Sudden Hearing Loss in Multiple Sclerosis: Case Report
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Abstract
This case illustrates the occurrence of a sudden hearing sensitivity loss that, in all likelihood, was the result of brainstem disorder resulting from multiple sclerosis (MS). Subject LD is a young woman who developed a sudden hearing loss while hospitalized for exacerbation of symptoms related to multiple sclerosis. By her own report, she had normal hearing in her left ear at the time of hospitalization. Four days after admission, she developed a hearing loss in her left ear, accompanied by roaring tinnitus and fullness. An audiologic evaluation revealed a substantial high-frequency sensitivity loss in the left ear. The combination of absent acoustic reflexes, depressed speech understanding, abnormal Bekesy audiometry, and an abnormal auditory brainstem response (ABR) was consistent with brainstem site of disorder. Over the next 2 weeks, hearing sensitivity recovered to within normal limits. This change in hearing sensitivity coincided with the recovery of acoustic reflexes, improvement in speech understanding, and partial recovery of the ABR.

Key Words: Auditory brainstem response (ABR), multiple sclerosis (MS), sudden hearing loss

Multiple sclerosis (MS) is a neurologic disease characterized by multiple focal demyelinating plaques. These plaques can occur throughout the brain, but have a predilection for the periventricular white matter of the brain stem. Because of the nature of the disease, all sensory and motor systems, including the auditory system, can be affected to a greater or lesser extent. Because the number and loci of the lesions within the brain stem vary considerably among patients, sequelae vary considerably as well.


Less well understood, however, are the effects of multiple sclerosis on hearing sensitivity (e.g., Stach et al, 1990). There appears to be general agreement that chronic hearing sensitivity loss may occur in patients with multiple sclerosis (Von Leden and Horton, 1948; Noffsinger et al, 1972; Cohen and Rudge, 1984; Musiek et al, 1989). The audiometric pattern appears to be as variable as the lesion site, and no single configuration emerges as being characteristic of multiple sclerosis. Indeed, the audiometric configuration of chronic hearing sensitivity loss has been variously described as unilateral and bi-
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There have also been reports of sudden hearing loss associated with multiple sclerosis (Von Leden and Horton, 1948; Phillips, 1952; Hallberg, 1956; Citron et al, 1963; Daugherty et al, 1983; Fischer et al, 1985; Shea and Brackmann, 1987; Barrett et al, 1988; Franklin et al, 1989). In most cases, it is unclear as to whether the transient hearing loss occurs as a result of sclerotic lesions of the brain stem or as a result of some idiopathic sudden hearing loss of cochlear origin. Since the presence of multiple sclerosis in a patient does not preclude a sudden hearing loss of some other etiology, when a hearing loss does occur in such a patient, its cause is often obscured.

Recent studies of acute hearing loss in multiple sclerosis have used measures of central auditory nervous system function, such as the auditory brainstem response (ABR), in an effort to determine the site of the lesion responsible for the hearing loss. However, since patients with MS can have abnormal ABR in the presence of normal peripheral sensitivity, an abnormal ABR in the presence of a sensorineural hearing loss may be unrelated. For example, Franklin et al (1989) reported two cases of acute hearing loss in patients with multiple sclerosis. Although the ABR was abnormal in both, there was no evidence to suggest that such an abnormality did not predate the hearing loss. In other studies, hearing sensitivity returned to normal levels, while the ABR remained abnormal (Fischer et al, 1985; Shea and Brackmann, 1987). In such cases, the notion that the ABR was abnormal before the sensitivity loss occurred must be entertained. Perhaps the most convincing evidence of acute hearing impairment emerges from patients who have experienced coincidental fluctuation of hearing sensitivity and retrocochlear signs (Fischer et al, 1985).

The present paper provides an illustrative example of a hearing sensitivity loss that, in all likelihood, occurred as a result of multiple sclerosis. The patient, a 22-year-old woman with multiple sclerosis, developed a sudden hearing loss while hospitalized for exacerbation of the disease. Results of audiologic evaluations, both at the time of her hearing loss and after her hearing returned to normal, suggested that the hearing sensitivity loss was retrocochlear in nature. This case supports the notion that sudden hearing loss can result from brainstem disorder associated with multiple sclerosis.

CASE REPORT

Description of Subject and Clinical Course

Subject LD, a 22-year-old female, was diagnosed with multiple sclerosis 6 months prior to the time of her sudden hearing loss. The initial diagnosis was made on the basis of abnormal MRI scans and abnormal visual evoked potentials. At that time, her ABR was noted to be normal.

On June 2, 1990, following 2 weeks of vertigo and left-sided numbness, LD was admitted to the hospital with severe vertigo, nausea, vomiting, and diplopia. She was placed immediately on intravenous therapy for dehydration. For the next 4 days, she underwent a series of tests to complete a diagnostic battery for multiple sclerosis.

A thorough neurophysiologic assessment was carried out on June 4 as part of the test battery. Results from the auditory evoked potential assessment showed a normal ABR in the right ear, with an absolute wave V latency of 5.44 msec and I–V interwave interval of 3.73 msec. The ABR in the left ear was abnormal, with an absolute wave V latency of 6.21 msec and I–V interval of 4.64 msec. The major portion of the delay was in the III–V interval. Although an audiologic evaluation was not carried out at that time, LD was noted to have normal behavioral thresholds to click stimuli.

During the 4 days of testing, the diplopia, vertigo, and nausea began to subside. On the morning of June 6, however, LD developed a sudden hearing loss in the left ear, accompanied by simultaneous numbness of her jaw and ear, roaring tinnitus, and a sense of fullness. She also stated that her diplopia had worsened.

Data and Observations

Initial Audiologic Evaluation

An audiologic evaluation was carried out on June 7 at the Methodist Hospital Audiology Service in Houston, Texas. The evaluation consisted of pure-tone, immittance, and speech audiometry. Immittance audiometry included measures of tympanometry, static immittance, and acoustic reflex thresholds. Speech audiometry consisted of single-syllable, phonetically balanced (PB) words, presented in quiet, and the Synthetic Sentence Identification (SSI) test (Jerger et al, 1968), presented with ipsilateral
competition at a message-to-competition ratio of 0 dB.

Immittance audiometry was consistent with normal middle ear function bilaterally, characterized by normal tympanograms, normal static immittance, and normal right uncrossed and right crossed acoustic reflexes. However, acoustic reflexes with sound to the left ear (left uncrossed and left crossed) were absent at all frequencies.

Results of pure-tone and speech audiometry are shown in Figure 1. Right ear results were consistent with normal hearing sensitivity through 4000 Hz and a mild sensitivity loss at 6000 and 8000 Hz. Speech understanding was normal. Left ear results showed a dramatic, steeply sloping, high-frequency sensorineural hearing loss. In addition, speech understanding was significantly depressed for both PB words and the SSI test. When scores on both measures were compared to expected performance for cochlear hearing loss of similar degree (Yellin et al., 1989), they fell below the lower limits of normal. That is, the expected scores for a cochlear hearing loss with a pure-tone average of this degree are better than those attained by this subject.

Because we were unsure about whether or not we would be able to record an ABR in an ear with such a substantial high-frequency hearing loss, we reverted to a traditional site-of-lesion test and carried out diagnostic Bekesy audiometry. Results were consistent with retrocochlear disorder. LD's thresholds by Bekesy audiometry to a pulsed tone were similar to conventional behavioral thresholds. However, significant auditory adaptation was present in response to a continuous tone, resulting in a Type III Bekesy audiogram.

Results of ABR testing are shown in Figure 2. ABRs were recorded in response to clicks of alternating polarity presented at 90 dB nHL at a rate of 11.1/sec. The right ear ABR was normal. Wave V latency was 5.4 msec, and the I-V interwave interval was 3.9 msec. The left ear ABR was strikingly abnormal, characterized by the presence of waves I and II only. In addition, wave I was noted to be of excessive amplitude in comparison to the left ear. Because of the paradox of being able to record a wave I in the presence of such a substantial hearing sensitivity loss, the wave I was tracked to its threshold. Results are shown in Figure 3. Wave I was followed down to 50 dB nHL. Because a wave I
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Figure 3  Auditory brainstem response from the left ear of a 22-year-old female with a diagnosis of multiple sclerosis. Testing was completed on June 8, 1990, 2 days after she reported a sudden hearing loss in her left ear. Stimuli were alternating clicks presented at three intensities at a rate of 11.1/sec.

Pure Tone Audiometry

80 dB nHL
60 dB nHL
50 dB nHL

Figure 4  Air-conduction (AC) pure-tone audiometry, acoustic reflex thresholds, and speech audiometric results in a 22-year-old female with a diagnosis of multiple sclerosis. Testing was completed on June 29, 1990, 23 days after she reported a sudden hearing loss in her left ear. Phonetically balanced (PB) word testing was carried out in quiet, and the Synthetic Sentence Identification (SSI) test was carried out at a message-to-competition ratio (MCR) of 0 dB.

Audiologic Re-evaluation

LD was discharged from the hospital several days later. On June 11, she noticed that her hearing began to return and that the roaring tinnitus decreased. By June 16, the tinnitus and vertigo had subsided. Her only residual complaint at this time was that of an “echo” in her left ear.

On June 29, LD was re-evaluated. Audiologic results showed that hearing sensitivity, acoustic reflex thresholds, and speech understanding had all returned to normal in the left ear. Immittance audiometry was consistent with normal middle ear function bilaterally, characterized by normal tympanograms, normal static immittance, and normal crossed and uncrossed acoustic reflexes. Acoustic reflexes with sound to the left ear had returned to normal levels.

Results of pure-tone and speech audiometry are shown in Figure 4. Left ear results had improved dramatically. Both right and left ear pure-tone thresholds were consistent with normal hearing sensitivity through 6000 Hz. In addition, speech understanding in the left ear was now at normal maximum levels.

ABR results are shown in Figure 5. ABRs were recorded in response to clicks of alternating polarity presented at 80 dB nHL at a rate of 11.1/sec. The right ear ABR remained normal, with a wave V latency of 5.4 msec and a I-V interwave interval of 3.8 msec. The left ear ABR remained abnormal, although wave V reappeared at a latency of 7.8 msec. Results of the three ABR evaluations of the left ear are shown in Figure 6. The initial ABR test, carried out prior to the onset of the sudden hearing loss, was the most well formed. The second ABR test, carried out in the presence of the sudden loss, showed an absence of the later waves. The final measurement, carried out after hearing sensitivity had returned, showed a re-emergence of wave V.
Figure 5 Auditory brainstem responses, with component peaks I, III, and V, from the right and left ears of a 22-year-old female with a diagnosis of multiple sclerosis. Testing was completed on June 29, 1990, 23 days after she reported a sudden hearing loss in her left ear. Stimuli were alternating clicks presented at an intensity of 80 dB nHL and a rate of 11.1/sec.

Figure 6 Auditory brainstem response from the left ear of a 22-year-old female with a diagnosis of multiple sclerosis. Testing was completed on three dates: (1) June 4, 1990, 2 days before she reported a sudden hearing loss in her left ear; (2) June 8, 1990, 2 days after she reported the loss; and (3) June 29, 1990, 23 days after she reported the loss.

COMMENT

D's hearing sensitivity loss appeared to be due to retrocochlear disorder. Audiologic evidence included absent acoustic reflexes with sound presented to the left ear, depressed speech understanding, and abnormal auditory adaptation. Following resolution of the sensitivity loss, wave V returned, although its latency remained abnormal. Thus, the ABR fluctuated along with the hearing sensitivity loss.

Of course, the fact that a patient has multiple sclerosis that is at an exacerbated stage does not preclude that person from having a sudden idiopathic hearing loss of cochlear origin. If the loss were of cochlear origin, however, the pattern of audiologic results would most likely have been one of better speech understanding, a measurable acoustic reflex at 500 Hz, no auditory adaptation, and an absent ABR.

One possible mechanism for sudden hearing loss in a patient with multiple sclerosis is related to the course of the disease process. Probably as a result of altered immune regulation, lymphocytes escape from the bloodstream, penetrate brain tissues, and destroy myelin. During the destruction process, transient edema develops in the tissue around the demyelinating lesions. This can affect transmission through axons, which results in exacerbation of symptoms. Once the inflammatory action subsides, swelling is reduced, and remission begins.
(Rivera, 1990). Thus, if the demyelination process occurs in the auditory brain stem, it could result in an auditory disorder that has an acute onset during the inflammation process, followed by a reduction in symptoms as the swelling subsides. LD's hearing loss reflected the disease course that is typical of multiple sclerosis. She entered the hospital as a result of a relapse of the disease. During the height of the exacerbation of her symptoms, she developed a hearing sensitivity loss that appeared to be retrocochlear in nature. Following discharge, and as her other symptoms began to remit, her hearing returned to normal.

Although sudden hearing loss is seldom reported in patients with multiple sclerosis, this case demonstrates that the disease, probably as a result of demyelination at some precise point in the auditory nervous system, can cause a hearing sensitivity loss. That the hearing returned during remission further implicates multiple sclerosis as the cause.

REFERENCES


