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One-and-a-half syndrome as first clinical manifestation of multiple sclerosis — a case report and literature review

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To the Editors

One-and-a-half syndrome is a rare neuro-ophthalmic disorder characterised by complete horizontal gaze palsy in one eye with medial gaze palsy in the other eye secondary to injuries of VI nucleus, paramedian pontine reticular formation and median longitudinal fasciculi [1, 2]. Herein, we describe a patient with one-and-a-half syndrome as the first clinical manifestation of MS. To the best of our knowledge, there have been few reported cases of one-and-a-half syndrome manifesting as the first presentation of MS.

In the present study, we describe a patient with one-and-a--half syndrome as the first clinical manifestation of MS that was managed with prednisolone. Due to no clinical improvement, plasmapheresis was initiated.

A previously healthy 35-year-old female presented with sudden onset diplopia. On physical examination, horizontal movement of both eyes was absent except for lateral gaze of the right eye, which was accompanied by nystagmus. Vertical gaze and convergence were intact. Neurological examination revealed impaired coordination tests and hyperactive tendon reflexes in the left upper extremity.

Brain MRI (magnetic resonance imaging) showed multiple T2 hyperintense lesions in juxtacortical and subcortical white matter. Multiple infratentorial lesions were also present in the pons, cerebellum and medulla. Paramedian pontine reticular formation and left medial longitudinal fasciculus were involved. Brain MRI fulfilled the McDonald criteria for dissemination in space and time. The patient received intravenous methyl prednisolone 1,000 mg/day for eight days with partial improvement of ophthalmoplegia followed by six cycles of plasmapheresis every other day, resulting in complete recovery of all neurological symptoms with no relapse over three years of follow up. Her gaze movements were normal and new MRI showed no enhancing lesion in the pontine area.

Multiple sclerosis is the most common autoimmune disorder of the central nervous system. It is characterised by axonal loss, white matter lesions and inflammation. It affects both sexes, with a female preponderance [3, 4]. Optic neuritis is the most common first presentation of MS. Other presentations of MS include sensory or motor deficits, cerebellum dysfunctions, and hyperreflexia [5]. Ocular presentations of MS may include diplopia, oscillopsia, decreased or even loss of vision, uveitis, nystagmus, internuclear ophthalmoplegia, and, rarely, one-and-a-half syndrome [5, 6].

One-and-a-half syndrome is a term coined by Fisher in 1967 to describe a disorder of horizontal gaze movement with clinical manifestation of lateral gaze palsy in one direction with unilateral internuclear ophthalmoplegia (INO) in the other eye [7]. The most frequent causes of one-and-a-half syndrome are vascular diseases such as brainstem ischaemic/ /haemorrhagic stroke, MS, pontine tumours and arteriovenous malformations [2].

To date, five MS patients whose first clinical presentation was of one-and-a-half syndrome have been described in the literature (Suppl.Tab. 1) [1, 2]. In 1983, Wall and Wray reported 20 cases of one-and-a-half syndrome with comprehensive physical examinations and follow ups. They proposed MS as the most common cause of one-and-a-half syndrome as

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14 out of the 20 cases were diagnosed as MS (and two were possibly MS). The most common visual symptom was diplopia, followed by blurred vision and oscillopsia. Interestingly, most concomitant cranial nerve lesions in these patients were unilateral loss of trigeminal sensory and peripheral palsy of facial nerve. Also, eight out of 10 patients with one-and-a--half syndrome experienced full recovery of ocular gaze after 4-16 weeks. Surprisingly, Wall and Wray reported four patients with one-and-a-half syndrome as the first clinical presentation of MS. These patients presented with right side INO. Three of them were female. Gaze movements were fully normalised in two patients after 4–9 weeks, with partial improvement in a third patient after nine weeks [2].

In 2008, Espinosa [1] reported a fifth case of one-and--a-half syndrome as the first clinical presentation of MS, a previously healthy 28-year-old man with sudden diplopia made worse by looking to the right, which was associated with complete left gaze palsy. MRI had showed periventricular and pontine lesions in the white matter and the patient was diagnosed with MS. The patient was not followed up, and therefore no data regarding the recovery status of gaze movement was reported. Our case experienced full recovery of ocular gaze after plasmapheresis, and no relapse of disease has yet been seen.

The treatment strategy of one-and-a-half syndrome depends on the cause of this condition. As vascular and autoimmune diseases are the usual cause, treatments of underlying diseases can lead to improvements in one-and-a-half syndrome. Treatments include, but are not limited to, corticosteroids, immunomodulatory agents and plasmapheresis in autoimmune diseases, thiamin in Wernicke encephalopathy, and surgery or radiation therapy in cases of tumours and arteriovenous malformations. Also, lumbar puncture, the injection of botulinum toxin and supportive treatments have been suggested in some conditions [8, 9]. Martyn et al. reported an MS patient with one-and-a-half syndrome who was managed with dexamethasone. Gaze movements recovered almost completely after six months, but pontine demyelination was found in magnetic resonance imaging [10]. However, we did not find any pontine lesion in our case after three years. Although we observed a dramatic response after plasmapheresis, Garcia-Martin et al. did not find an improvement in their case after plasmapheresis [11].

To conclude, one-and-a-half syndrome can appear as the first presentation of MS. Ophthalmologists as well as neurologists should be aware of this possibility and should consider MS in a differential diagnosis for any patient presenting with one-and-a-half syndrome as a clinically isolated syndrome. They should evaluate the condition meticulously by taking a history, making a clinical examination, and appropriate neuroimaging.

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