



Case Report:

A Case of Multiple Sclerosis Presenting as Eight and Half Syndrome.

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Abstract: Multiple sclerosis (MS) is a chronic disease characterized by inflammation, demyelination, gliosis (scarring), and neuronal loss; the course can be relapsing-remitting or progressive. Manifestations of MS vary from a benign illness to a rapidly evolving and incapacitating disease requiring profound lifestyle adjustments. We report a 24 year old female who presented with right internuclear ophthalmoplegia with right lower motor neuron facial nerve palsy which is called eight and half syndrome. The etiology in our patient was multiple sclerosis which was confirmed by radio-imaging studies. Patient improved on pulse therapy of methyl prednisolone and tapering dose of steroids.

Key Words: Eight and half syndrome; Multiple sclerosis; Fifteen and half syndrome.

Introduction

Multiple sclerosis is a demyelinating disease affecting the myelin and can involve any part of central nervous system. We report an interesting case of eight and half syndrome affecting young female which was due to multiple sclerosis confirmed on magnetic resonance imaging. She improved on pulse therapy and tapering dose of steroids.

Case Report:

A 24 years female patient from Shimla presented in medicine OPD with complaints of acute onset of double vision which was non progressive. It was present only on left side eye movements. On examination, there was medial rectus palsy of right eye (Fig. 1) along with nystagmus in the abducting eye when the patient was asked to see at extreme left. There were also findings of right lower motor neuron facial palsy in the form of obliterated skin folds over forehead and prominent left nasolabial fold (Fig. 2a & 2b). Rest of the clinical neurological examination was normal. Patient was investigated with magnetic resonance imaging of brain and

spinal cord. MRI showed hyperintensities on T2 weighted images oriented perpendicular to the ventricular surface, corresponding to the pathologic pattern of perivenous demyelination (Dawson's fingers) and hyperintensities in the dorsal tegmentum of caudal pons (Fig. 3a & 3b). Patient was diagnosed to be multiple sclerosis. She was treated with intravenous methylprednisolone 1000mg for 5 days followed by oral prednisone tapered over 2 weeks. Patient signs improved during hospital stay. On follow up, patient had no complaints.



Fig. 1: Right medial rectus palsy

Discussion:

Multiple sclerosis (MS) is a chronic disease characterized by inflammation, demyelination, gliosis (scarring), and neuronal loss; the course can be relapsing-remitting or progressive. Lesions of MS typically occur at different times and in different CNS locations (i.e., disseminated in time and space). Manifestations of MS vary from a benign illness to a rapidly evolving and incapacitating disease requiring

profound lifestyle adjustments. MS is approximately threefold more common in women than men. The age of onset is typically between 20 and 40 years (slightly later in men than in women)¹, but the disease can present across the life span. Our patient was female with age of 24 years which was consistent with normal age and sex presentation.



Fig. 2a: Absence of skin folds over right half of forehead



Fig. 2b: Prominent left nasolabial fold



Fig. 3a: Dawson's fingers and hyperintensity in dorsal tegmentum of lower pons.



Fig. 3b: Hyperintensities in periventricular area.

Eight-and-a-half syndrome is caused by a lesion in the dorsal tegmentum of the caudal pons involving the PPRF or abducens nucleus and the medial longitudinal fasciculus(MLF), as well as the nucleus and fasciculus of the facial nerve.² It is characterized by Internuclear Ophthalmoplegia (INO) in addition to horizontal gaze palsy (one-and-a-half syndrome) and ipsilateral lower motor neuron-type facial palsy. Our patient had same signs on right side. Patients with one-and-a-half syndrome and facial nerve palsies may develop oculopalatal myoclonus weeks to years after the onset of the ocular motility problem.³ The one-and-a-half syndrome has also been described with facial diplegia (the fifteen and half syndrome [$1\frac{1}{2} + 7 + 7 = 15\frac{1}{2}$]).⁴ It may also be associated with supranuclear facial weakness on the same side as the gaze palsy and internuclear ophthalmoplegia with lesions of the paramedian aspect of the dorsal pontine tegmentum, providing evidence for the existence of corticofugal fibers that extend to the facial nucleus in the dorsal paramedian pontine tegmentum.⁵ The one-and-a-half syndrome is most often caused by multiple sclerosis, infarcts, hemorrhages, trauma, basilar artery aneurysms, brainstem arteriovenous malformations, and tumors.⁶⁻⁸

Symptoms of MS are extremely varied and depend on the location and severity of lesions within the CNS . Examination often reveals evidence of neurologic dysfunction, often in asymptomatic locations. On MRI, characteristic abnormalities are found in >95% of patients although more than 90% of the lesions visualized by MRI are asymptomatic.¹ Lesions are frequently oriented perpendicular to the ventricular surface, corresponding to the pathologic pattern of perivenous demyelination (Dawson's fingers). Lesions are multifocal within the brain, brainstem, and spinal cord. Lesions larger than 6 mm located in the corpus callosum, periventricular white matter, brainstem, cerebellum, or spinal cord are particularly helpful diagnostically.

Diagnostic criteria for clinically definite MS require documentation of two or more episodes of symptoms and two or more signs that reflect pathology in anatomically noncontiguous white matter tracts of the CNS. Symptoms must last for >24 h and occur as distinct episodes that are separated by a month or more. At least one of the two required signs must be present on neurologic examination. The second may be documented by abnormal paraclinical tests such as MRI or evoked potentials (EPs)¹. Our patient had signs of eight and half syndrome ald lasted for more than 24 hours. Second part was documented on MRI(1 attack; objective clinical evidence of 2 or more lesions). So our patient had definite multiple sclerosis.

References:

1. Hauser SL, Goodin DS. Multiple Sclerosis and other demyelinating diseases. Harrison's principles of internal medicine 18th edition New York: McGraw Hill; 2012. p. 3395-3409.
2. Eggenberg E. Eight-and-a-half syndrome. *J Neuroophthalmol* 1998;18:114-116.
3. Wolfe GI, Galetta SL, Mollman JE. Spontaneous remission of papilledema and sixth nerve palsy in acute lymphoblastic leukemia. *J Neuro-Ophthalmol* 1994;14:91-94.
4. Bae JS, Song HK. One-and-a-half syndrome with facial diplegia: the 15 1/2 syndrome? *J Clin Neuroophthalmol* 2005;25:52-53.
5. Anderson CA, Sanberg E, Filley CM, et al. One and one-half syndrome with supranuclear facial weakness. *Arch Neurol* 1999;56:1509-1511.
6. Kim JS. Internuclear ophthalmoplegia as an isolated or predominant symptom of brainstem infarction. *Neurology* 2004;62:1491-1496.
7. Ohta K, Gotoh F, Fukuuchi Y, et al. Midpontine tegmentum infarction with one-and-a-half syndrome□ demonstrated by magnetic resonance imaging. *Keio J Med* 1994;43:164-165.
8. Wall M, Wray SH. The one-and-a-half syndrome. A unilateral disorder of the pontine tegmentum: a study of 20 cases and review of the literature. *Neurology* 1983;33:971-980.