Sensorineural Hearing Loss: More than Meets the Eye?

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PURPOSE: To assess the value of MR in patients with sensorineural hearing loss (SNHL) caused by lesions other than acoustic neuromas. METHODS: MR studies of 51 patients with SNHL were retrospectively reviewed; patients with acoustic neuroma were excluded to focus on the more uncommon causes. RESULTS: Twenty patients had labyrinthine lesions. Six patients had viral labyrinthitis, one patient had bacterial labyrinthitis, and one patient had luetic labyrinthitis. Three patients had hemorrhage in the labyrinth, two posttraumatic and one spontaneous from an adjacent temporal bone tumor. Only one of the two patients with traumatic labyrinthine hemorrhage had evidence of a fracture on high-resolution CT. In one patient with CT-proved cochlear otosclerosis, pericochlear foci of enhancement were seen on contrast-enhanced MR. Four patients had presumed labyrinthine schwannomas. A middle ear cholesteatoma in one patient invaded the cochlea and resulted in marked cochlear enhancement due to granulation tissue. Thirteen patients had intracanalicular and cerebellopontine angle lesions. The lesions included arteriovenous malformations (three patients), sarcoidosis (three patients), metastasis (two patients), lymphoma (two patients), lipomas (two patients), and postshunt meningeal fibrosis (one patient). Eighteen patients had intra-axial lesions responsible for SNHL. The most common intra-axial lesions were brain stem infarcts and multiple sclerosis. Traumatic lesions in the inferior colliculi, sarcoidosis, lymphoma, and extrinsic compression of the colliculi from a pineal tumor were also noted. CONCLUSION: MR can demonstrate numerous lesions responsible for SNHL other than acoustic neuromas. The entire acoustic pathways, including the labyrinth, internal auditory canal, cerebellopontine angle, and brain stem should be carefully scrutinized for lesions in patients with SNHL. The use of contrast media markedly increases the yield of MR in this clinical situation by demonstrating inflammatory and neoplastic labyrinthine lesions and meningeal pathology (both neoplastic and inflammatory) in the internal auditory canal and cerebellopontine angle cistern.

Index terms: Cerebellopontine angle, neoplasms; Brain stem, neoplasms; Hearing, loss; Temporal bone, magnetic resonance

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Sensorineural hearing loss (SNHL) due to internal auditory canal/cerebellopontine angle (IAC/CPA) lesions, especially acoustic neuroma, has been extensively described (1–14). Contrast-enhanced magnetic resonance imaging (MR) has made a major impact in the detection and diagnosis of acoustic neuroma, meningioma, and epidermoid of the IAC/CPA area (15–20). While the evaluation of the CPA and the IAC has received a great deal of attention, little emphasis has been paid to the identification of the more uncommon lesions causing SNHL. Lesions within the bony and membranous labyrinth and the more proximal acoustic pathway have received the least amount of attention (21–25).

Recently a series of patients with inflammatory lesions of the membranous labyrinth in whom the labyrinth enhanced during contrast-enhanced MR were reported (25, 27). This report reemphasized the importance of carefully evaluating images of the entire acoustic pathway in patients with SNHL. In the present report, we describe our MR experience with lesions of the bony and membranous labyrinth producing SNHL. In addition, we will discuss new observations regarding the more
uncommon lesions of the IAC/CPA area and intraaxial acoustic pathway.

Although patients with SNHL most commonly have either normal MR or acoustic neuroma, careful scrutiny of the bony and membranous labyrinth and each of the components of the central acoustic pathway is necessary to identify many of the more unusual causes of SNHL on MR imaging (21).

Material and Methods

MR studies of 51 patients with SNHL were retrospectively evaluated in this study. Patients were from multiple institutions; their ages ranged from 3 to 79 years. Cases of acoustic neuroma, meningioma, and epidermoidoma were excluded from the study group to focus on the more uncommon causes of SNHL. The imaging protocol utilized included pre-gadolinium sagittal T1-weighted images (300/20), post-gadolinium axial T1-weighted images (600/20) with 3-mm thick sections on 0 or 0.5-mm intersection gap, 192 or 256 X 256 matrix, 14 to 18-mm field of view, and 2-4 excitations, and axial proton density and 5-mm thick, 2.5-mm gap T2-weighted images (2000/30, 80) through the whole brain. Thirty-eight patients also underwent pre-gadolinium T1-weighted imaging through the posterior fossa in addition to the above sequences, while 24 patients underwent post-gadolinium coronal T1-weighted imaging (600/20) with 3-mm thick sections.

Two neuroradiologists reviewed all studies according to a specific protocol, involving evaluation of the bony and membranous labyrinth, the IAC, the CPA cistern, the brain stem, and the superior temporal gyrus.

Results

The results are summarized in Table 1. Twelve patients have been included in previous articles (25, 27). Twenty patients had labyrinthine lesions. Six patients had viral labyrinthitis (Figs. 1 and 2), one patient had bacterial labyrinthitis from adjacent middle ear infection (Fig. 3), whereas one patient had luetic labyrinthitis. In two of these patients with inflammatory labyrinthitis, isolated enhancement of the cochlea was present (Fig. 1). In the remainder, both the cochlea and the vestibule enhanced on the symptomatic side (Fig. 2).

Three patients had hemorrhage in the labyrinth, two posttraumatic (Fig. 4) and one spontaneous from an adjacent temporal bone adenomatous tumor (Fig. 5). Only one of the two patients with traumatic labyrinthine hemorrhage had evidence of a fracture on high-resolution computed tomography (CT).

In one patient with CT-proved cochlear otosclerosis, MR demonstrated pericochlear foci of enhancement corresponding to the lytic area seen on CT (Fig. 6). Four patients had presumed la-
Fig. 3. Bacterial labyrinthitis. Middle-aged man with left middle ear infection, acute hearing loss, and facial nerve paralysis. Gadolinium-enhanced axial T1-weighted image (600/20) demonstrates enhancement of the left cochlea (arrow) and tympanic segment of the facial nerve (open arrow). Adjacent mastoiditis is seen as high signal in the mastoid air cells (curved arrow).

Fig. 2. Labyrinthitis presumed to be viral in a patient 2 weeks following acute onset of SNHL.
A. Enhanced T1-weighted (600/20) axial image of the right temporal bone shows enhancement of the first (long arrow) and second (short arrow) turns of the cochlea as well as the vestibule (arrow; V).
B. Normal left ear taken with same photographic technique shows nonenhancing cochlear first (long arrow) and second (short arrow) and vestibule (arrow; V).

Byrinnhine schwannomas. Three patients had vestibular schwannomas (Fig. 7) and one patient had a small acoustic neuroma extending into the basal turn of the cochlea.

A middle ear cholesteatoma in one patient invaded the cochlea, resulting in both marked cochlear enhancement and enhancement of the meninges in the IAC and CPA secondary to surgically proved granulation tissue (Fig. 8). Three patients had postmeningitis labyrinthine ossifications (Fig. 9). No fluid signal from the membranous labyrinth was seen in these cases.

Thirteen patients had IAC/CPA lesions. Three patients had arteriovenous malformation (Fig. 10). One of the three had associated acute intraxial hemorrhage. Three cases of sarcoidosis were identified (Fig. 11). Two metastatic lesions and two cases of lymphoma were seen. Two lipomas and one case of postshunt meningeal fibrosis were also identified.

Eighteen patients with intraaxial lesions thought to be responsible for SNHL were found. The most common intra-axial lesions were brainstem infarct (Fig. 12A) and multiple sclerosis (Fig. 12B). One case of multiple sclerosis, in addition to having the typical findings of periventricular lesions, had dramatic enhancement of the cochlear membranous labyrinth on the side of the hearing loss (Fig. 12C). Traumatic changes along the central acoustic pathway also caused SNHL (Fig. 13).

Discussion

SNHL remains a challenging clinical-radiologic problem despite the existence of a broad range of clinical and radiologic tests that may be employed in the search for precise causes of this symptom. MR imaging plays a pivotal part in the workup of such patients. The MR study in the patient with SNHL is necessarily focused on the IAC and CPA since the acoustic neuroma, the most statistically common cause of this problem, is found there. Although the search in this area for acoustic neuroma remains central to the radiologic task, multiple other diseases cause SNHL (21–27). Many of these disease processes are
Fig. 4. MR in patient 3 days following head trauma with SNHL.

A, Coronal nonenhanced T1-weighted MR image (600/20) shows high signal (arrow) presumed to represent methemoglobin in the vestibular aspect of the membranous labyrinth.

B, Enhanced T1-weighted MR image (600/20) again shows high signal within the two turns of the cochlea (arrow).

Fig. 5. Adenomatous tumor within medial temporal bone fistulizes membranous labyrinth with subacute blood in vestibule.

A, Axial CT image of the left temporal bone at the level of the vestibule (arrow) shows the adenomatous tumor (open arrow) along its medial surface.

B, T1-weighted axial MR image (600/20) at the same level as A reveals the mixed signal tumor (open arrow) with high signal within the vestibular membranous labyrinth (arrow) presumed to represent methemoglobin.

found in the labyrinth and along the more central intraaxial acoustic pathway. Only with systematic evaluation of the entire acoustic pathway from the labyrinth level to the superior temporal gyrus centrally will these lesions be discovered in the search for causes of SNHL (21).

The area of the bony and membranous labyrinth is commonly overlooked in the evaluation of the MR images in a patient with SNHL. Although high-resolution MR imaging of this area has been available for a number of years, only recently have any reports of pathology in the labyrinth been reported (22–27). Although lesions of the bony labyrinth are probably best seen with high-resolution CT, those affecting the membranous labyrinth will only be seen with focused MR imaging. Inflammation from any cause, hemorrhage, and smaller intralabyrinthine schwannoma are the three lesions of the membranous labyrinth that can be seen only by MR imaging (22–27).

Inflammatory lesions of the membranous labyrinth have primarily been reported in the pathologic literature.

The eight cases of inflammatory lesions of the membranous labyrinth reported in this study all demonstrate varying degrees of enhancement of the membranous labyrinth parts that could not be seen with CT imaging. The case of bacterial labyrinthitis resulted from the spread of middle ear infection, whereas luetic labyrinthitis was seen as part of systemic syphilis. Six cases of presumed viral labyrinthitis in which contrast-en-
enhanced MR demonstrated enhancement of the membranous labyrinth were seen. In these patients, the enhancement always correlated with severely impaired labyrinthine function. The presence of enhancement was highly specific for labyrinthine injury, but its sensitivity remains unknown. It is likely that many patients with mild labyrinthitis will not show enhancement (25, 27).

Hemorrhage in the membranous labyrinth most commonly results from fractures through the temporal bone region. Because the fracture may not always be identifiable on CT, MR imaging may be the only imaging study sufficient to explain the patient’s hearing loss. High-resolution CT remains the study of choice in the setting of suspected temporal bone fracture (26, 28). However, if CT does not explain the patients’ symptoms (hearing loss, vertigo, facial nerve paralysis), MR has an important adjunctive role.

When a temporal bone tumor fistulizes the membranous labyrinth, it may hemorrhage into this space. Spontaneous hemorrhage is rare but can be readily diagnosed by unenhanced MR. In any case, high signal seen within the membranous labyrinth in the absence of contrast enhancement probably represents subacute blood. In our limited experience, this high signal seems to persist up to months after the original insult producing the hemorrhage.

Labyrinthine schwannomas have, until recently, been reported only in the pathologic literature. Two recent radiologic articles have demonstrated such lesions in vivo, both on unenhanced and enhanced studies (22, 23). This diagnosis can be suggested in the case of a patient with long-standing symptoms that do not remit over time, where a focal mass is seen within the labyrinth. The schwannoma is of higher signal intensity than normal membranous labyrinth fluid signal on nonenhanced MR images and dramatically enhances when contrast is administered.
Labryrinthinne ossificans. Axial T1-weighted MR image (600/20) of the right ear in a profoundly deaf patient following an episode of meningitis shows no signal where the cochlear membranous labyrinth would be expected to be (arrows). Modiolus -small arrow.

Arteriovenous malformation of the CPA cistern. Axial T2-weighted MR image (2600/80) of the low CPA cistern reveals the malformation nidus (arrow) within the cistern, compressing the inferior cerebellar peduncle (open arrow) where the cochlear nuclei are known to reside. n = normal opposite inferior cerebellar cistern.

This case can be compared with that of a patient with labyrinthitis, where the fluid signal within the membranous labyrinth is normal before contrast administration and more subtle, nonfocal enhancement is seen after contrast is given. However, only follow-up imaging demonstrating persistence of an enhancing focus within the membranous labyrinth with interval growth can definitively indicate the presence of schwannoma.

In the case of cochlear otosclerosis and labyrinthine ossificans, both CT and MR can identify the changes in the bony labyrinth that support these diagnoses. In active cochlear otosclerosis, CT shows lytic areas in the perilabyrinthine bone blurring the normally sharp margins of the bony labyrinth. The MR case of pericochlear enhancement in a patient with CT-proved cochlear otosclerosis suggests an inflammatory process in the bone, resulting in leakage of the contrast from the vascular spaces. To our knowledge, pericochlear enhancement in cochlear otosclerosis has not been previously reported. All three cases of labyrinthine ossificans revealed ossification of the membranous labyrinth on both CT and MR. The MR diagnosis is easily missed unless a search for the missing fluid signal of the membranous labyrinth is undertaken.

Most enhancing lesions in the IAC are acoustic neuromas. With focused MR imaging, this diagnosis is easily made in most cases. Care must be taken not to misdiagnose other causes of enhancement in the IAC as "early acoustic neuroma" (30). The converse is also true; lesions that appear as more linear "neuritis" may transform to more globular acoustic neuroma over time (Fig. 14). Hemangioma and arteriovenous malformation are two lesions that may enhance in the IAC, thereby mimicking acoustic neuroma. Furthermore, because they can present with hemorrhage, precontrast T1-weighted images become important in study interpretation to differentiate enhancement from subacute hemorrhage.

Lipomas of the IAC can also mimic enhancing lesions and will be readily diagnosed on the precontrast T1-weighted images or on the fat-suppression images. The meninges may enhance in a variety of inflammatory (sarcoid, postshunt meningeal fibrosis) and neoplastic (lymphoma, metastasis) conditions (30-33). Symptomatic CNS sarcoidosis has been reported in 5% of patients with sarcoidosis, and meningeal involve-
Fig. 12. Intraaxial causes of SNHL.
A, T2-weighted axial MR image (2600/80) in a patient with acute onset of bilateral hearing loss worse in the left ear reveals a pontine cerebrovascular accident (open arrow).
B, A multiple sclerosis plaque ($P$) is identified on this T2-weighted MR axial image (2600/80) at the level of the pons.
C, Another multiple sclerosis patient with right-sided hearing loss shows enhancement of the cochlear aspect of the membranous labyrinth (arrow).

Fig. 13. Tectal contusion causing bilateral SNHL following trauma. Contrast-enhanced axial T1-weighted MR image (600/20) demonstrates bilateral enhancing contusions of the inferior colliculi (arrows). The precontrast images were normal.

Intraaxial lesions proximal to the cochlear nuclei on the dorsolateral medulla generally present with bilateral SNHL, worse on the side opposite the lesion (21). The more proximal the lesion along the central acoustic pathway, the more difficult it is to be sure the lesion is causing the hearing loss. The intraaxial lesions included in our series underscore the importance of obtaining CPA cistern. The thickened, enhanced meninges may touch each other in the midline, mimicking an acoustic neuroma. It is important to scrutinize the remainder of the meninges in the posterior fossa and supratentorial level to find other evidence of meningeal enhancement that would point to the correct diagnosis. Repeat MR examination after steroids can demonstrate marked reduction in the enhancement (as seen in one of our patients) and convert the homogeneously enhancing lesion filling the IAC into two thin layers of enhancement along the meninges of the IAC and/or CPA.

Intraaxial lesions proximal to the cochlear nuclei in eight of 12 patients in a recent article (33). The enhancement may involve only the IAC or extend into the IAC from the adjacent
a whole brain T2-weighted MR sequence as part of the imaging protocol for patients with SNHL. Any intraaxial disease process that disrupts the normal function of the ascending fibers within the central acoustic pathway may cause SNHL. Primary and metastatic brain tumor, stroke, vascular malformations, and multiple sclerosis make up the bulk of lesions that present at least in part with SNHL (21). In the case of tumor, stroke, and vascular malformations, the lesion is usually found directly along the ascending fibers of the central acoustic pathway. In contrast, multiple sclerosis may be seen only as plaques within the supratentorial white matter, with the plaque causing the hearing loss presumably remaining too small to be visible by present MR techniques (24).

Because of the retrospective nature of our study, it was not possible to determine what proportion of patients with SNHL had an identifiable cause by contrast MR. Clearly, despite these recent advances in imaging, a large number of cases of SNHL remain unexplained.

In conclusion, any MR imaging search for a cause of SNHL must include careful scrutiny of both the labyrinthine components and the pieces of the central acoustic pathway. Only with this complete MR examination and careful inspection of the images produced will the multiple nonacoustic causes of hearing loss be identifiable.

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