AJNR

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AJNR Am J Neuroradiol 1998, 19 (8) 1433-1436
http://www.ajnr.org/content/19/8/1433

AJNR Am J Neuroradiol 19:1433–1436, September 1998

Sudden Hearing Loss: Frequency of Abnormal Findings on Contrast-Enhanced MR Studies

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BACKGROUND AND PURPOSE: **Our purpose was to determine the frequency of abnormal findings on contrast-enhanced high-resolution MR imaging studies in patients with sudden hearing loss.**

METHODS: **Seventy-eight consecutive patients with sudden hearing loss underwent contrast-enhanced MR imaging of the temporal bone, cerebellopontine angle, and brain. Additional tests included audiologic examination, electrocochleography, fistula tests, and serologic tests for viral agents and autoimmune disorders.**

RESULTS: **Probable causes of the sudden hearing loss in these patients included viral or immune-mediated disease, Meniere disease, vascular disorder, syphilis, neoplasm, multiple sclerosis, and perilymphatic fistula. Twenty-four (31%) of the 78 patients were found to have abnormal imaging results early in the course of their work up and treatment.**

CONCLUSION: **The prevalence of abnormal findings on contrast-enhanced MR studies is higher than previously reported in patients with sudden hearing loss.**

The diagnosis of sudden hearing loss (SHL) has engendered much controversy in the literature (1–5). SHL has been described as "a sensorineural hearing loss with at least a 30-dB decrease in threshold in three contiguous test frequencies occurring over a 24- to 72-hour period" (6). The debate has centered on both the origin and treatment of SHL, which is understandable given the multiplicity of causes of this syndrome (6, 7), which makes it impossible for any one treatment to be effective for every patient.

In most cases, the exact probable cause of SHL remains unknown; however, a specific diagnosis is especially critical in the first 7 to 14 days, when treatment is most helpful. Therefore, any diagnostic test that could narrow the possibilities within the first few days of onset would be highly desirable. Contrast-enhanced high-resolution MR imaging has become the imaging method of choice for the detection of acoustic neuromas, and it has recently been used to detect the presence of inflammatory conditions in the labyrinth (8–12).

The most commonly reported presumed cause of enhancement of the cochlea to date has been viral infection (9–12), but immune-mediated inner ear disease (10, 13) and perilymphatic fistulas (10) have also been suggested as a possible cause of labyrinthine enhancement. When enhancement is limited to the endolymphatic sac, a specific diagnosis cannot be made, but this finding has been associated with Meniere disease (11). While advances in imaging have helped our understanding of sensorineural hearing loss (SNHL) in specific cases in which contrast-enhanced MR studies have been positive, the prevalence of positive studies in these patients has not been reported. The purpose of our study was to determine the frequency of abnormal MR imaging findings in a series of consecutive patients with acute SNHL to asses the diagnostic yield of this imaging technique.

Methods

Over a period of 6 years, from January 1989 through January 1995, we examined and treated 173 patients with SHL. Patients were eliminated from this study if the cause of their SHL (eg, herpes zoster oticus, traumatic perilymphatic fistula, preexisting Meniere disease, etc) was determined after the initial history and physical examination were completed. Patients who had no contrast-enhanced MR temporal bone study performed or interpreted by the authors were also excluded. In the end, 78 cases were available for a comparison of presumed diagnosis, degree of initial hearing loss, and imaging findings. The patients ranged in age from 13 to 79 years, with a median age of 45 years. Thirty-eight patients were women and 40 were men.

The MR protocol included standard high-resolution sequences through the labyrinth in the axial and coronal projections. T1-weighted (600/13/4 [TR/TE/excitations]) and T2-weighted (2400/30, 80/1) images through the whole brain were obtained before and after contrast administration with a 14-cm field of view, a 256 × 192 matrix, and 3-mm-thick sections. The images were obtained on a 1.5-T imaging unit with a quadrature head coil. No phase-array coils were used.

Additional diagnostic examinations usually included subjective and platform fistula tests; electronystagmography; electrocochleography; acute and convalescent antibody titers for her-

Received October 23, 1997; accepted after revision March 11, 1998.

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pes zoster, measles, mumps, rubella, cytomegalovirus, and influenza A and B; and an autoimmune screen (sedimentation rate, antinuclear antibodies, rheumatoid factor, quantitative immunoglobulins, and C1q binding factor).

Except for the patients with acoustic neuromas who underwent surgery, the following presumptive causes of SHL were established indirectly:

Viral Labyrinthitis/Cochleitis.—This diagnosis was made most confidently when a patient showed an initial rise in an antibody titer on the acute test and a fourfold decrease at the 1- or 2-month convalescent test. These are accepted criteria for the diagnosis of a variety of viral illnesses. A viral cause was also suspected if a patient had an obvious viral illness within 2 weeks of onset of SHL and no other probable cause was found. Two patients who had viral booster shots within 2 weeks of SHL onset were believed to have a probable viral cause.

Meniere Disease.—This diagnosis was made if the patient subsequently experienced a fluctuating hearing loss with episodic vertigo and tinnitus. An electrocochleogram with a summating potential/action potential ratio of more than .50 was considered strongly suggestive of endolymphtic hydrops in this group (14).

Immune-Mediated Labyrinthitis.—This diagnosis was suspected when serologic tests were positive for a known autoimmune disease, when an abnormal lymphocyte transformation test for inner ear tissue was obtained, or when there was clinical evidence of a defined systemic autoimmune disease (eg, Sjögren disease with a positive tissue biopsy.

Vascular Disease.—This diagnosis was made if there was imaging evidence of a brain infarct, if vascular compression could be demonstrated by a combination of MR imaging and MR angiography, or if the patient was over 70 years of age with known cerebrovascular disease and no other etiologic factor was found. The vascular compression syndrome is somewhat controversial, as a number of asymptomatic patients may show similar radiologic findings.

Syphilis.—These patients had clear serologic evidence of active syphilis.

Schwannoma.—These patients had typical contrast-enhanced MR imaging evidence of a schwannoma. Three were found, and all were surgically removed.

Multiple Sclerosis.—These patients had typical findings of multiple sclerosis on contrast-enhanced MR studies, a pontine lesion, and no previous diagnosis of multiple sclerosis.

Perilymphatic Fistula.—Two patients had no historical data to indicate trauma but their workups showed a positive platform pressure test in the ear with SHL. For logistical reasons, this test was usually performed 1 to 2 weeks after the initial examination.

Unknown.—Finally, when none of the above criteria were met, the cause of SHL was classified as unknown (idiopathic).

Results

The Table compares the presumed diagnosis (probable cause) of SNL with findings on the contrast-enhanced MR studies. Twenty-four (31%) of 78 patients presenting with SHL had abnormal imaging findings that most likely accounted for the patient's symptoms. Eighteen (75%) of 24 patients with an abnormal MR study had severe or profound SHL, indicating significant damage to the auditory system, usually the cochlea. Sixteen patients had enhancement in the cochlea and vestibule. The highest proportion of positive studies was seen in patients with presumed immune labyrintitis (66%), followed by those with presumed viral labyrintitis (39%). There was no difference in the labyrinthine enhancement pattern among different presumptive diagnoses.

The frequency of schwanomas (acoustic neuromas) in our series was 4%.

Discussion

The role of early contrast-enhanced MR imaging of the temporal bone in patients with SHL has become more clear as experience has increased (13, 15–18). Numerous disorders of the temporal bone and cerebellopontine angle have been documented in this and other reports as the cause of SHL, including immune-mediated SHL (10, 12, 15, 19–24), viral diseases (2, 5–18, 25–28), Meniere disease (11), vascular insufficiency (1–4, 29–35), syphilis (36, 37), schwannoma (38), multiple sclerosis (39), and perilymphatic fistula (40).

Our experience with contrast-enhanced MR studies of the temporal bone in 78 consecutive patients with SHL resulted in an abnormal study in 31%, a higher prevalence than previously reported (40). Still, 22 (28%) of these 78 patients had no known etiologic agent, despite aggressive attempts to determine the cause of SHL.

The patients with immune-mediated SHL had the highest frequency (66%) of abnormal MR findings. Our patients included those with Sjögren disease and prelymphoma lymphocytic infiltrate of the meninges, relapsing polychondritis, systemic lupus erythematosus, neurosarcoidosis, and immune-mediated inner ear disease.

Patients with a presumed probable viral cause constituted the largest group, and their MR findings were

Final Diagnosis	No. of Cases	MR Findings		
		Abnormal	Normal	Characteristics
Immune labyrinthitis	9	6	3	Labyrinth enhancement
Viral labyrinthitis	19	7	12	Labyrinth enhancement
Meniere disease	13	1	12	Endolymphatic sac enhancement
Vascular	5	2	3	Basilar artery in cerebellopontine angle
Syphilis	3	1	2	Labyrinth enhancement
Schwannoma	3	3	0	Internal auditory canal enhancing mass
Multiple sclerosis	2	2	0	Periventricular plaques
Perilymphatic fistula	2	0	2	Normal
Idiopathic	22	2	20	Labyrinth enhancement
Total (%)	78 (100)	24 (31)	54 (69)	

abnormal 40% of the time. Only four of the 19 patients with a probable viral cause had serologic evidence, the rest had preexisting viral illnesses or a viral vaccination prior to the onset of SHL. In 13 patients, the initial episode of Meniere disease presented like SHL, but with time, the true diagnosis became apparent. When the degree of hearing loss prompted an electrocochleogram to be obtained, this test was often helpful in establishing the specific origin of the SHL. Vascular causes were established by contrast-enhanced MR studies in three patients, and schwannomas were identified most readily by this imaging technique.

We also established the presence of labyrinthine hemorrhage in several patients with sudden SNHL, a diagnosis that can now be made in vivo on unenhanced MR studies (17). However, since these patients were not seen in the acute stage, they were not included in this study.

The higher prevalence of positive studies in our series is clearly attributed to the use of intravenous contrast material. Indeed, without contrast administration, only 10 patients would have had positive studies, assuming the three acoustic neuromas would have been confidently diagnosed on high-resolution T2-weighted images.

Cost-benefit ratios are now more than ever being applied to expensive tests such as contrast-enhanced MR studies. Several authors have proposed limited (less expensive) studies of the internal auditory canal with noncontrast high-resolution T2-weighted imaging to exclude acoustic neuromas (41). However, it is clear from our study that such an approach would yield a very low percentage of positive studies in patients with acute SNHL (even though such a study may be appropriate in other contexts).

The choice of imaging study will depend on each clinician's approach to patients with SHL. Although the cost of the contrast-enhanced MR study is known, what is not known is the cost of not doing the study; that is, the liability of missed diagnoses, inappropriate or unnecessary treatment, and so on. If one takes a nihilistic approach to the treatment of SHL, as some have done (2, 42, 43), then no diagnostic tests need be performed. And if one's therapeutic approach to all cases of SHL is only corticosteroid treatment (5), then the contrast MR study may also be superfluous. If, however, one wants to fashion a treatment plan specifically designed for a known probable cause, then an early contrast-enhanced MR study has the best chance of yielding a positive result and of providing the clinician with an anatomic basis for such a therapeutic approach. Our findings suggest that the clinical impact of contrast-enhanced MR studies on the clinical management of patients with SHL deserves further evaluation.

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