A Case of Multiple Sclerosis Presenting as Eight and Half Syndrome

Sekizbuçuk Sendromu ile Karşımıza çıkan Multiple Skleroz Vakası

Özden KAMIŞLI[1], Mehmet TECELLİOĞLU[1], Abdülcemal ÖZCAN[2]

[1] İnönü Üniversitesi Tıp Fakültesi, Nöroloji Anabilim Dalı, MALATYA
[2] Malatya Devlet Hastanesi, Nöroloji Kliniği, MALATYA

ABSTRACT

We present a case young woman who presented with right gaze limitation and right eye limitation of adduction with horizontal nystagmus on abduction left side (one-and-a-half syndrome) in addition to a right-sided lower motor neuron facial nerve palsy which is called eight-and-a-half syndrome. The etiology in our patient was multiple sclerosis which was confirmed by medical history and magnetic resonance imaging.

Keywords: *Multiple sclerosis, eight-and-half syndrome, one-and-a-half syndrome*

ÖZET

Bu olguda sol tarafta abdüksiyon ile, sağda horizontal nistagmus (sekizbuçuk sendromu) ve sağ göz adduksiyon kısıtlılığı ile birlikte sağ bakış kısıtlılığına ek olarak sağ taraf alt motor nöron fasiyal sinir paralizisi ile karşımıza çıkan sekiz buçuk sendromlu bir kadın hastayı sunduk. Hastamızın etiyolojisi medikal öykü ve manyetik rezonans görüntüleme ile doğrulanmış olup; multiple skleroz tanısı mevcuttu.

Anahtar Kelimeler: *Multiple skleroz, sekizbuçuk sendromu, birbuçuk sendromu*

INTRODUCTION

One-and-a-half syndrome is an unusual disorder of eye movement, which is characterized by lateral gaze palsy in one direction and internuclear ophthalmoplegia (INO) in the other eye. The combination of one-and-a-half syndrome plus an ipsilateral facial nerve palsy (lower motor neuron type) was referred to eight-and-a-half syndrome by Eggenberger (1).

This syndrome is caused by a lesion in the dorsal tegmentum of the caudal pons involving the parapontine reticular formation (PRF) and the medial longitudinal fasciculus (MLF), as well as the nucleus and the fasciculus of the facial nerve. To our knowledge there are only a few case reports of "eight and half syndrome" due to multiple sclerosis (MS) (2,3).

KÜ Tıp Fak Derg 2015; 17(3): 34-38
Geliş Tarihi / Received: 04.09.2015
Kabul Tarihi / Accepted: 17.11.2015

Yazışma Adresi / Correspondence: Mehmet TECELLİOĞLU
Malatya Deevlet Hastanesi, Nöroloji Kliniği, MALATYA
E-posta: mehmettecelli@hotmail.com Tel: 0422 4445634

Here, we present a unique case of eight-and-a-half syndromes due to MS, which lesions lead to a combination of bilateral horizontal gaze palsy and unilateral peripheral facial palsy.

CASE REPORT

A 22-year-old woman presented with right gaze limitation and right eye limitation of adduction with horizontal nystagmus on abduction left side (one-and-a-half syndrome) in addition to a right-sided lower motor neuron facial nerve palsy. In the medical history she had been suffered from a vision loss one year ago. Magnetic resonance imaging showed hyperintense signal changes in the periventricular area, right cerebellar peduncle and paramedian pontine territory-ventrally to the fourth ventricle (Figure 1, 2, 4). The lesion's characteristic was found demyelinating, and it was clearly demonstrated on fluid-attenuated inversion recovery sequences (Figure 3). Lesions are multifocal within the brain, brainstem, and spinal cord. Patient's clinical and radiological features were consistent with MS.

This acute attack was treated with high dose steroid therapy (metilprednisone 1000 mg/day for 7 days). Within a month following therapy, an improvement in the gaze palsy was seen and ocular motility was partially restored.

Figure 1-4: Magnetic resonance imaging showed hyperintense signal changes in the periventricular area, right cerebellar peduncle and paramedian pontine territory-ventrally to the fourth ventricle

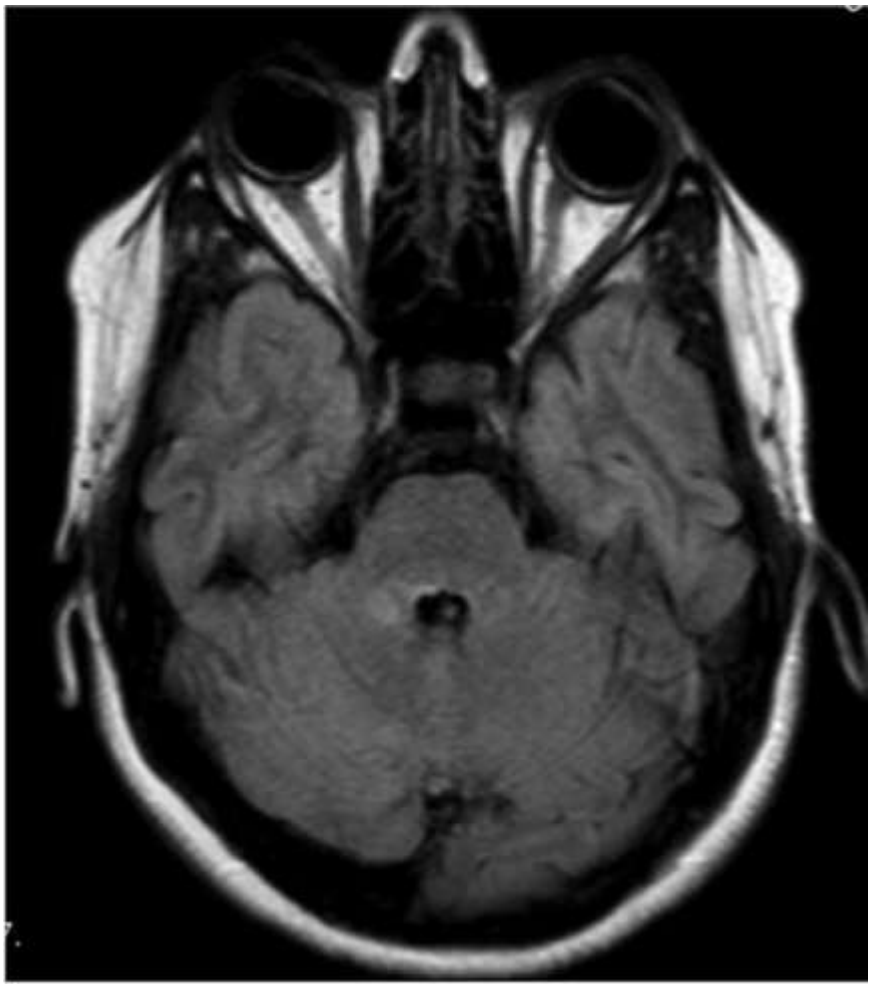

Figure 1

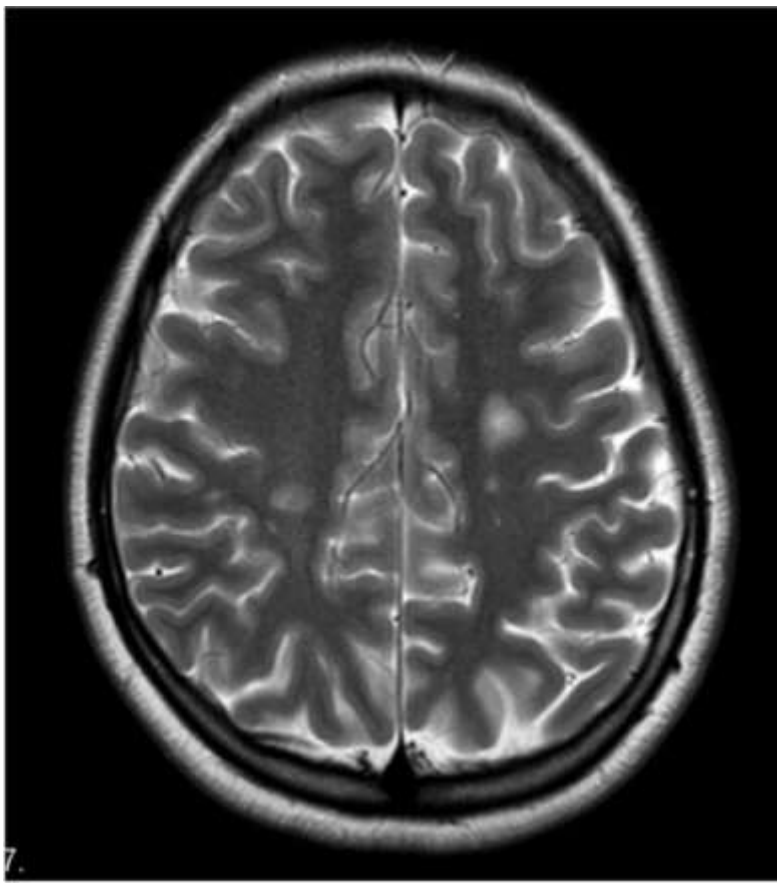

Figure 2

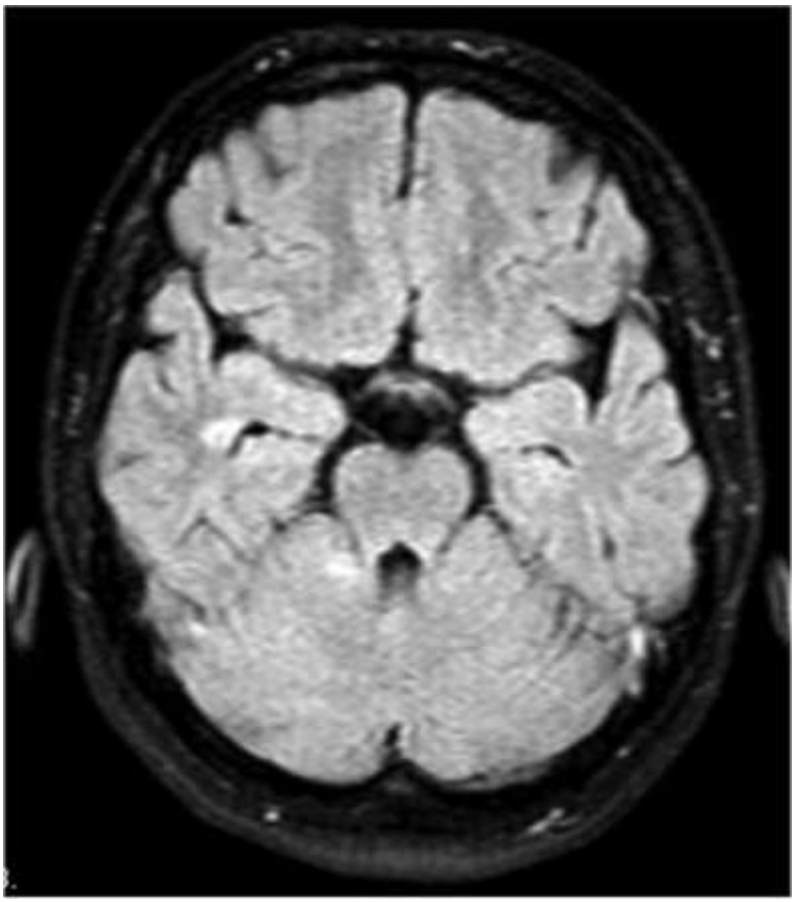

Figure 3

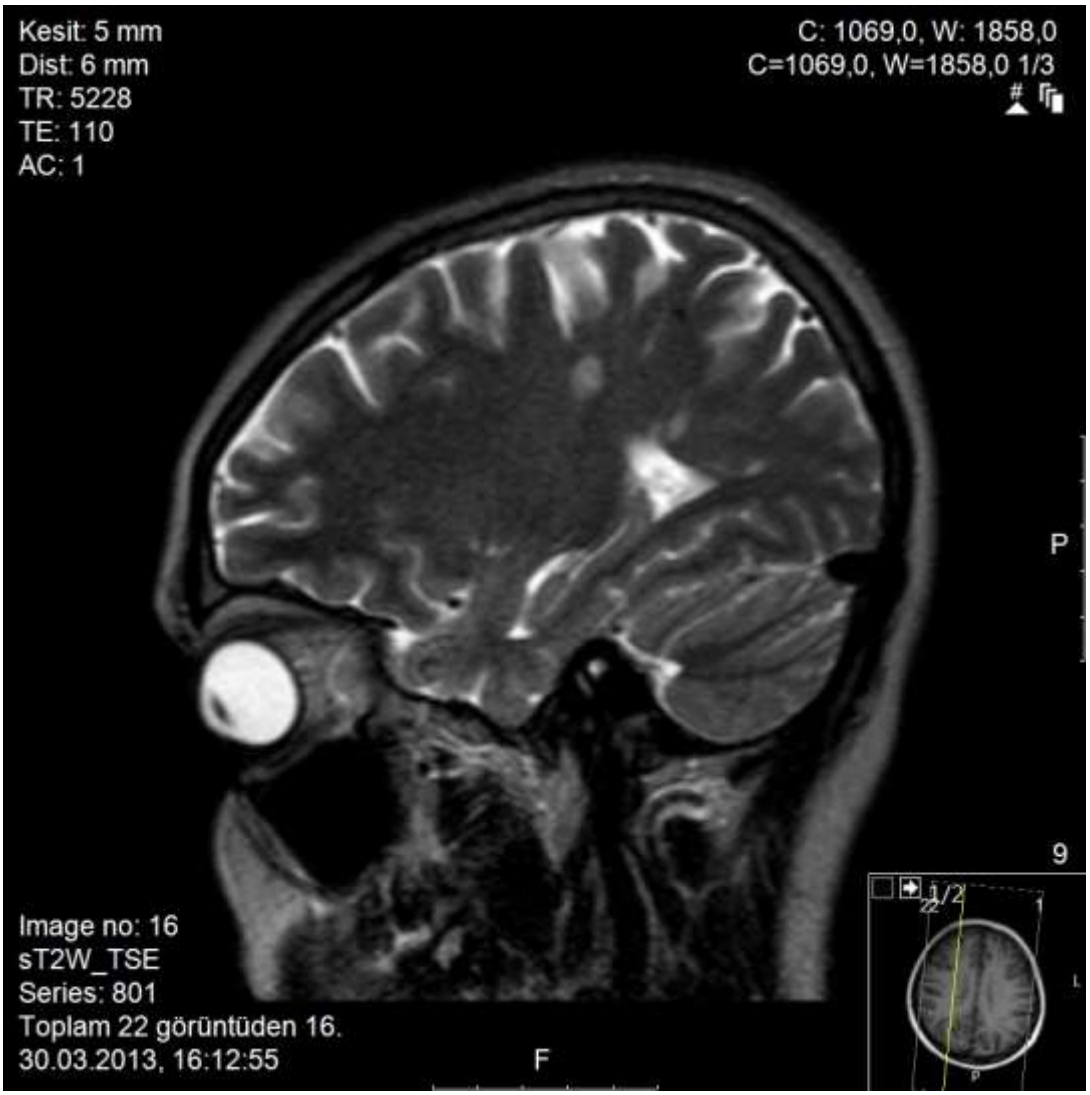

Figure 4

DISCUSSION

MS primarily affects the white matter of the brain and spinal cords, which is characterized by inflammation, demyelination, gliosis (scarring), and neuronal loss. Brainstem lesions, in particular intrapontine lesions, occur in significant frequency in patients with multiple sclerosis. Eye movement abnormalities are relatively common clinical manifestation of MS that occur from disruption of critical pathways in the brainstem, cerebellum, and cerebral hemispheres (4).

Horizontal eye movements are regulated in the pons by the abducens nucleus, MLF, and the paramedian pontine reticular formation (PPRF). INO is caused by a lesion in the MLF, which manifests with adduction deficit of the lesion side and abducting nystagmus of the contralateral side. A lesion affecting pontine tegmentum with an ipsilateral horizontal gaze palsy and INO is known as one-and-a-half syndrome (5). Eight-and-a-half syndrome is also a very rare syndrome and caused by a lesion in the pontine tegmentum involving the PPRF or abducens nucleus and the MLF, extended to the nucleus and fasciculus of the facial nerve (1). The eight-and-a-half syndrome is most often caused by brain stem infarcts, hemorrhages, trauma, basilar artery aneurysms, brainstem arteriovenous malformations, and tumors. It is rarely related with multiple sclerosis (2,3,6,9). Thus we reported this case for to point out the wide range of symptomatology in MS.

REFERENCES

1. Eggenberger E. Eight-and-a-half syndrome: one-and-a-half syndrome plus cranial nerve VII palsy. J Neuroophthalmol. 1998; 18(2): 114-6.
2. Mortzos P, Nordling MM, Sørensen TL. Eight-and-a-half syndrome as presenting sign of childhood multiple sclerosis. J AAPOS. 2014; 18(5): 490-2.
3. Raina R, Kaushik M, Mahajan SK, Raghav S, Sharathbabu NM. A case of multiple sclerosis presenting as eight and a half syndrome. Online J Health Allied Scs. 2012; 11(4): 1-3.
4. Prasad S, Galetta SL. Eye movement abnormalities in multiple sclerosis. Neurol Clin. 2010; 28(3): 641-55.
5. Anderson CA, Sandberg E, Filley CM, Harris SL, Tyler KL. One and one-half syndrome with supranuclear facial weakness: magnetic resonance imaging localization. Arch Neurol. 1999; 56(12): 1509-11.
6. Bocos-Portillo J, Ojeda JR, Gomez-Beldarrain M, Vazquez-Picon R, Garcia-Monco JC. Eight-and-a-Half Syndrome. JAMA Neurol. 2015; 72(7): 830.
7. Sampath Kumar NS, Raju CG, Kiran PR, Kumar TA, Gopal BV, Khaseem DB. Eight-and-a-half syndrome: a rare presentation of pontine infarction. J Stroke Cerebrovasc Dis. 2014; 23(8): e389-e391.
8. Nandhagopal R, Krishnamoorthy SG. Neurological picture. Eight-and-a-half syndrome. J Neurol Neurosurg Psychiatry. 2006; 77(4): 463.
9. van Toorn R, Schoeman JF, Donald PR. Brainstem tuberculoma presenting as eight-and-a-half syndrome. Eur J Paediatr Neurol. 2006; 10(1): 41-4.