

Unusual primary manifestations of multiple sclerosis: A case report

Yaşar Altun¹, Hacı Taner Bulut², Ali Arık¹

¹Department of Neurology, Medical Faculty of Adiyaman University, Adiyaman, Turkey

²Department of Radiology, Medical Faculty of Adiyaman University, Adiyaman, Turkey

ORCID ID of the author(s)

YA: 0000-0001-7013-0618
HTB: 0000-0002-8152-2497
AA: 0000-0002-0091-5724

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].

Corresponding Author

Yaşar Altun

Department of Neurology, Medical Faculty of Adiyaman University, Adiyaman, Turkey
E-mail: yasaraltun02@gmail.com

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.

Financial Disclosure

The authors declared that this study has received no financial support.

Published

2021 May 15

Copyright © 2021 The Author(s)

Published by JOSAM

This is an open access article distributed under the terms of the Creative Commons Attribution-Non Commercial-NoDerivatives License 4.0 (CC BY-NC-ND 4.0) where it is permissible to download, share, remix, transform, and buildup the work provided it is properly cited. The work cannot be used commercially without permission from the journal.



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.

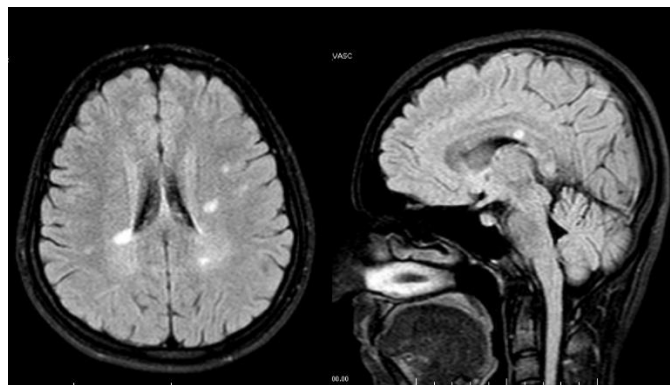
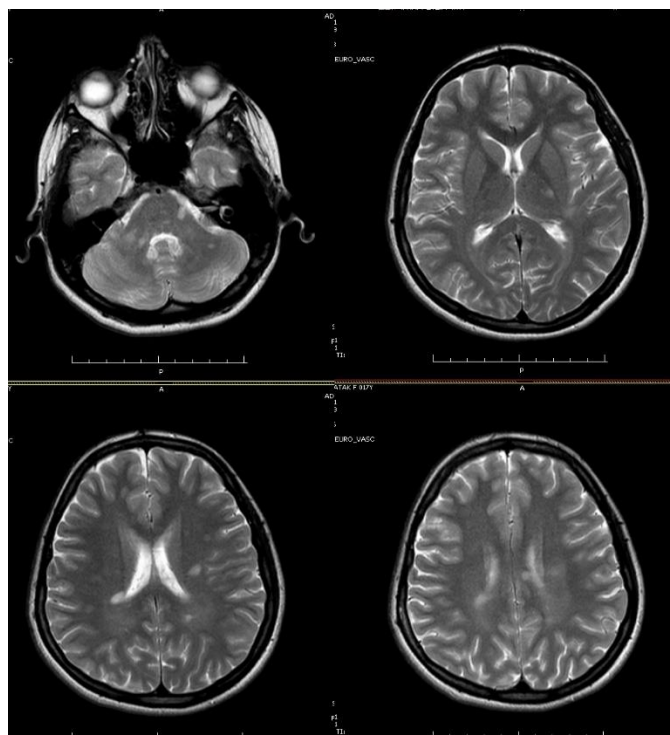


Figure 3: T2 A images are negative for plaque formation in the left pontobulbar junction, right inferior cerebellar peduncle level, left periaxial, callosal, and both periventricular deep white matter and juxta cortical placement.



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.

References

- Köseoğlu M, Çelik RGG, Tütüncü M, Bingöl A, Erbaş B, Deringöl D, et al. Sexual dysfunction and associated risk factors in multiple sclerosis. *J Surg Med.* 2020;4(9):779-83.
- Park KA, Oh SY, Min JH, Kim BJ, Kim Y. Cause of acquired onset of diplopia due to isolated third, fourth, and sixth cranial nerve palsies in patients aged 20 to 50 years in Korea: A high resolution magnetic resonance imaging study. *J Neurol Sci.* 2019 Dec 15;407:116546. doi: 10.1016/j.jns.2019.11.011. PMID: 31551555.
- Commins DJ, Chen JM. Multiple sclerosis: a consideration in acute cranial nerve palsies. *Am J Otol.* 1997 Sep;18(5):590-5. PMID: 9303155.
- Ratnayake EC, Caldera M, Perera P, Gamage R. Isolated trigeminal nerve palsy with motor involvement as a presenting manifestation of multiple sclerosis in an equatorial region-a case report. *Int Arch Med.* 2012 May 30;5(1):17. doi: 10.1186/1755-7682-5-17. PMID: 22647398; PMCID: PMC3403846.
- Kwon JY, Kim JY, Jeong JH, Park KD. Multiple sclerosis and peripheral multifocal demyelinating neuropathies occurring in a same patient. *J Clin Neurol.* 2008 Mar;4(1):51-7. doi: 10.3988/jcn.2008.4.1.51.
- Fukazawa T, Moriwaka F, Hamada K, Hamada T, Tashiro K. Facial palsy in multiple sclerosis. *J Neurol.* 1997 Oct;244(10):631-3. doi: 10.1007/s004150050158.
- Critchley EP. Multiple sclerosis initially presenting as facial palsy. *Aviat Space Environ Med.* 2004 Nov;75(11):1001-4. PMID: 15559004.
- Zadro I, Barun B, Habek M, Brinar VV. Isolated cranial nerve palsies in multiple sclerosis. *Clin Neurol Neurosurg.* 2008 Nov;110(9):886-8. doi: 10.1016/j.clineuro.2008.02.009.
- Sorgun MH, Kocer B, Kaplan F, Yılmaz N, Yücesan C. Cranial Neuropathy in Multiple Sclerosis. *Turk J Neurol.* 2011;17:137-42.
- Ebers GC, Sadovnick AD. The geographic distribution of multiple sclerosis: a review. *Neuroepidemiology.* 1993;12(1):1-5. doi: 10.1159/000110293.
- Kahana E, Leibowitz U, Alter M. Brainstem and cranial nerve involvement in multiple sclerosis. *Acta Neurol Scand.* 1973;49(3):269-79. doi: 10.1111/j.1600-0404.1973.tb01299.x.
- Thömke F, Lensch E, Ringel K, Hopf HC. Isolated cranial nerve palsies in multiple sclerosis. *J Neurol Neurosurg Psychiatry.* 1997; 63: 682-85.
- Hooze JP, Redekop WK. Trigeminal neuralgia in multiple sclerosis. *Neurology.* 1995 Jul;45(7):1294-6. doi: 10.1212/wnl.45.7.1294.
- Gass A, Kitchen N, MacManus DG, Moseley IF, Hennerici MG, Miller DH. Trigeminal neuralgia in patients with multiple sclerosis: lesion localization with magnetic resonance imaging. *Neurology.* 1997 Oct;49(4):1142-4. doi: 10.1212/wnl.49.4.1142.
- Yetimalar Y, Secil Y, Inceoglu AK, Eren S, Basoglu M. Unusual primary manifestations of multiple sclerosis. *N Z Med J.* 2008 Jul 4;121(1277):47-59. PMID: 18677330.

This paper has been checked for language accuracy by JOSAM editors.

The National Library of Medicine (NLM) citation style guide has been used in this paper.