

Unusual primary manifestations of multiple sclerosis: A case report

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Abstract

Isolated cranial nerve involvement is rarely seen in multiple sclerosis patients. A 17-year-old female patient presented with complaints of numbness in the right half of her face, difficulty in chewing with the right jaw, having the right corner of her mouth drooping to the right, and blurring in both eyes. She had loss of sensation, left central facial paralysis, and frust monoparesis in the left arm in the right trigeminal nerve dermatome. Following systemic steroid treatment, her left central facial paralysis and chewing difficulties regressed, and loss of sensation improved. As in this case, MS may present with multiple instances of cranial nerve paralysis in addition to the involvement in the extremities. The association of the fifth and seventh nerve palsy in MS is rarely seen in the literature.

Keywords: Multiple sclerosis, Facial paralysis, Cranial nerve neuropathy

Introduction

Multiple sclerosis (MS) is a chronic, autoimmune, demyelinating disease of the central nervous system (CNS) [1]. Although brain stem involvement is common at the onset of MS and during the course of the disease, MS patients rarely have isolated cranial nerve involvement. In fact, of all MS patients, isolated cranial nerve palsy appeared as the first finding in only 1.6% to 5.2% [2]. Although some report that brain stem demyelination is one mechanism, isolated cranial nerve palsy is insufficient to explain MS pathogenesis [3]. Recently, the first clinical picture of recurrent MS has been described as “clinical isolated syndrome” (CIS) [4]. Therefore, isolated cranial nerve palsy with characteristic imaging patterns is now in this category. It is likely that, diplopia follows a partial sixth nerve lesion, while mild facial paralysis is observed in the partial seventh nerve lesion. As a result, the sixth nerve lesion is likely to be more prominent than the seventh nerve lesion. In the literature, there are case reports of MS patients with facial paralysis [5-7]. In the early stages of multiple sclerosis, facial hypoesthesia is observed as a result of the fifth nerve lesion, but it usually does not cripple the patient. However, magnetic resonance imaging (MRI) may miss some brainstem lesions. In patients with MS, the third and fourth nerve palsy (internuclear ophthalmoplegia and convergence disorder) show up without brainstem lesions. In one study, 19.6% of Japanese patients with MS had facial paralysis during the course of the disease [6]. However, in a Croatian cohort, the prevalence of facial paralysis was reported as 3.7% [8]. In a recent study conducted in Turkey, 5.3% of MS patients reported peripheral facial paralysis during the attack period [9].

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Informed Consent

The authors stated that the written consent was obtained from the patient and the parents of the patient presented with images in the study.

Conflict of Interest

No conflict of interest was declared by the authors.

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Case presentation

A 17-year-old previously healthy female was admitted to hospital with complaints of numbness in the right half of the face, difficulty chewing on the right side, dropping in the right mouth corner, and blurring in both eyes. She did not complain of any limb weakness, imbalance tendency to fall, but she did have weakness in the left arm. Her complaints started suddenly about two days prior to seeking treatment, and when the symptoms did not pass, she went to the hospital. The patient did not describe any complaints of diplopia, painful eye movements, hearing impairment, facial rash, dysphagia, or dysarthria. The patient had no history of traveling to a foreign country. Her neurological examination revealed normal orientation of place, person, and time. Although facial asymmetry was uncertain at first glance, when the patient was asked to show her teeth, there was retraction in the right mouth corner and indistinctness in the left nasolabial sulcus. She could lift her left eyebrow but not her right (Figure 1).

Figure 1: The first examination of the patient revealed that although facial asymmetry due to seventh nerve paralysis was uncertain at first glance, when the patient was asked to show her teeth, there was drooping in the right mouth corner, indistinctness in the left nasolabial sulcus, and she could not move her left eyebrow.



As a result of the current clinical findings, the patient was considered to have right trigeminal and left central facial paralysis. There was no saliva secretion or facial rashes. There was no weakness in the temporal and masseter muscles with palpation. Sensory examination revealed hypoesthesia on the right side of the face, characteristically matching the distribution of the trigeminal nerve in the skin, including the maxillary and mandibular parts of the trigeminal nerve. Other cranial nerve examinations were normal. Ophthalmoscopic examination revealed no optic atrophy or papillitis. Reflex examinations revealed no demonstrable pathological involvement affecting the extremities in the corticospinal, spinothalamic, and posterior column pathways. Cerebellar tests were normal. Brain and spinal MRI were performed (Figure 2, 3).

Figure 2: T2 FLAIR images show multiple hyperintense plaques in the periventricular deep white matter, the juxtacortical area, and in the plaque lesion located in the corpus callosum.

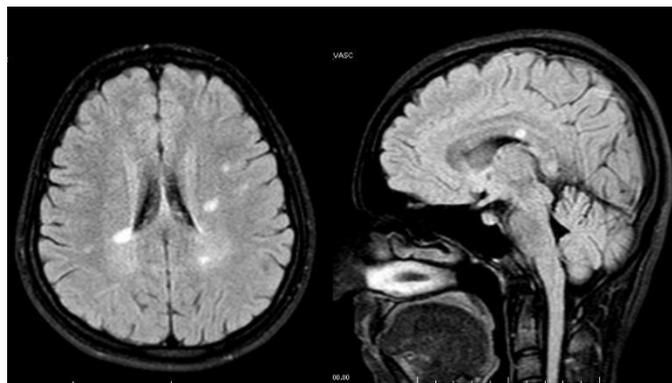
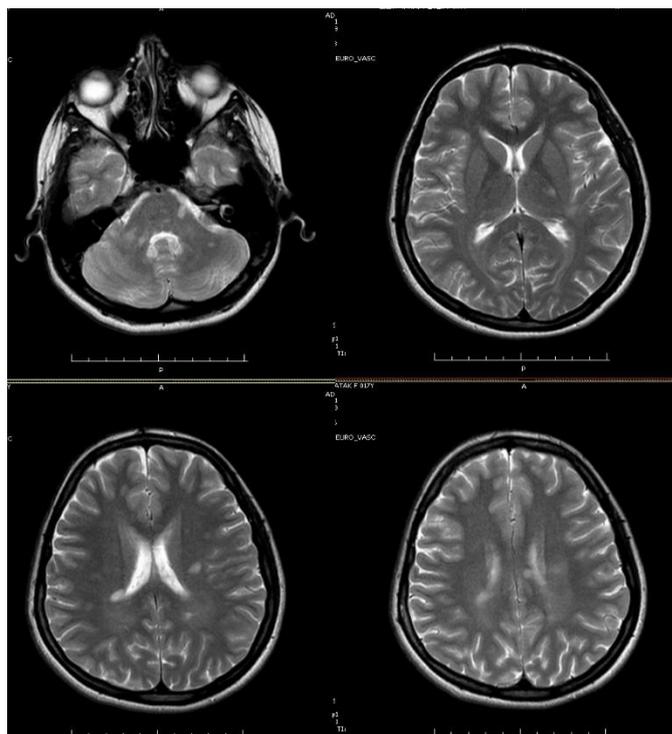


Figure 3: T2 A images are negative for plaque formation in the left pontobulbar junction, right inferior cerebellar peduncle level, left periaxial, callosal, and both periventricular deep white matter and juxta cortical placement.



There was no lesion in the spinal cord. Cerebrospinal fluid (CSF) analysis revealed normal protein levels without cells. Oligoclonal band (OCB) type 2 (three bands) was positive. Anti-myelin oligodendrocyte glycoprotein (MOG) and neuromyelitis optica (NMO) antibodies were negative. VEP, tibial, and median SEP of evoked potentials were within normal limits. All other basic biochemistry results were normal, including inflammatory markers. The patient was HLA B27 (-) and HLA B51 (+). The patient was treated with intravenous methylprednisolone (1000 mg / day) for 7 days. On the seventh day of hospitalization, the patient's left facial paralysis and chewing difficulties regressed and loss of sensation improved (Figure 4). The patient consented to participate in this study.

Figure 4: The left central facial paralysis regressed after treatment



Discussion

Isolated cranial nerve strokes can result from many diseases, including vasculitis, basal meningitis, and many other inflammatory conditions of the brain stem. The pathogenesis of isolated cranial nerve paralysis in multiple sclerosis is uncertain. Some reports suggested that brain stem demyelination plays a role [3], and the case presented herein had a brain stem lesion on MRI. Isolated cranial nerve paralysis is rarely seen in demyelinating diseases, including MS [10, 11].

While brain stem involvement is common in MS, isolated cranial nerve palsy is rare. In their retrospective study, Thömke et al. [12] reported that isolated cranial nerve palsy is a rare clinical finding in MS, affecting only 1.6% of patients in their series. Trigeminal neuralgia was reported as the first sign of MS in 0.3% and 1.9% of cases during the disease [3, 13, 14]. The case presented herein was admitted with motor and sensory symptoms due to trigeminal nerve involvement. The pathogenetic mechanism of trigeminal sensory neuralgia in MS patients is usually caused by demyelinating lesions affecting the pontine trigeminal pathways. Trigeminal sensory neuropathy secondary to MS preferably affects the second and third division of the trigeminal nerve, and in our case, there was hypoesthesia in the second and third branch of the trigeminal nerve. However, she did not have trigeminal neuralgia. In the literature, MS patients with facial paralysis have mostly been reported as case reports [5, 6]. In a case presented by Critchley, the first sign of MS disease was facial nerve paralysis [7]. Another study reported facial paralysis in 21 (19.6%) of 107 MS patients [6]. Zadro et al. [8] reported that 2.7% of patients presenting with seventh cranial nerve paralysis and 3.5% presented with trigeminal nerve paralysis as the first clinical symptoms. In the study of Thömke et al. [12] facial paralysis was the first finding of admission in three patients. Yetimalar et al. [15] reported peripheral facial paralysis in one of 21 MS patients who began having with unusual symptoms. The case presented herein was admitted with left central facial paralysis.

Conclusion

Since MS is characterized by multiple neurological symptoms, early diagnosis and treatment are critical to its

prognosis and course. Therefore, MS should be considered in the differential diagnosis of young adult patients presenting with isolated cranial nerve palsy.

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