

A Case of Eight-and-a-Half-Syndrome as a Presenting Manifestation of Late Onset Multiple Sclerosis

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Abstract

We describe a 74-year-old woman presenting with right Internuclear Ophthalmoplegia (INO), horizontal gaze palsy and ipsilateral peripheral facial nerve palsy known as eight-and-a-half-syndrome. MRI and CSF findings led to diagnosis of Clinically Isolated Syndrome (CIS). One month later, the patient suffered acute myelitis.

Our patient represents a rare case of late onset Relapsing-Remitting Multiple Sclerosis (RRMS) presenting with an eight-and-a-half-syndrome. Apart from the clinical rarity of eight-and-a-half-syndrome, this case serves as a reminder that MS can occur in every period of life and should be considered in differential diagnosis irrespective of age at onset of neurological symptoms [1].

Keywords: Eight-and-a-half-syndrome; Multiple sclerosis; Case report; Late onset; Eye movements

Case Report

A 74-year-old woman was admitted to our neurology service five days after sudden onset of diplopia and unsteadiness of gait.

On neurological examination, the patient showed right Internuclear Ophthalmoplegia (INO) and right-sided horizontal gaze palsy consistent with a one-and-a-half-syndrome. Further, there was right-sided peripheral facial nerve palsy, the rest of her neurological examination was normal. Her past medical history included hypercholesterolemia and euthyroid struma.

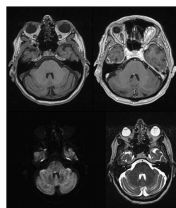


Figure 1: A hyperintense signal and there was a slight gadolinium contrast enhancement, consistent with an inflammatory origin.

Cranial MRI with Diffusion Weighted Imaging (DWI) revealed a slight diffusion restriction in the right paramedian pontine region ventrally to the fourth ventricle and a corresponding low signal on the Attenuated Diffusion Coefficient (ADC) map arguing against an ischemic aetiology. On coronal and transversal T2-weighted images the lesioned area showed a hyperintense signal and there was a slight gadolinium contrast enhancement, consistent with an inflammatory origin (Figure 1).

In addition, there were multiple periventricular, pericallosal and subcortical T2 hyperintense lesions arguing for chronic demyelinating disease (Figure 1).

Magnetic Resonance Angiography (MRA) and sonography of the supraaortal vessels showed slight calcification without evidence of any intra- or extracranial stenoses. Cerebrospinal fluid (CSF) investigation revealed one cell per microliter and the presence of numerous intrathecal Oligoclonal Bands (OCB) which were not found in serum.

Overall, clinical presentation, Magnetic Resonance Imaging (MRI) and CSF findings were consistent with Clinically Isolated Syndrome (CIS) [2]. Over the first few day of diagnostic work-up spontaneous improvement occurred and the patient was discharged after complete remission after 10 days.

One month later, the patient was re-admitted because of new onset weakness of her left upper and lower limb. Neurological examination showed a left-sided sensorimotor hemiparesis (BMRC 3-4/5) with left-sided hyper-reflexia and a positive Babinski sign. Cranial nerve findings were normal at this time. MRI of the spinal cord revealed a T2 hyperintense lesion at the C3/C4 level with slight contrast enhancement suggestive of acute myelitis (Supplemental Figure 2). After admission of intravenous corticosteroids (cumulative dose 3000 mg) over 3 days complete remission of symptoms occurred after 5 days.

At follow up six months later, the patient had not experienced another relapse and had a normal neurological examination. Considering the patient's age, complete remission of symptoms and low MRI lesion load, we refrained from initiation of disease modifying therapy.

Discussion

One-and-a-half-syndrome was first introduced by Fischer et al. in 1967 to describe a combination of horizontal gaze palsy and internuclear ophthalmoplegia [3]. It is defined as a horizontal gaze palsy due to unilateral damage to the Parapontine Reticular Formation

(PPRF), and an ipsilateral Internuclear Ophthalmoplegia (INO) causing a deficit in adduction of the ipsilateral bulb due to an interruption of internuclear fibres of the ipsilateral Medial Longitudinal Fasciculus (MLF) after crossing the midline.

One-and-a-half-syndrome in combination with peripheral facial nerve palsy was described as eight-and-a-half syndrome by Eggenberger et al. [4], a condition mostly caused by a vascular or demyelinating lesion [5,6].

Anatomical proximity of PPRF, MLF, abducens nucleus and facial nerve in the dorsal pontine area is responsible for simultaneous occurrence of INO and horizontal gaze palsy as well as peripheral facial palsy [7].

Clinically Isolated Syndrome (CIS) is defined as a first clinical event suggestive of Multiple Sclerosis (MS) without clinical or radiological evidence of dissemination in time [1,8]. In general, median age at MS onset is approximately 30 years, but ranges from early childhood to older age, with 4-9% of patients having a first clinical event beyond the age of 50 years, often labelled as Late Onset MS (LOMS) [9]. Twenty to eighty-five percent of patients diagnosed with CIS will develop Relapsing-Remitting MS (RRMS) depending on the number of MRI lesions and the presence of CSF OCB at baseline [10,11].

In our patient, multiple periventricular, pericallosal and subcortical T2 hyperintense lesions were already detected at the time of the first inflammatory demyelinating event, thus fulfilling the MS MRI criteria of dissemination in space but not in time. However, a second neurological event four weeks later and the evidence of a new T2 hyperintense lesion now fulfil the criteria for diagnosis of RRMS [1]. Our patient thus represents a rare case of a late onset RRMS in a 74-year-old woman presenting with an eight-and-a-half-syndrome.

Apart from the clinical rarity of eight-and-a-half-syndrome as such, this case serves as a reminder that MS can occur in every period of life

and should be considered in differential diagnosis irrespective of age at onset of neurological symptoms.

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