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Case Report

MRI에서 내이의 비정상적 조영증강을 보이는 진주종성 미로 누공 1예

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MRI Findings of a Cholesteatomatous Labyrinthine Fistula Showing Abnormal Inner Ear Enhancement

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A 59-year-old male patient presented with sudden onset of vertigo and hearing loss. Labyrinthitis due to lateral semicircular canal fistula caused by cholesteamatous otitis media was suspected from temporal bone computed tomography (CT) and clinical symptoms. The patient was treated with canal wall down mastoidectomy with removal of the cholesteatoma and lateral semicircular canal occlusion. Preoperative gadolinium-enhanced magnetic resonance imaging (MRI) images of the inner ear revealed increased signal in the cochlea as well as vestibule. Correlation of the MRI findings and the inner ear involvement in labyrinthine fistula is discussed.

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Key Words: Chronic otitis media; Labyrinthine fistula; MRI

Introduction

Introduction of newer antimicrobial treatments and advanced diagnostic technology has decreased the incidence of serious complications of cholesteatomatous chronic otitis media. Labyrinthine fistula remains a common complication, with frequency of 4-12%.[1-4] Lateral semicircular canals are the most common site of the fistula, although other sites of the vestibule or cochlea may be involved. Confirmation of labyrinthine fistula can be made intraoperatively and surgical management is required. Nevertheless, preoperative evaluation is mandatory. Clinical signs of sensorineural hearing loss and/or vertigo and the fistula test render limited information regarding inner ear involvement. Accurate diagnosis of bony defect in labyrinthine fistula due to cholesteatoma is facilitated by routine preoperative imaging study using high resolution computed tomography (HRCT).[5,6] However, HRCT only suggests the extent of inner ear disturbance in labyrinthine fistula patients. Magnetic resonance imaging (MRI) can determine the changes in fluid composition such as cerebrospinal fluid, and enhanced visualization of smaller structures such as the inner ear became more feasible. We report a case of labyrinthine fistula of the lateral semicircular canal caused by cholesteatomaous otitis media, and discuss correlation of the MRI findings and the inner ear disturbance.

Case Report

A 59-year-old Korean male patient with a longstanding history of chronic otitis media presented with acute onset vertigo. His chief

complaints were complained of severe rotatory dizziness and aggravation of hearing loss in the right ear. Otoscopic examination revealed a large-sized central perforation of right tympanic membrane with pulsatile otorrhea (Figure 1A). On physical examination, left beating horizontal nystagmus was observed under Frenzel's glasses. Pure tone audiograms revealed deafness in the right ear with normal hearing in the left ear (Figure 1B). Temporal bone computed tomography (CT) scans revealed a large cholesteatoma filling antrum and middle ear cavity and bony erosion over the lateral semicircular canal in the right ear (Figure 2). Temporal MRI was performed using Achieva, 3.0T (Phlilps Medical System, Best, The Netherlands). Cholesteatoma induced inflammation in the middle ear cavity was noted to communicate with inner ear fluid-filled space in T2-weighted images (Figure 3A). T1 weighted images obtained 5 minutes after intravenous gadolinium administration, with slice thickness of 2 mm, revealed areas of pathological enhancement in the lateral canal and cochlea on the right side compared to the left side (Figure 3B). Prominent enhancement of lateral canal and entire length of cochlea was identified more clearly in when T1 weighted images were obtained with slice thinckness of 1 mm and no interslice gap, to enhance resolution (Figure 3C). After a week of intravenous antibiotics and conservative care, subjective dizziness ameliorated and otorrhea decreased. Closed-loop Caloric test was performed after spontaneous nystagmus subsided, and caloric response was severely decreased on the right side (right side canal paresis 86%) (Figure 4). The patient underwent canal wall down mastoidectomy with removal of the cholesteatoma and lateral semicircular canal occlusion. A huge cholesteatoma filled the mastoid antrum and middle ear cavity. Careful removal of cholesteatoma matrix

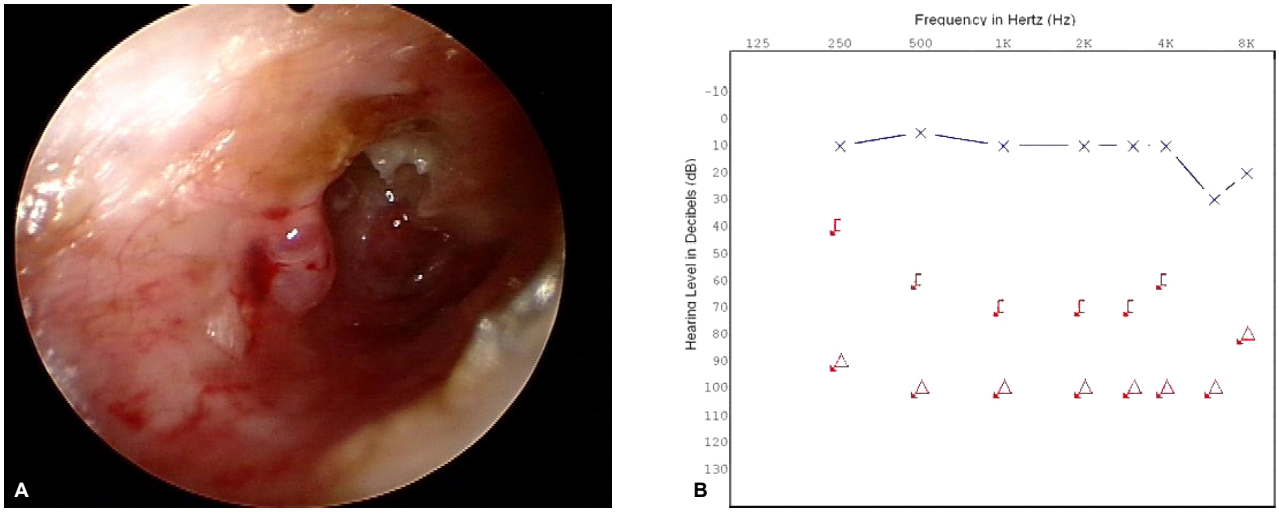

Figure 1. At initial presentation, otoscopic examination revealed a large-sized central perforation of right tympanic membrane with pulsatile otorrhea (A), and pure tone audiograms revealed deafness in the right ear with normal hearing in the left ear (B).

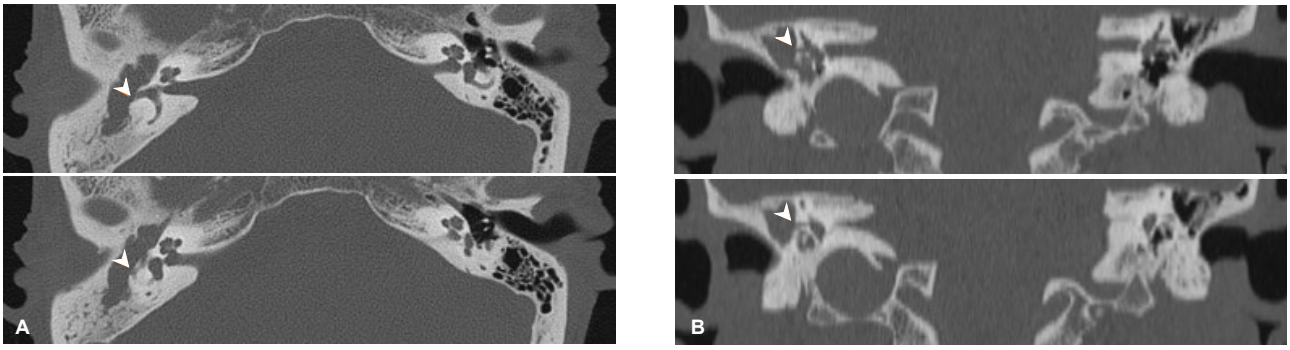

Figure 2. Computed tomography (CT) of the temporal bone revealed a fistula of the right lateral semicircular canal related to cholesteatoma. Arrowheads indicate the defect in the labyrinthine bone surrounding the lateral semicircular canal in axial (A) and coronal (B) images.

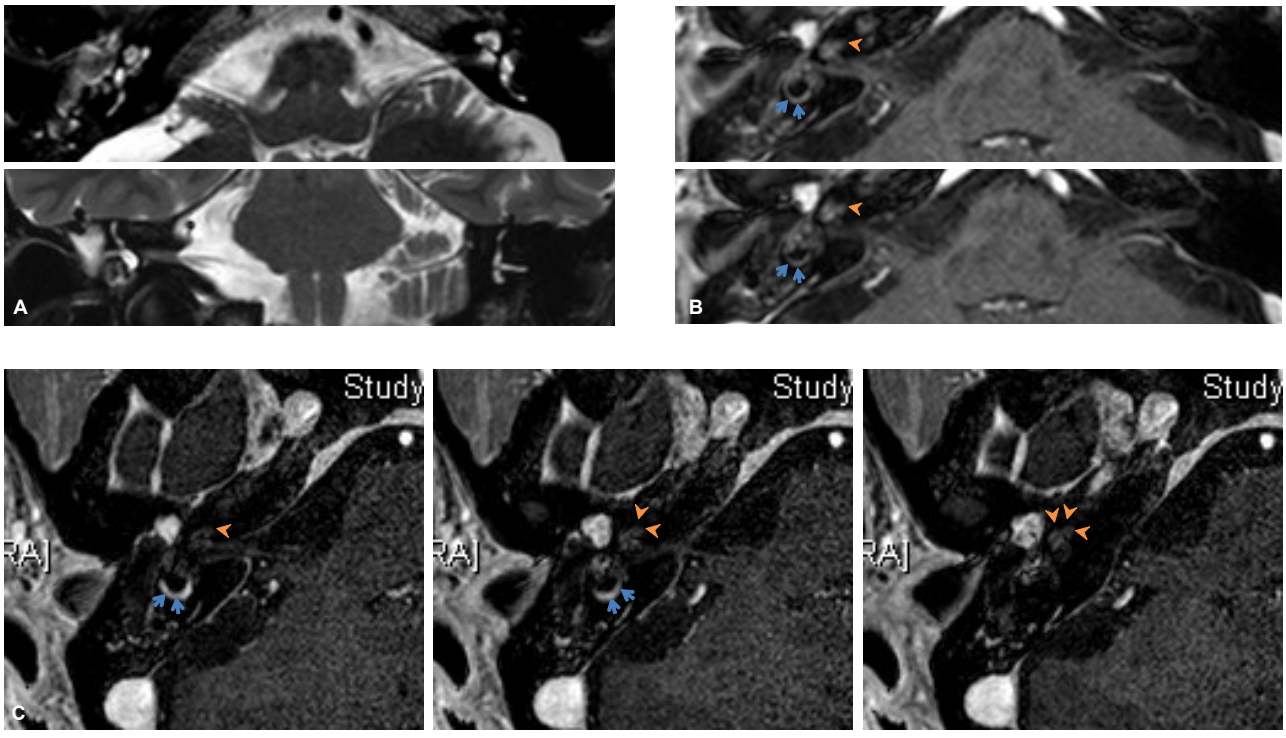

Figure 3. Magnetic resonance imaging (MRI) findings. (A) T2-weighted images revealed cholesteatoma induced inflammation in the right middle ear with abnormal communication with inner ear fluid-filled space. (B) Routine T1 weighted images after intravenous gadolinium administration revealed areas showed pathological enhancement in the lateral canal and cochlea on the right side. (C) High-resolution T1 weighted scans demonstrate prominent enhancement of lateral canal and entire length of cochlea more clearly (Cochlea, indicated by arrowheads; lateral semicircular canal, arrows).

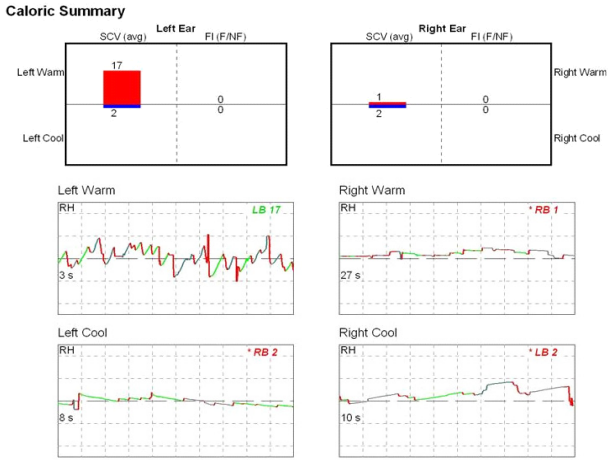

Figure 4. Caloric response in the right ear was severely decreased (canal paresis 85%) on closed-loop caloric test. SCV, slow component velocity; FI, fixation index; F/NF, fixed/not fixed; RH, right horizontal; LB, left beating; RB, right beating; S, second; Avg, average.

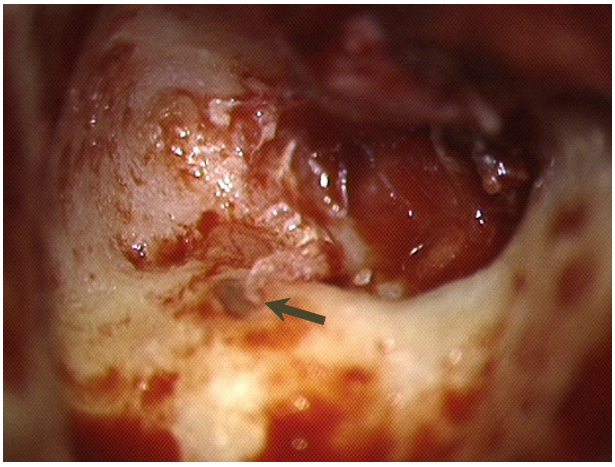

Figure 5. About 4 mm-sized fistula over lateral semicircular canal was identified. Cholesteatoma matrix covering the opening was adherent to membranous labyrinth (arrow) with extension into the lateral canal.

revealed erosion of facial nerve bony canal over the length of tympanic segment. The ossicles were eroded and only stapes footplate was identified. About 4 mm-sized fistula in the otic capsule over lateral semicircular canal was found. When cholesteatomatous matrix was carefully peeled away from the fistula site, it was found to be adherent to the membranous labyrinth with extension into the

canal (Figure 5). A portion of adherent membranous labrytinth was removed with the matrix and the lateral semicircular canal was obliterated using pieces of temporalis muscle fascia. The opening was covered with bone dust and fibrin glue. After the surgery, the patient complained of aggravation of dizziness which subsided over about 2 months. The patient's hearing showed no change postoperatively.

Discussion

Clinical manifestations of labyrinthine fistula in cholesteatomatous chronic otitis media vary from asymptomatic to severe dysfunction of cochleovestibular system. Patients usually present with varying degrees of hearing loss and otorrhea. Accompanying symptoms include vertigo and tinnitus[4] Surgical treatment entails complete removal of cholesteatoma and reliable management of fistula (exteriorization or seal-off), and aims to preserve of inner ear function. To prevent the risk of surgery-related sensorineural hearing loss upon removal of cholesteatoma matrix covering the fistula opening, clinical suspicion and preoperative evaluation using HRCT are recommended.[1,2,4,6,7] Recent studies using three dimensional fluid-attenuated inversion recovery (3D-FLAIR) sequence in MRI have reported enhanced visualization of the fluid-filled inner ear in patients with various inner ear diseases.[8-10] In patients with labyrinthine fistula due to cholesteatoma, 3D-FLAIR images have shown areas of high signal intensity after intravenous gadolinium administration in the inner ear.[9] The authors have suggested that involvement of inner ear fluid-filled space results in high signal intensity on images due to the breakdown of blood-labyrinthine barrier (BLB). Likewise, in our patient, a fistula over lateral semicircular canal due to middle ear cholesteatoma was identified from HRCT, which would explain the presentation of sudden onset vertigo and hearing loss. Loss of both cochlear and vestibular function was evident from the audiometry and closed-loop caloric test results. We performed MRI scans to evaluate inner ear involvement using specific sequence to enhance resolution. Compared to routine temporal MRI scans, the fluid-filled inner ear space is delineated more clearly using thinner slice thickness of 1mm for more comprehensive assessment. After intravenous gadolinium injection, routineT1 images showed abnormal enhancement of the lateral semicircular canal and cochlea in the right ear compared to the normal side (Figure 3B). High resolution scans demonstrate inner ear fluid in the lateral semicircular canal and the entire length of cochlea showed signal intensity higher than brain parenchyme, suggesting disruption of inner ear fluid homeostasis.

A classification of labyrinthine fistulas has been proposed according to the extent or depth of involvement of the labyrinth.[1] Type I fistula is considered to be an erosion of the bony labyrinth with an intact endosteum. Type II fistula is a true fistula with an opened perilympharic space. Type III fistula is an opened perilymphatic space with concomitant involvement or destruction of the underlying membranous labyrinth. Sensorineural hearing loss and vertigo are observed more often in type II and III, which implies the presence of inner ear dysfunction. Anatomical stages of labyrinthine fistula may be correlated with MRI findings. For example, it would be unlikely to find any abnormality in the inner ear fluid in type I fistula limited to the bony labyrinth. On the other hand, inflammation and subsequent fibrosis of the membranous labyrinth due to labyrinthitis can result in obliteration of the inner ear fluid-filled space and non-visualization on T2 images. Also, abnormal enhancement after gadolinium administration may be expected in cases of the BLB breakdown, even without direct penetration of membranous labyrinth. In our patient, disruption of bony labyrinth was evident from CT scans and T2-weighted MRI images showed presence of inner ear fluid in the bony canal of the lateral semicircular canal, suggesting acute involvement of the inner ear rather than a chronic process. T1-weighted MRI images taken 5 minutes after gadolinium administration revealed increased signal intensity not only in the lateral semicircular canal in direct connection with middle ear cholesteatoma, but also in the entire length of cochlea, and it can be presumed that compositional changes in the inner ear fluid was not limited to the vestibular organ. Involvement of the cochlea fluid-filled space observed in the contrast-enhanced MRI scans corresponds to loss of hearing in our patient, and it may predict poorer hearing outcome. Further studies utilizing refined imaging techniques may render more information to explain the mechanism underlying progressive inner ear dysfunction in labyrinthine fistula.

Conclusion

In conclusion, preoperative evaluation of gadolinium-enhanced

MRI of the inner ear as well as temporal bone CT and audio-vestibular tests provides additional information regarding inner ear involvement in labyrinthine fistula due to middle ear cholesteatoma.

중심 단어: 만성중이염, 미로누공, 자기공명영상

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