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Sudden Sensoryneural Hearing Loss As a Rare Attack Type in Multiple Sclerosis

Multipl Sklerozda Nadir Bir Atak Türü Olarak Ani Sensorinöral İşitme Kaybı

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Multiple sclerosis (MS) is a chronic, multifocal, inflammatory, demyelinating disorder of the central nervous system. In addition to common symptoms like optic neuritis (ON), sensorial, pyramidal, and cerebellar dysfunction, a wide range of rare symptoms could be a feature of MS. Rare symptoms could be the presenting symptom of MS. Herein, we report a case with MS with a very rare symptom; unilateral vestibulocochlear nerve involvement and aimed to emphasize the importance of electrophysiological studies in the diagnosis.

A 35-year-old male patient who had been following with MS was admitted to hospital with the complaint of acute hearing loss. He had diagnosed with definite MS two years ago. The presenting symptom was left sided weakness, and hypoesthesia on the left upper extremity. At that time, the magnetic resonance imaging (MRI) of the brain revealed demyelinating lesions in periventricular white matter, corpus callosum and left capsula interna. Cervical MRI showed C3-C4 intramedullary demyelinating plaque. None of the lesions had contrast enhancement.

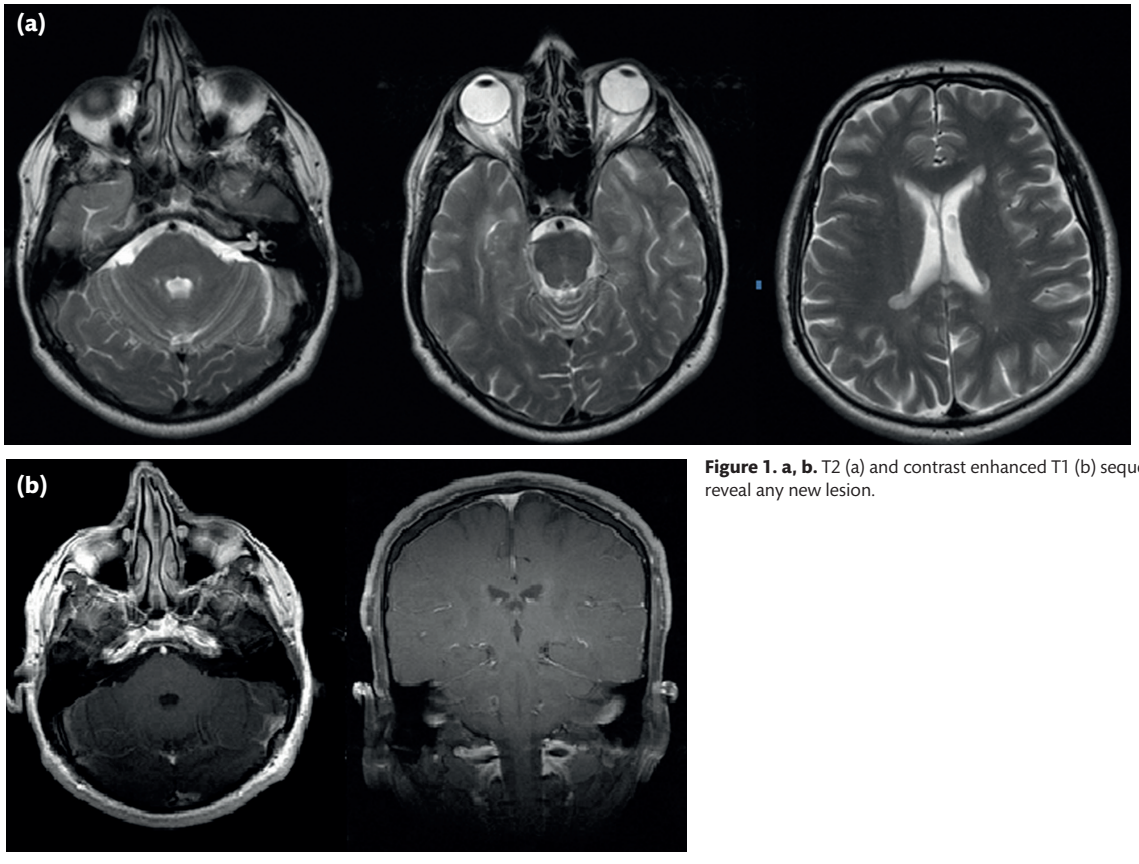


Figure 1. a, b. T2 (a) and contrast enhanced T1 (b) sequences of the brain MRI does not reveal any new lesion.

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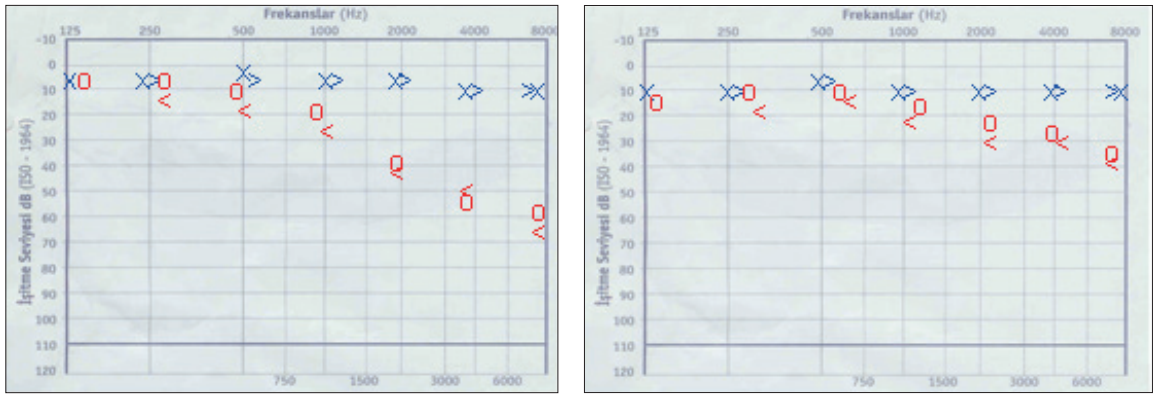


Figure 2. a, b. Pure tone audiogram performed at the symptom onset shows sensorineural hearing loss (a). Audiological assessment demonstrates significant improvement soon after the pulse steroid therapy (b).

Cerebrospinal fluid (CSF) protein and glucose levels, IgG index were within normal limits. Oligoclonal band was not detected, and vasculitis markers were negative. The symptoms totally disappeared with five dose pulse methyl prednisolone treatment (with a dose of 1 gr/day). He had been followed without any exacerbation for two years. He had not received any treatment with his decision when he was admitted with acute hearing loss on the right side and diplopia. The neurological examination showed right-beating nystagmus on the primary, rightward and vertical gazes. Weber test was localized to the left and Rinne test was positive, suggesting of a sensorineural loss on the right. He underwent to a brain MRI which showed chronic lesions and no new lesion was detected (Fig. 1). There was no contrast enhanced lesion or any lesion on the brainstem. Pure tone audiogram showed sensorineural hearing loss (Fig. 2a). Immediate brainstem auditory evoked potentials (BAEP) localized the lesion of acoustic nerve at the distal of the cochlear nucleus (Fig. 3a). The patient presented noticeable improvement in auditory symptoms after five dose pulse methyl prednisolone therapy (with a dose of 1 gr/day). Audiological assessment demonstrated significant improvement soon after the steroid therapy (Fig. 2b) while BAEP did not show any change. Eight months after the attack BAEP showed improvement (Fig. 3b). Because we decided to report our experience with a single case the informed consent was obtained from the patient.

A wide range of rare symptoms can be observed in the course of MS due to the extensive involvement of the central nervous system. The most frequent rare symptom group is cranial nerve (CN) abnormalities (except ON). 6th CN is the most common and the 3rd CN is the second one which is affected in MS. It was reported that all cranial nerves could be involved in MS (1).

The incidence of hearing loss in MS is variable in the literature (0.4–4.35%) (2, 3) Dizziness, vertigo and tinnitus might be accompanying symptoms. Cases with bilateral deafness were also reported (4). Since our case had the diagnosis of MS, clarifying the etiology of the hearing loss wasn't difficult. However, it could be a challenging condition if it was the presenting manifestation of MS or another neurological disorder (5, 6). Hearing loss was also declared in other demyelinating disorders such as neuromyelitis optica (7). For this reason, patients between the ages of 10 to 50 (most frequent ages for demyelinating disorders) should be referred to a Neurologist after the more common causes of hearing loss such as Meniere disease, ototoxic drugs, diabetes etc. are excluded.

Auditory tests are easily accessible tests which determine the kind and the amount of hearing loss. BAEP is a useful tool to detect the side of damage in the pathway. Brainstem lesions causing hearing loss are almost always detected in the brain MRI. However, as our patient, the lesions which effect the auditory pathway at level more distal than brainstem mostly could not detected by imaging (3). In that case, electrophysiological tests are more valuable for the localization of the damage. The knowledge from the literature suggests that the side of the lesion could be in any part of the auditory pathway. Bermagashi et al. described a patient with acute unilateral hearing loss and tinnitus. Wave I was absent in the affected side, which is suggestive of a damage at the acoustic nerve. The imaging demonstrated edema and hyperintense signal in the eight nerve within the auditory canal (8). Cases were reported whose Wave I was preserved while other waves were absent at the affected side (9, 10). The presence of Wave I shows normal cochlear function and the absence of other waves reveals that the lesion is very close to the root of the eight cranial nerve (9). The lesions in the brainstem which cause hearing loss are usually detected by the MRI (5, 11).

It is well known that BAEP abnormalities could seen in many patients with MS without a hearing problem and performing BAEP at the time of the diagnosis would have been helpful in our case. Auditory tests and BAEP are also useful to screen the improvement after the treatment.

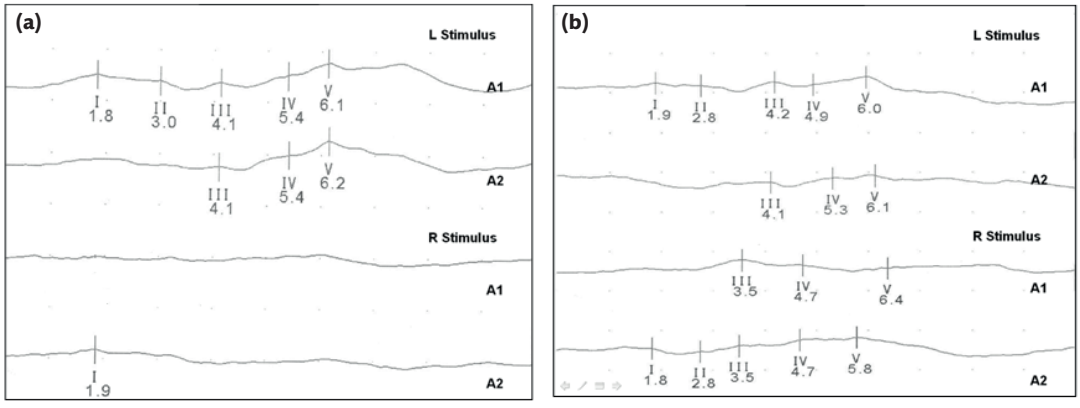


Figure 3. a, b. Immediate brainstem auditory evoked potentials (BAEP) localized the lesion shows only the formation of Wave I (a). Formation of the waves II, III, IV and V eight months after the incident strongly supports the recovery.

It was stated that hearing loss in patients with MS in the literature is generally short lasting and has a good prognosis (3). Significant clinical and audiological recovery was seen after pulse steroid treatment in our patient. BAEP did not show any changes soon after the treatment, but formation of the waves II, III, IV and V eight months after the incident which were not recorded in the initial study strongly supports the recovery. It was noted that BAEP abnormalities may persist more than a year despite hearing ameliorates (9).

In conclusion, hearing loss might be a feature of demyelinating disorders. This should be kept in mind and especially young patients with sensorineural hearing loss should be consulted to the Neurology department. As well as MRI, BAEP is also a very useful test for the localization of the lesion.

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