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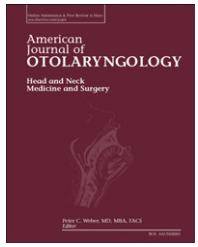

Isolated deafness in multiple sclerosis patients☆

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ABSTRACT

Isolated cranial nerve involvement in multiple sclerosis (MS) patients is not frequent. Deafness is considered to be uncommon in MS patients. We have reviewed the sensorineural hearing loss episodes that had been thoroughly investigated in our hospital in the last 5 years. We present three cases of sensorineural hearing loss in patients with MS and compare them with other previously reported and discuss this uncommon symptom. The cases that we present were firstly evaluated by an otolaryngologist. A lesion is seen at the root-entry zone of the eighth cranial nerve in only one case, but no lesions are seen in the other cases. A retrocochlear demyelinating disorder was demonstrated in the two patients in whom brainstem auditory evoked potentials were performed. All patients recovered at least partially their hearing functions.

1. Introduction

Isolated cranial nerve involvement is uncommon in multiple sclerosis (MS). It was present in 10.4% out of 483 patients in one study [1], more often as a presenting symptom (7.3%) than as a manifestation of a relapse (3.1%). Fifth cranial nerve is most often involved [1,2], followed by facial, abducens and oculomotor nerves.

Deafness is considered to be rare in MS. Unilateral deafness has been described in up to 4.35% of patients [1,3]. Sequential bilateral hearing loss has been reported too [4]. Other hearing disturbances, such as central hyperacusis with phonophobia [5], are less well characterized.

We have reviewed the sensorineural hearing loss episodes that had been thoroughly investigated in our hospital in the last 5 years. We have found only three cases of sensorineural hearing loss in patients with MS.

2. Case reports

2.1. *Patient 1*

A 29-year-old woman with 5-year history of relapsing–remitting MS reported a right side hearing disturbance, which had progressed in hours from tinnitus to complete hearing loss. The patient was firstly evaluated by an otolaryngologist 10 days after symptoms onset. An audiometry revealed right side sensorineural hearing loss. Examination additionally showed horizontal right-beating nystagmus. Oral treatment with corticosteroids was initiated. Three days later, brain MRI showed nonenhancing hyperintense T2 lesions. One of them was in the right eighth cranial nerve root-entry zone in the brainstem (Fig. 1). Brainstem auditory evoked potentials (BAEPs) found a right side retrocochlear demyelinating process (Table 1).

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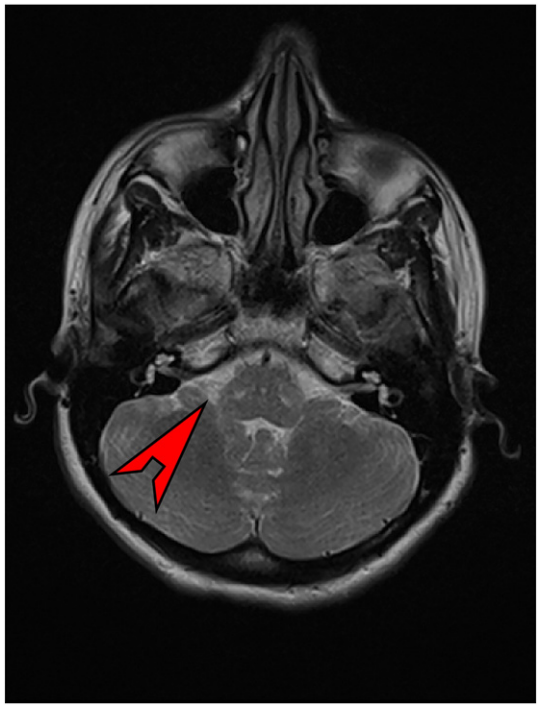

Fig. 1 – Cranial 1.5 T MRI of patient 1, showing a T2-hyperintense lesion in the right eighth cranial nerve root-entry zone in the brainstem (arrow).

The patient was referred to a neurologist that treated her with a 5-day course of intravenous methylprednisolone (1 g/day), with good recovery.

2.2. *Patient 2*

A 29 year-old woman presented with partial loss of vision in her right eye. She did not seek for medical attention until one week later, when she developed sudden left side hearing loss. Visual acuity in her right eye was 0.8. Visual evoked potential (VEP) was compatible with a demyelinating disturbance in both optic nerves, more severe in the right one. An audiometry demonstrated sensorineural hearing loss in the left side. BAEPs showed a left side retrocochlear demyelinating process (Table 1).

Brain MRI showed multiple T2-hyperintense lesions in the supratentorial area with periventricular and juxtacortical localizations. One lesion was showed in the right superior cerebellar peduncle, but no lesions were identified in the left eighth cranial nerve root-entry zone in the brainstem (Fig. 2); some of the supratentorial lesions were gadolinium-enhancing. CSF examination showed an elevated IgG index with six oligoclonal bands. All tests in serum and cerebrospinal fluid in order to rule out other potential etiologies were negative.

As the previous patient, she was firstly evaluated by an otolaryngologist, who initiated treatment with oral steroids and referred her to a neurologist. She received a 5-day course of intravenous methylprednisolone (1 g/day). She totally recovered her visual function but she still had a residual left side hearing loss of 25% with tinnitus some months later. In the following years she had clinical relapses and more lesions appeared in the MRI.

Table 1 – BAEPs showing a retrocochlear demyelinating disorder.

	dB	I ms	III ms	V ms	III–V ms
Patient 1					
L-BAEPs	80	1.74	3.68	5.72	3.98
R-BAEPs	80	1.94	4.58	6.68	4.74
Patient 2					
L-BAEPs	80	1.7	4.14	6.50	4.8
R-BAEPs	80	1.62	3.66	5.32	3.7

Right side in patient 1, left side in patient 2. It is verified by the increased latencies of waves I–V (compared with the expected latencies).

2.3. *Patient 3*

A 25 year-old woman complained about sudden left side hearing loss. When she was 22, a left cerebellopontine angle epidermoid cyst was partially removed. She was evaluated by an otolaryngologist. The audiometry was compatible with sensorineural hearing loss in the left side. The otolaryngologist decided to order an infratentorial area MRI to assess if the hearing disturbance was in relationship with the epidermoid cyst. The MRI showed no changes compared with the previous MRI; there were not lesions that could explain the acute hearing disorder (Fig. 3). The supratentorial region of the brain was not properly evaluated. BAEPs were not performed. The auditory deficit progressively and completely resolved without treatment. The hearing disturbance was considered to be idiopathic.

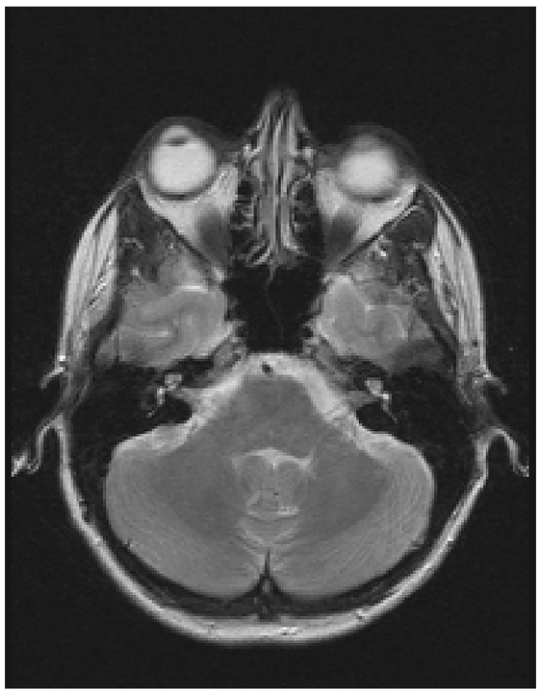

Fig. 2 – Cranial 1.5 T MRI of patient 2, showing no T2 lesions in the brainstem.