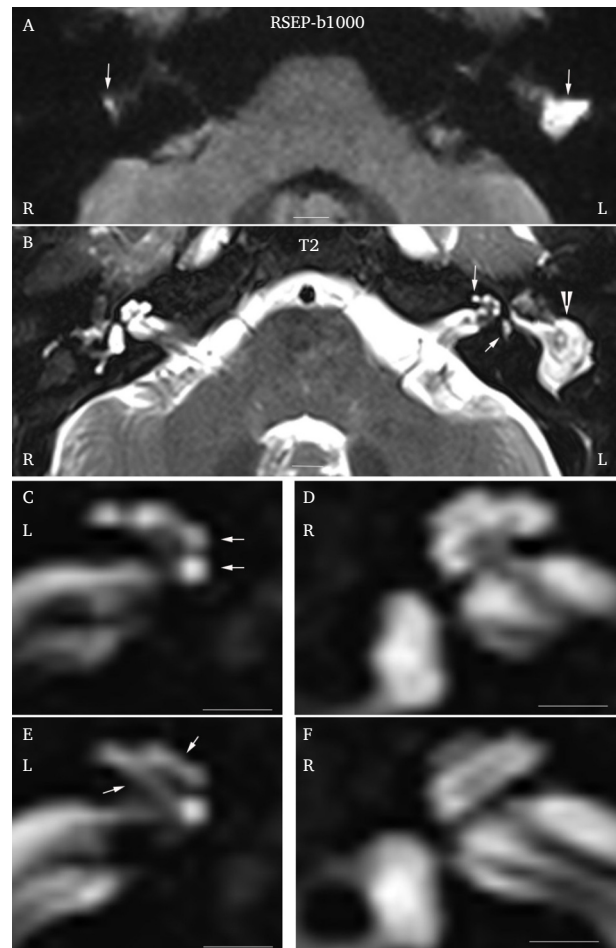


Figure 2 Non-contrast MR image of the temporal bone before surgery in the right ear in 2023. A positive cholesteatoma signal was detected on both sides via a readout-segmented echo-planar (RSEP) imaging-based technique (arrows in A). A nodular bright signal was detected in the left cochlea and vestibule (arrows in B), and a large piece of bright signal was detected in the left middle ear cavity (arrowhead in B) on the T2-weighted MR image. The bright signal in the left cochlea was confirmed as endolymph by T2-sampling perfection with application-optimized contrasts via a flip angle evolution (SPACE) sequence (arrows in C and E) compared with the right side (D and F). RSEP-b1000: b value = 1000 during imaging. Scale bars = 6.0 mm (A-B) and 3 mm (C-F).

3 Discussion

Pneumolabyrinth, or air in the cochlea, vestibule, and/or semi-circular canals on CT, has been reported as a radiological sign of PLF, but its sensitivity and specificity are limited (Mafee et al., 1984). Later, Dubrulle et al. reported that a local high signal at the level of the round window membrane on intravenous gadolinium-enhanced MR images was considered a sign of PLF (Dubrulle et al., 2020). However, the round window membrane is too thin to be exactly identified on MRI, and a local area of high signal may also be generated in the hook region of the cochlea. In the present case, MRI via the SPACE sequence demonstrated that specific signs of missing perilymph represent direct evidence of PLF, which was confirmed by surgery for the first time. Fibrosis or granulation may demonstrate a low-signal region in the inner ear, but it may also appear in the endolymphatic space. The slight refilling of the perilymph 4 d after round window repair indicates that the recovery of the perilymph circulation is slow or that the ability to produce perilymph has been compromised in PLF.

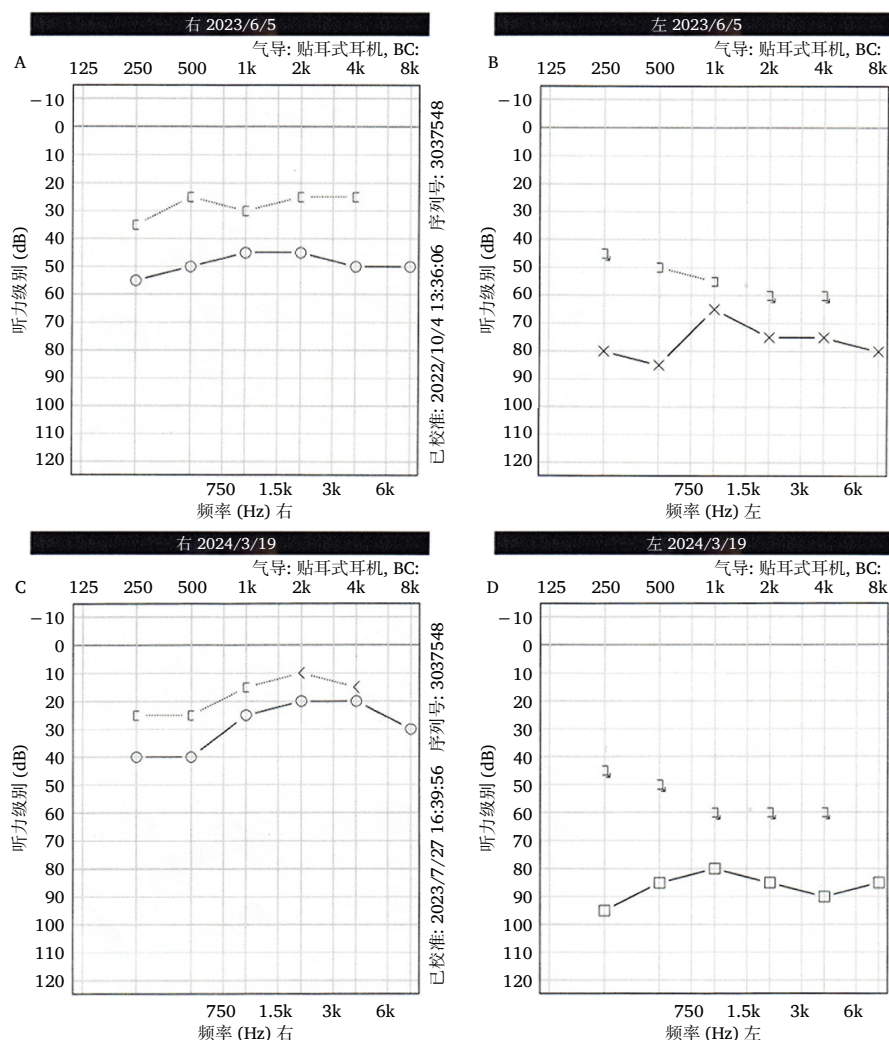


Figure 3 Pure tone audiogram before (A) and after (B) surgery in the right ear.

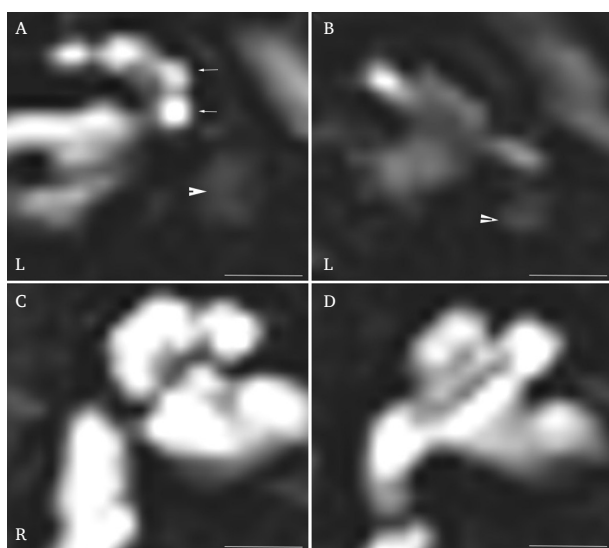


Figure 4 MRI of T2-sampling perfection with application-optimized contrasts via a flip angle evolution (SPACE) sequence of the inner ear 4 d after round window repair. Slight refilling of the perilymph in the left cochlea (arrows in A) and vestibule (arrowheads in A and B), which has not reached the same level as the right ear (C and D). Scale bar = 3.0 mm.

patients. The disappearance of the pneumolabyrinth in the left vestibule on postoperative CT images supported the MRI results. However, no pathological changes were detected in the cochlea via CT imaging, indicating that the sensitivity of CT detection of PLF is very low. In addition to its specificity and high resolution, MRI via the SPACE sequence is superior to the previous report because it avoids intravenous injection of gadolinium, which should be reduced as much as possible in the clinic owing to potential adverse effects.

4 Limitations

The image of the round window membrane was not captured because of a failed camera connection after incidental bumping on the microscope stand during surgery by a young man. However, an obvious perforation filled with mucoid fluid was identified on the round window membrane, and both headache and dizziness resolved after repair, which confirmed the existence of the PLF.

5 Conclusion

The missing perilymph sign on MRI indicates PLF. In addition to its specificity and high resolution, MRI via the SPACE sequence is superior to previously reported modalities because it avoids intravenous injection of gadolinium, which should be reduced as

much as possible in the clinic due to potential adverse effects.

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Declaration of competing interest

The authors have no competing interests to declare that are relevant to the content of this article.

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