Otoacoustic Emissions in Sudden Unilateral Hearing Loss Associated with Multiple Sclerosis

Michael J. Cevette*
Martin S. Robinette†
Jonathan Carter*
Joost L. Knops†

Abstract

Sudden unilateral hearing loss may result from etiologies affecting cochlea, eighth nerve, or more central auditory tracts. Two case studies are presented in which the measurement of otoacoustic emissions helped rule out outer hair cell cochlear pathology. In both cases, the final diagnosis was sudden unilateral hearing loss associated with multiple sclerosis (MS). For one case, the sudden hearing loss was the first clinically recognized presenting sign of MS.

Key Words: Multiple sclerosis (MS), otoacoustic emissions (OAEs), sudden unilateral hearing loss

Evoked otoacoustic emissions (EOAEs) are acoustic responses measured in the ear canal that originate from the cochlea. They were first described by Kemp (1978) and are believed to be produced by an "active" mechanical process of outer hair cell (OHC) electromotility (Brownell, 1990). Many investigators have demonstrated that EOAEs are present in essentially all ears with normal middle ear and cochlear function (Bonfils et al, 1988; Stevens, 1988; Robinette, 1992a), and generally absent in ears with cochlear hearing loss exceeding 30 to 35 dB HL (Kemp, 1978; Kemp et al, 1986; Probst et al, 1987; Bonfils and Uziel, 1989).

The presence of EOAEs reflects normal cochlear function; they are generally absent with even a mild cochlear hearing loss. It has been suggested that the presence of EOAEs in patients with moderate to severe hearing loss is diagnostic of a retrocochlear auditory disorder, when a nonorganic component has been ruled out. Indeed, several investigators have measured the presence of EOAEs in moderate to profound confirmed or suspected retrocochlear hearing loss (Bonfils and Uziel, 1988; Lutman et al, 1989; Ohlms et al, 1991; Prieve et al, 1991; Robinette and Facer, 1991; Robinette, 1992a, b; Robinette et al, 1992; Patuzzi, 1993; Cane et al, 1994).

There are, however, other patients suffering idiopathic sudden hearing loss (ISHL) who reportedly often have EOAEs despite sensorineural hearing loss of 35 dB HL or greater across the frequency range of 1000 through 4000 Hz. Sakashita et al (1991) conducted transient EOAE evaluations on a group of 46 patients (50 ears) with long-standing sensorineural hearing loss of unknown etiology and a group of 42 patients (43 ears) within 14 days of onset of ISHL. In all patients, the presence of middle ear and retrocochlear lesions was ruled out by performing other audiologic tests such as acoustic immittance measurements, auditory brainstem responses (ABRs), and roentgenologic examinations such as computed tomography scans. Seven of the 50 ears with long-standing hearing loss had thresholds for the frequencies of 0.5 through 4 kHz that exceeded 35 dB HL. None of these 7 ears had EOAEs. On the other hand, 39 of the 43 ears with ISHL had thresholds for the frequencies of 0.5 through 4 kHz that exceeded 35 dB HL. Of these 39 ears, 20 had
measurable EOAEs. Indeed, 18 of these patients with EOAEs had hearing losses greater than 40 dB HL and 3 had hearing losses greater than 80 dB HL for frequencies of 0.5 through 4 kHz. The authors suggest that for patients with ISHL and EOAEs the inner ear injury is not to the OHCs but to the stria vascularis. In a follow-up report, Truy et al (1993) studied 24 patients with ISHL and agreed that the auditory damage might be to the stria vascularis, but failed to measure EOAEs for the 9 patients for whom hearing thresholds at the best frequency exceeded 40 dB HL.

Historically, it has been difficult to separate sudden hearing loss involving the cochlea from sudden hearing loss involving the eighth nerve or more central auditory tracts. In that cochlear (OHC) function is reflected by otoacoustic emissions, the addition of EOAE measures to other auditory tests may assist in the assessment of site of auditory lesion for some patients having sensorineural hearing losses. We present the diagnostic value of EOAEs for two patients with multiple sclerosis (MS) who suffered sudden unilateral sensorineural hearing loss.

CASE REPORTS

Case 1

History

The patient is a 41-year-old female evaluated at Mayo Clinic, Scottsdale in July 1993 for a relapse of MS that began 3 weeks earlier with sudden hearing loss in the right ear and vertigo. A previous evaluation at another institution in March 1992 led to a diagnosis of MS based in large part on an MRI scan revealing several areas of increased T2 signal in the periventricular white matter of both cerebral hemispheres and areas of increased signal in the right cerebral peduncle and the left medulla. She received four injections of adrenocorticotropic hormone (ACTH) at another facility shortly after the onset of the most recent symptoms. At the time of her visit to our facility, the patient felt that she had regained a little hearing in the right ear; no previous audiogram was available. The vertigo was almost resolved at the time of the July 1993 evaluation.

Audiologic Test Results

The results of pure-tone audiometry indicated a moderately severe reverse-sloped sensorineural hearing loss in the right ear and a slight high-frequency hearing loss in the left ear (Fig. 1, A). Her word recognition scores (W-22 lists) were 12 percent for the right ear and 100 percent for the left ear at a sensation level of 40 dB above her speech reception threshold. Tympanometry indicated normal middle ear function bilaterally. Ipsilateral and contralateral acoustic reflexes were absent for pure-tone stimuli to the right ear; pure-tone stimulation of the left ear produced normal ipsilateral and contralateral acoustic reflex thresholds and no acoustic reflex decay (Silman and Gelfand, 1981).

Transient evoked otoacoustic emissions were measured using the Otoacoustic Analyzer Transient Evoked ILO88 module and software (Kemp et al, 1986, 1990) controlled by an IBM PC. Stimuli were presented in the default mode (differential nonlinear test paradigm). Data were collected in the “quick screen” mode consisting of 260 blocks (8 clicks per block) of 80 μsec rectangular pulses presented at 80/sec and at about 86 dB peSPL in the right ear and 81 dB peSPL in the left ear. The pulses were presented through a transducer in a probe fitted to the patient’s ear canal with an immittance probe tip. Emissions were collected and averaged in a 10-msec time window from 2.5 through 12.5 msec between stimulus clicks.

The canal noise floor was 27.1 and 39.4 dB SPL in the right and left ears, respectively.

Figure 1  Audiogram and TEOAEs for case 1, a 41-year-old female with sudden right ear hearing loss. A, pure-tone audiogram; B, OAE frequency spectrum (light shaded area = noise floor, dark shaded area = emission in dB SPL).
TEOAE amplitudes were 10.4 dB SPL in the right ear and 10.6 dB SPL in the left ear. The emission frequency spectrum extended from 800 through 5000 Hz in the right ear and at 1000 through 2800 Hz and again at 4000 to 4500 Hz in the left ear (see Fig. 1, B). The waveform reproducibility was 98 percent and 69 percent in the right and left ears, respectively. These test results were interpreted as showing normal to near normal cochlear function for the emission frequencies tested.

ABR latencies for the patient's right ear were interpreted as normal when MS was diagnosed in March 1992 (Fig. 2, top tracings). However, following the right ear sudden hearing loss in July 1993, ABR right ear measures were abnormal. As shown in Figure 2 (middle and bottom tracings), the right ear response was consistent with retrocochlear abnormality as exemplified by absent wave components beyond wave II, while the left ear ABR was within normal limits.

Interpretation

In summary, the test results are as follows: normal left ear, moderate-to-severe right ear sensorineural hearing loss, normal tympanograms bilaterally indicating normal middle ear function, and normal otoacoustic emissions in the right ear suggesting normal OHC cochlear function. Otoacoustic emissions in the left ear are present but not as robust as in the right ear. This was due in part to the combination of lower stimulus level and higher noise level during the left ear recording (see the noise in frequency spectrum portion of Fig. 1, B). Absent acoustic reflexes (both ipsilateral and contralateral) with stimulation to the right ear and absent ABR wave components beyond wave II on the right side suggested dysfunction of the right afferent auditory pathway. The MRI of March 1992 showed increased signal in the right cerebral peduncle and the left medulla, indicating prior brainstem involvement from MS. However, at that time, both clinically and by ABR, there was no evidence of involvement of the auditory system.

While sudden unilateral hearing loss is an uncommon manifestation of MS, the combined evidence for this case suggests that the unilateral sensorineural hearing loss is associated with exacerbation of MS involving myelinated portions of the auditory nervous system.

Case 2

History

The patient is a 33-year-old male evaluated at Mayo Clinic, Rochester in March 1994 with a complaint of a sudden hearing loss in his left ear that occurred 4 weeks previously. He reported no previous history of ear or hearing problems, and he had no complaint of vertigo or tinnitus. He indicated that there had been some improvement in his hearing over the last 2 days.

Audiologic Test Results

The results of pure-tone audiometry revealed a mild-to-moderate sensorineural hearing loss in the left ear. The audiometric configuration for the right ear showed a slight low-frequency loss.
at 500 Hz and a mild high-frequency hearing loss above 3000 Hz (Fig. 3, A). His word recognition score (W-22 lists) for the left ear was 80 percent, compared to 100 percent for the right ear at a sensation level of 40 dB above his speech reception threshold. Tympanometry indicated normal middle ear function bilaterally. Ipsilateral and contralateral acoustic reflexes were absent for pure-tone stimuli to the left ear. With pure-tone stimulation to the right ear, contralateral reflexes were elevated for 500 Hz and normal for 1000 and 2000 Hz; ipsilateral reflexes for the right ear could not be measured due to artifacts. Acoustic reflex decay was absent at 1000 Hz for the right ear.

Transient evoked otoacoustic emissions were measured using the Otoacoustic Analyzer Transient Evoked IL088 module and software controlled by an IBM PC. Stimuli were presented in the default mode. Data were collected in the "original" mode, which consisted of 260 blocks of 80 µsec rectangular pulses presented at 50/sec and at about 80 dB and 81 dB peSPL to the right and left ears, respectively. Emissions were collected and averaged for a 17.5-msec time window of 2.5 through 20 msec between clicks. The canal noise floor was 33.6 dB SPL for the right ear and 33.7 dB SPL for the left ear. TEOAE amplitude for the right ear was 3.6 dB above the noise floor, and the emission frequency spectrum extended from 1200 through 2300 Hz (see Fig. 3, B). The waveform reproducibility was 66 percent. Left ear TEOAE amplitude was 4.8 dB above the noise floor and the emission frequency spectrum extended from 600 through 1200 Hz; the waveform reproducibility was 76 percent. Distortion product OAEs were also measured using the IL088-XP. Primary tones F1 and F2 were presented at 70 dB SPL at an F1/F2 ratio of 1.224. The evoked distortion product measured at the F1-2F2 place was displayed graphically at the F2 place (see Fig. 3, C). The measurements were made in 1/4-octave steps from 700 through 6000 Hz. In the right ear, DPOAEs were elicited from about 1200 through 1800 Hz and again from 2500 through 4000 Hz. DPOAEs in the left ear were elicited from about 800 through 6000 Hz, with the possible exception of the quarter octave between 3000 and 4000 Hz, where the response was obscured by noise artifact. The combination of these otoacoustic measures is interpreted as showing normal to near normal cochlear function for the majority of the frequency range from 1000 through 4000 Hz in each ear, despite the marked difference in pure-tone thresholds between ears.

Auditory brainstem response measures were accomplished employing a rarefaction click stimulus presented at 85 dB nHL at a rate of 11.1 per sec. As shown in Figure 4, the interpeak latency between wave I and wave V for the right ear was 4.40 msec, which lies near the upper limit of normal (normal being <4.54 msec). The left ear response showed an abnormal wave I absolute latency of 2.16 msec (normal being <1.88 msec). No other repeatable waveform peaks could be identified for the left ear.

**Medical Test Results**

An MRI scan of the brain showed no lesions in the area of the internal auditory canals but did show several small to moderate size T2 signal abnormalities in the periventricular white matter of both cerebral hemispheres (some of which were periventricularly located) and, to a lesser extent, portions of the superior basal ganglia-thalamic regions and a portion of the right pons-middle cerebellar peduncle. Spinal fluid analysis,
Figure 4 ABR tracings for case 2. The top tracings are replicated right ear normal waveforms. The bottom tracings are the replicated abnormal left ear waveforms.

Interpretation

In summary, the test results are as follows: mild-to-moderate unilateral sensorineural hearing loss, normal tympanograms indicating normal middle ear function, normal DPOAEs suggesting normal to near normal OHC cochlear function, absent acoustic reflexes (both ipsilateral and contralateral) with stimulation to the left ear, and abnormal ABR tracings indicating a disorder of the left afferent auditory pathway. The MRI showed increased signal in the superior basal ganglia-thalamic regions and the right pons and middle cerebellar peduncle.

While sudden unilateral hearing loss is rare as the initial presentation of MS, the evidence suggests that it was the first clinically recognized presenting sign of MS for this patient.

Past medical history revealed that 10 years earlier, he had an episode of a tingling sensation involving both feet that lasted approximately 2 weeks and resolved spontaneously. It is speculated that this event may have been his first exacerbation of disease, although not recognized at the time. Family history is also significant in that he has a maternal aunt with MS.

DISCUSSION

The prevalence of hearing loss as one of the initial symptoms associated with MS is estimated at about 1 percent (Kahana et al, 1973). Indeed, reports of sudden hearing loss as the only initial symptom are rare (Durlovic et al, 1994). The pattern and onset of hearing loss associated with MS may be as varied as the clinical consequences seen in other affected sensory and motor systems (Daugherty et al, 1983; Fischer et al, 1985; Shea and Brackmann, 1987; Barratt et al, 1988; Franklin et al, 1989; Stach and Delgado-Vilches, 1993). Since the disorder is associated with multiple focal demyelinating plaques throughout the white matter of the central nervous system, the central auditory pathway is susceptible to dysfunction at various sites and of differing degrees. The variability of type of auditory dysfunction in MS occurs for all standard measures of auditory function assessing peripheral, neural, and brainstem tracts such as immittance and ABR testing, as well as pure-tone and speech audiometry (Noffsinger et al, 1972; Hess, 1979; Stach and Jerger, 1984; Musiek et al, 1989; Jacobson and Jacobson, 1990). In order for one or more of the plaques of MS to produce unilateral loss that would show audiometrically, as occurred in these cases, it has been hypothesized that the lesion must lie distal to the cochlear nucleus complex and central to the neurilemmal-neuroglial junction on the auditory nerve (Hallpike, 1967; Antonelli et al, 1986; Robinette and Facer, 1991).

Evoked otoacoustic emission testing is a fast, objective screening method to assess normal OHC function. It can add valuable diagnostic information in determining cochlear versus retrocochlear involvement for patients with hearing losses of uncertain etiology. For the two MS patients described here, normal EOAEs helped rule out cochlear pathology as a possible cause of the sudden moderate sensorineural hearing losses.
REFERENCES


