Neuro-Behcet's Disease Presented with One-and-a-Half Syndrome

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Abstract: One-and-a-half syndrome is characterized by conjugate horizontal gaze palsy in one direction side and internuclear ophtalmoplegia in the other side. This syndrome occurs with lesions in the dorsal pontine tegmentum involving the ipsilateral PPRF or the abducens nucleus and have to extend the ipsilateral MLF.

The report describes a patient with Behcet disease who developed one-and-a-half syndrome as a result of pontine tegmentum involvement.

Keywords: Behçet disease, one- and -a-half syndrome, horizontal gaze, dorsal pons.

1. INTRODUCTION

Behcet Disease (BD) has been described as both a multi system inflammatory disorder and a vascular disease characterized by inflammatory vasculitis in the vessel wall and vasculopathy [1]. Neurological involvement of BD is a rare manifestation, only seen in 6% of the cases [2].

We report a case of one-and-a-half-syndrome caused by marked dorsal pontine tegmentum involvement of BD.

2. CASE REPORT

A 32 year-old man who had been followed with a diagnosis of BD, applied to our clinic for rapid onset of diplopia. He had been suffering from recurrent oral and genital aphtous ulcers in last few years and his Pathergy test was positive. On admission, neurological examination showed a conscious patient with intact cognitive functions.

Neuro-ophtalmological examination revealed that visual acuity, light reflex and fundoscopy were normal. On leftward gaze, he had an abduction deficit in the left eye and adduction deficit in the right eye. On rightward gaze, he had an adduction deficit in the left eye and right-beating nystagmus in the right eye, which persisted during saccades, pursuit and oculocephalic movements. (Figure **1**) Vertical eye movements and the findings of the rest of the neurological examination were normal.

Magnetic resonance imaging (MRI) showed an acute hyperintense lesion in midline and left dorsal pons. (Figure **2**) Elevated protein content and pleocytosis were detected in the cerebro-spinal fluid (CSF) examination and the patient's CSF was negative for oligoclonal bands.

Intravenous methylprednisolone (1g/day) was administered for 5 days, followed by an oral administration of prednisolone (1mg/kg/day). His clinical symptoms were completely improved within two weeks (Figure **3**).

3. DISCUSSION

One-and-a-half-syndrome consists of an ipsilateral conjugate horizontal gaze palsy in one direction (the "one") and an internuclear ophtalmoplegia in the other direction(the "one-half"), was first desribed by Fisher in 1967 [3]. Therefore, the only horizontal gaze movement preserved is abduction of the contrlateral eye.

This rare condition occurs with lesions in the dorsal pontine tegmentum that involve the ipsilateral pontine reticular formation (PPRF) and/or the sixth nerve nucleus have to extend superiorly to the ipsilateral medial longitudinal fasciculus (MLF) [4, 5].

Close proximity of the PPRF and MLF in the dorsal pontine tegmentum makes them vulnerable to vascular, demyelinating, neoplastic and infectious diseases [4]. These signs were not encountered in reports of Neuro-Behcet Disease. Differential diagnosis from multiple sclerosis (MS) can be difficult in patients with brainstem MRI hyperintensities. Our patient's MRI showed an acute lesion in the midline and left dorsal pontine tegmentum. In our case, the absence of MS plaques in cranial and cervical MRI and negative oligoclonal band of the cerebrospinal fluid helped us with the differential diagnosis of Neuro-Behcet Disease and MS.

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Figure 1: Photographs of the patient's bilateral horizontal gaze before the treatment; show that the patient has impaired movement of both eyes in leftward gaze. In right wars gaze, the patient has impaired adduction of the left eye when the abduction of the right eye and vertical gaze of both eyes are intact.

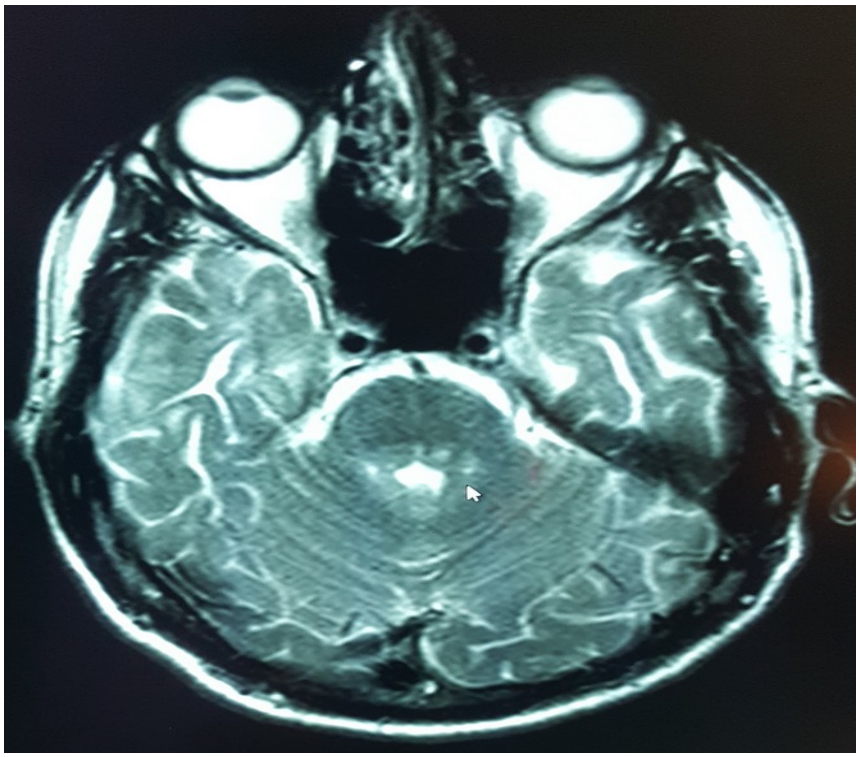

Figure 2: T2 weighted MRI revealed a hyperintense lesion in the midline and left dorsal pans.

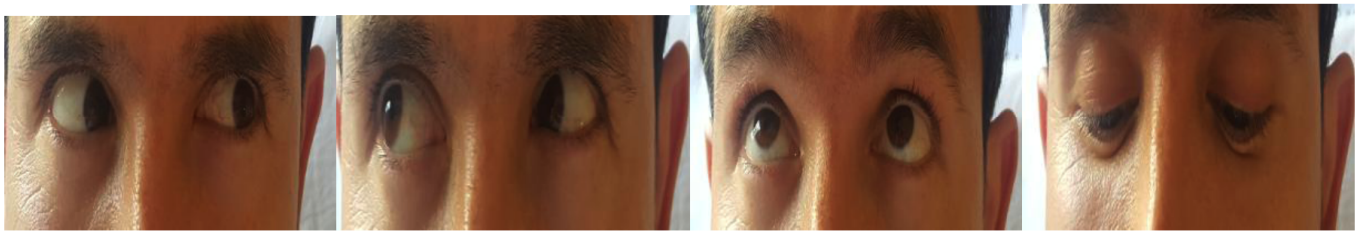

Figure 3: Photographs of the patient's bilateral horizontal gaze after the treatment showing that the patient's horizontal gaze palsy is improved.

From this report it seems that, in keeping with our experience the one-and-a-half syndrome may be an unusual manifestation of BD as a result of pontine tegmentum involvement.

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