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Case Report

Auditory Evoked Potential Inconsistency in Sudden Unilateral Hearing Loss with Multiple Sclerosis

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Sudden sensorineural hearing loss is a well-recognized clinical symptom in multiple sclerosis (MS). Acute inflammatory demyelination in the cochlear nerve or more central auditory tracts may cause sudden retrocochlear hearing loss. A 28-year-old male patient who was confirmed as having MS presented with suffering from dizziness as well as ongoing right-side hearing loss. We performed audiological tests, such as pure tone audiometry (PTA), otoacoustic emission, auditory brainstem response (ABR), and auditory steady-state response (ASSR). His clinical and audiological abnormalities disappeared with steroid therapy. However, each test showed different time courses of improvement. Although the results of the PTA and ASSR tests improved in exactly 1 month after the first attack, the results of the ABR reached 3 months to return to normal. To the best of our knowledge, this is the first case report of the time difference of hearing improvement shown in PTA, ASSR, and ABR tests.

KEYWORDS: Multiple sclerosis, sudden sensorineural hearing loss, auditory brain response

INTRODUCTION

Multiple sclerosis (MS) is a degenerative autoimmune disease of the central nervous system. Multiple focal demyelinating plaques develop throughout the brain in MS [1]. Various symptoms, such as double vision, paresthesia, motor weakness, and hearing loss, can occur according to the loci of these plaques [1]. In the presence of demyelinated plaques, which can affect the cochlear nerve and possibly more central auditory tracts, these developments may lead to sudden retrocochlear sensorineural hearing loss [2]. Previous studies have demonstrated auditory problems related to MS through several audiometric measurements [3]. A decreased speech understanding, an increased pure tone audiometry (PTA) threshold, an abnormal acoustic reflex threshold, and an abnormal auditory brainstem response (ABR) are some of the common findings of MS with symptoms of hearing problems [4].

In many previously reported cases of MS with hearing loss, physicians have used PTA and ABR tests. However, the results of auditory steady-state response (ASSR) tests are not described fully in most previous reports.

This case report represents the auditory findings and disease course of patients of hearing loss with MS, with different improvement courses between the three tests: PTA, ASSR, and ABR.

CASE PRESENTATION

A 28-year-old male patient presented to our clinic with right-side hearing impairment and vertigo. His vertigo symptoms started 1 month prior, and right-side hearing impairment developed 3 days prior to admission. On initial diagnosis, his brain magnetic resonance imaging (MRI) scan was abnormal. Brain MRI showed non-enhancing hyperintense signal lesions in the right cerebellar peduncle, the right lateral midbrain, the left corpus callosal splenium, and the left temporal periventricular white matter in the T2/FLAIR image (Figure 1). We evaluated his auditory function 3 days after administering steroid pulse therapy. On physical examination, both ears were normal. The PTA showed slight hearing impairment in the right ear, especially as the hearing threshold was 30 dB at 2000 Hz. The otoacoustic emission was normal bilaterally. However, the ASSR examination showed severe hearing loss in the right ear, with a hearing level (HL) between 70 and 110 dB HL. In addition, the ABR examination showed that only wave I was present at 70 to 90 dB HL, whereas waves III and V were absent at 90 dB HL. All audiological examinations in the left ear were normal (Figure 2). After 1 month, the symptoms of his hearing problem were relieved, and a follow-up PTA demonstrated all frequencies to be within the normal

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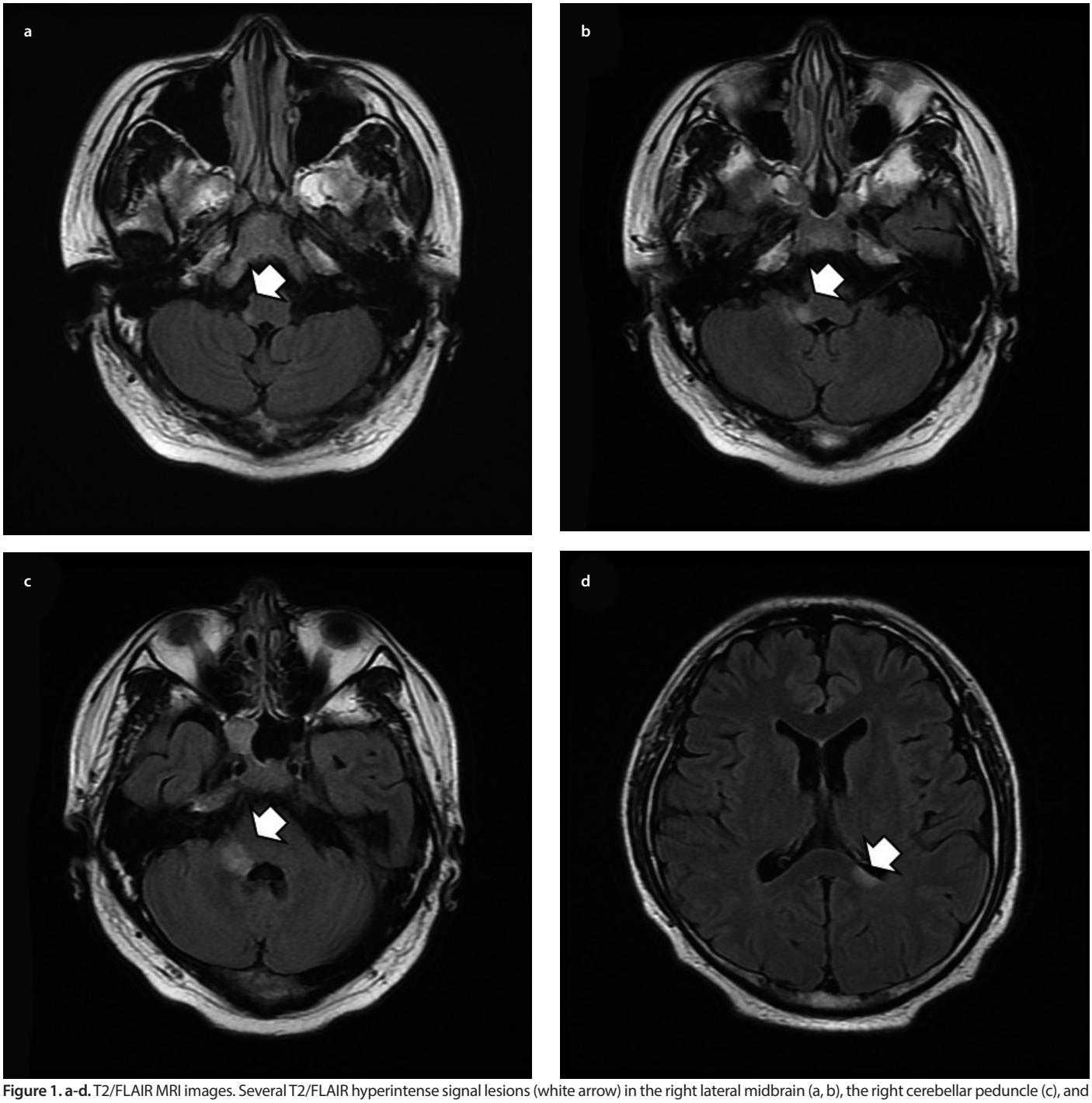


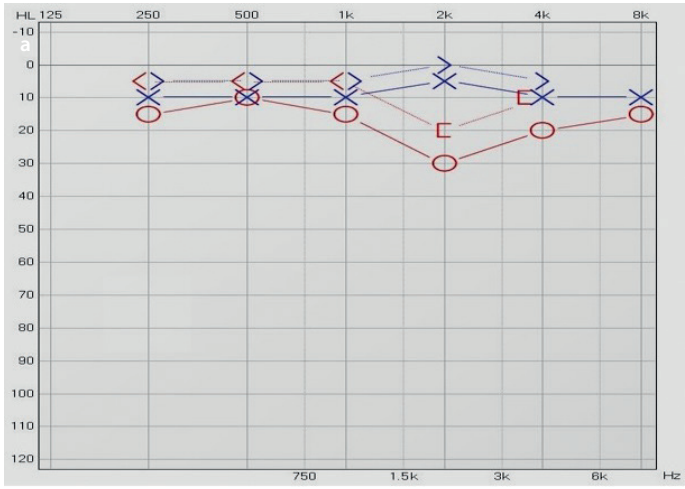
Figure 1. a-d. T2/FLAIR MRI images. Several T2/FLAIR hyperintense signal lesions (white arrow) in the right lateral midbrain (a, b), the right cerebellar peduncle (c), and the left temporal periventricular white matter (d).

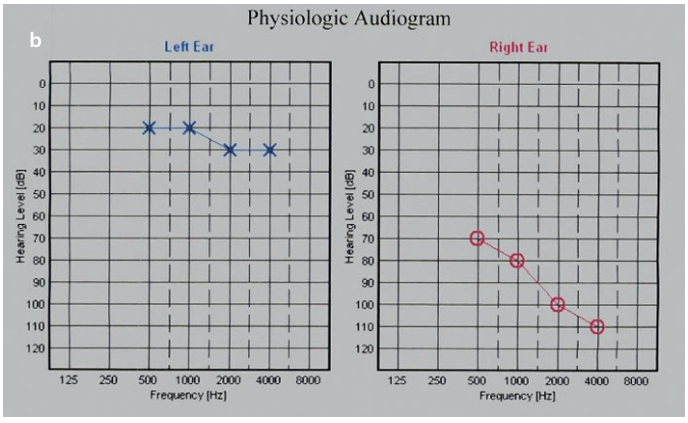
thresholds in the right ear. On ASSR examination, right-side hearing was improved, whereas the ABR result was unchanged (Figure 3).

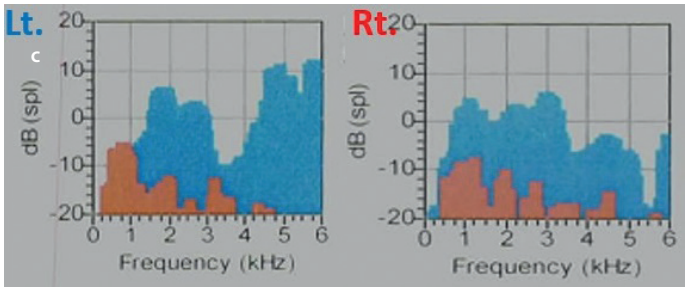
After 3 months, the patient's auditory symptoms were completely resolved. The following PTA test showed normal results as in the previous examination. For the ABR examination, the right-side wave V was recorded at an intensity of 30 dB HL, whereas a delayed wave III and V latencies were noted: the right-side wave III (4.29 ms) and wave V (6.58 ms) were delayed compared with the left-side wave III (3.62 ms) and wave V (5.50 ms) at an intensity of 90 dB HL (Figure 4). Verbal informed consent was obtained from the patient for the publication of this case report. This study complied with the Declaration of Helsinki human study and the recommendations of the Chonnam National University institutional review board.

DISCUSSION

The pathophysiology of MS remains unclear, but the underlying mechanism is thought to involve either destruction by the immune system or a failure of the myelin-producing cells. The demyelinated plaque usually affects the periventricular white substance of the central nervous system. In rare cases, the cerebellum, brainstem, and spinal cord can be affected by MS [5].







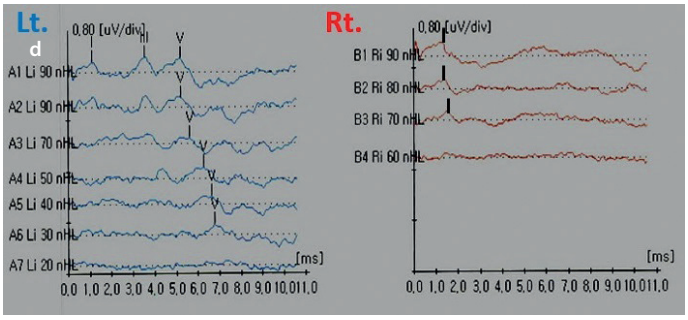
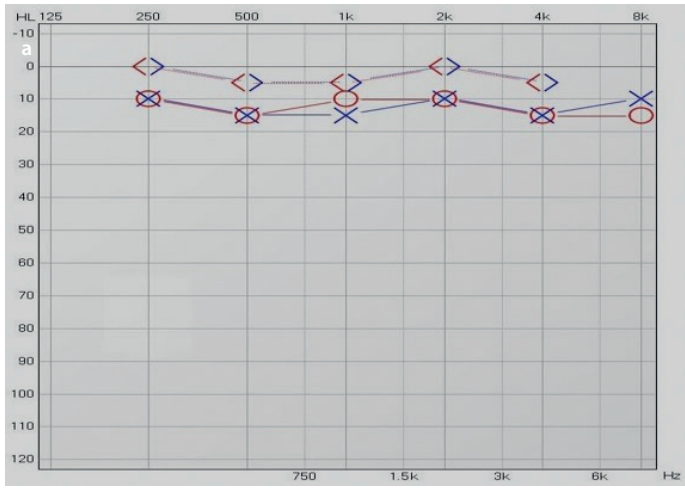
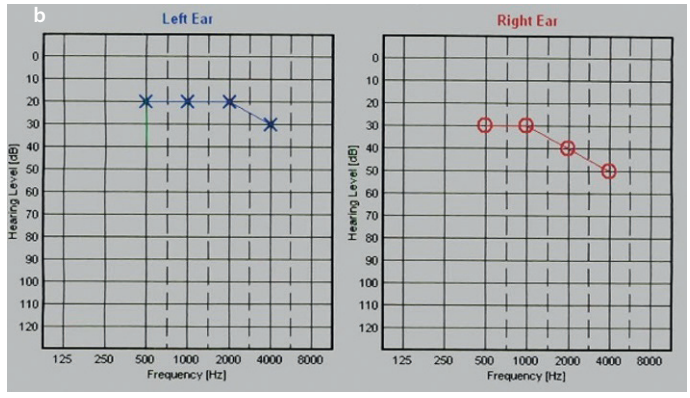


Figure 2. a-d. Audiological examination 1 week after the attack. Pure tone audiometry showed slightly high-frequency hearing loss in the right ear and normal hearing in the left ear (a). Auditory steady status response examination showed severe hearing loss in the right ear (b). Otoacoustic emission examination showed normal responses in both ears (c). Auditory brainstem response examination showed that only wave I was present at 70 to 90 dB HL, whereas waves III and V were absent at 90 dB HL in the right ear and a normal response in the left ear (d).
Lt: left-side ear; Rt: right-side ear.





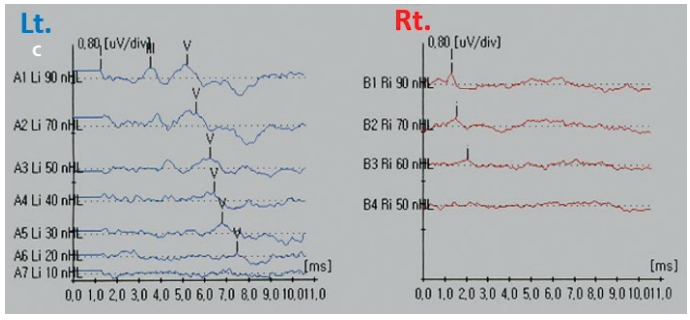


Figure 3. a-c. Audiological examination 1 month after the attack. Follow-up pure tone audiometry represented within normal threshold in all frequencies in the right ear (a). On auditory steady status response examination, right-side hearing was improved (b), whereas the auditory brainstem response result was unchanged (c).
Lt: left ear; Rt: right ear.

Hearing loss in patients with MS disease may be caused by plaques that are placed on the brainstem, any area of the entrance of the cochlear nerve into the brainstem, and the auditory cortex [6]. In our case, the hyperintense signal on the MRI at the right midbrain and the cerebellar peduncle is thought to be the main lesion that caused hearing loss. One consideration in this case is that hearing evaluation was performed 3 days after starting steroid therapy. Since acute brain lesion was found on MRI, we had to start steroid therapy immediately. However, this might have had some effect on hearing. According to ABR and ASSR, the threshold was still at severe hearing loss. Nonetheless, it would have been better to evaluate these prior to initiating any kind of treatment.

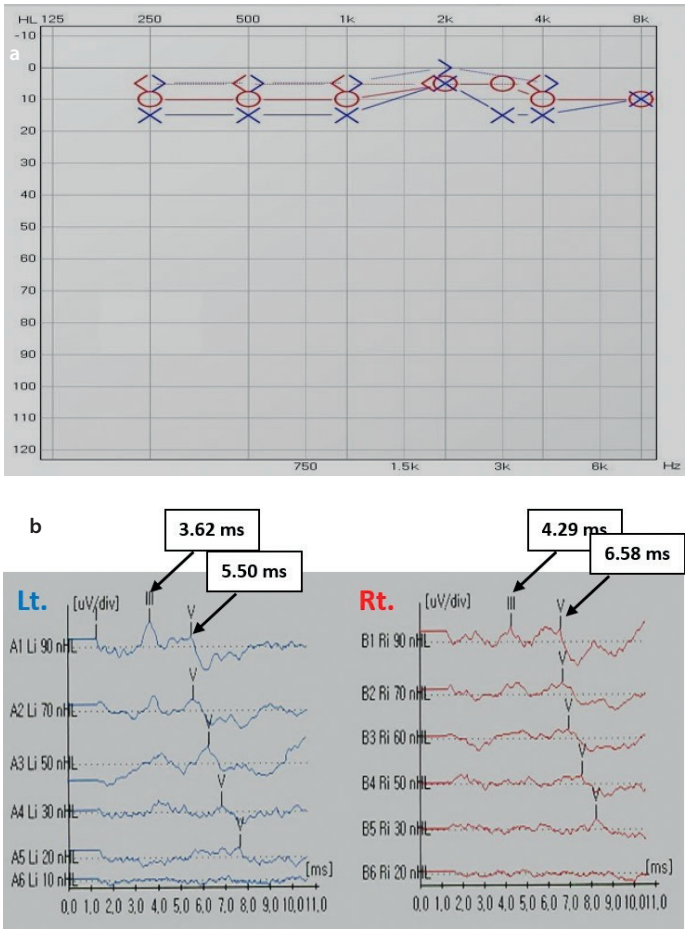


Figure 4. a-b. Audiological examination 3 months after the attack. Follow-up pure tone audiometry represented within normal threshold in all frequencies in the right ear (a). On auditory brainstem response examination, right-side wave V was recorded at an intensity of 30 dB HL, whereas delayed wave III and V latencies were noted (b).
Lt: left ear; Rt: right ear.

The ABR examination appears to be the most appropriate test for the diagnosis of sensorineural hearing loss in patients with MS. PTA or otoacoustic emission can frequently be normal in patients with MS [7, 8]. Moreover, researchers have reported that the use of evoked responses can provide a sensitive index of MS when compared with CT or even MRI [2]. Abnormalities in the auditory evoked potentials can be identified in patients with normal MRI, especially in cases that involve the brainstem. The relatively high cost of MRI, when compared with auditory evoked potentials, is an additional issue to consider in the clinical application of these two neurodiagnostic approaches in MS. ABR results frequently present abnormalities as absent waveforms except for wave I, delays in absolute latencies for waveforms, delays in interpeak latencies for I, III, and V, interaural latency of V and I–V, and poor waveform morphology and amplitudes of waves III and V [9]. An ABR can be normal for individuals with MS if the brainstem is not involved. A small plaque in the auditory brainstem pathway can alter the ABR results.

For the most part, previous reports of hearing loss related to MS have shown no significant relationship between the PTA and the ABR results. Franklin et al. reported two cases of acute hearing loss in patients with MS. Although the ABR was abnormal in both cases, there was no evidence on PTA to suggest that hearing problems existed [10]. In our case, the auditory findings of the hearing status and hearing improvement between the PTA and the ABR showed different results. This result also represents a typical relapsing–remitting pattern of MS, but the discrepancy between the PTA and the ABR was not fully understood.

ASSR is an auditory evoked response similar to ABR, but there has been no study on patients with MS using ASSR. Until now, ASSR has been known to have the advantage of obtaining a frequency-specific threshold as compared with ABR [11]. Meanwhile, the threshold correlation between the ABR and the ASSR of this patient has manifested as an unexpected result. The ABR and the ASSR showed a similar pattern immediately after the onset of MS symptoms, but the ASSR results showed a more rapid recovery of symptoms. Thus, if we used an ASSR in patients with MS who had difficulty hearing, we would be able to more rapidly determine whether the restoration of hearing had occurred or not.

CONCLUSION

This case report illustrates a typical relapsing–remitting course of MS and shows the diagnostic importance of the ABR and the ASSR as compared with the PTA. Further studies that analyze the combined ABR and ASSR findings of patients with MS would provide more information on retrocochlear pathophysiological hearing problems.

Informed Consent: Verbal informed consent was obtained from the patient.

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Conflict of Interest: The authors have no conflict of interest to declare.

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