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Neuro-Behçet: Pons Involvement with Longitudinal Extension to Midbrain and Hypertrophic Olivary Degeneration

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Key Words
Behçet's disease · Neuro-Behçet · Hiccups · Brainstem · Hypertrophic olivary degeneration

Abstract
A 21-year-old right-handed man developed progressive dysarthria and gait disturbance over 4 months (associated with intermittent hiccups). During that time, he also suffered from uveitis. A physical examination showed pseudobulbar and pyramidal signs and genital and oral ulcers. A brain MRI revealed an extensive lesion mainly located in the ventral pons, with an extension upwards to the midbrain. The inferior olivary nucleus also showed hyperintensity. After the treatment with intravenous methylprednisolone and pulses of cyclophosphamide, he improved. As observed on his MRI, his lesions also improved, except for an increase of the inferior olivary nucleus, consistent with hypertrophic olivary degeneration. Neuro-Behçet tropism for ventral brainstem explains the usual presentation with pyramidal syndrome. Hypertrophic olivary degeneration due to pons involvement could explain the hiccup attacks in a few known cases.

Introduction

Behçet's disease (BD) is a vasculitis with tropism for venules with the extension to multiple organs. The most widely accepted criteria for the diagnosis are the International BD Study Group criteria [1–3].

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The neurological involvement (neuro-Behçet) increases the morbidity and mortality due to BD. There are 2 categories of CNS involvement in BD: parenchymal and nonparenchymal. In the parenchymal one, meningoencephalitis with brainstem and diencephalon lesions usually occurs, whereas in the nonparenchymal category, the onset consists of a venous thrombosis and aneurysms [1, 2].

We report a patient who suffered from parenchymal neuro-Behçet with extensive pons involvement during the multisystemic debut of BD.

Case Report

A 21-year-old right-handed man presented with 4 months of progressive dysarthria, clumsiness in skilled hand tasks, such as writing or drawing, as well as difficulty in walking. He also suffered intermittent hiccup attacks. During that time, he had had 2 episodes of blurred vision in his right eye. This had been diagnosed as uveitis. When questioned, he also confirmed 2 bouts of oral aphthae.

On neurological examination, he suffered from spastic dysarthria with an exaggerated jaw jerk, generalized hyperreflexia with an increased muscle tone and with a right-sided predominance and bilateral extensor plantar reflexes. The gait was also spastic. He also presented scars of previous erythema nodosum and 2 ulcers, an oral one that was already in the process of healing, and a genital one that was still oozing. A 'skin prick test' confirmed a positive pathergy reaction.

A complete blood count showed a white blood cell count of 13,600/μl with 70% neutrophils. The erythrocyte sedimentation rate was 19 after 1 h. A lumbar puncture demonstrated 180/μl leukocytes (55% neutrophils) with normal glycorrhachia (83 mg/dl) and protein levels (37 mg/dl). An MRI showed an extensive lesion mainly localized in the ventral pons, with a longitudinal extension upwards to the midbrain. The inferior olivary nucleus also showed hyperintensity (fig. 1).

The patient was treated with a pulse of methylprednisolone 1 g per day during 5 days, followed by pulses of cyclophosphamide 1 g/m^2, monthly during the first 6 months and once every 3 months after that. At discharge, dysarthria, hands and gait clumsiness had improved. In addition, the hiccup attacks had disappeared. Four months later, a control MRI showed an improvement with less extension and swelling of the lesion. On the other hand, there was an increase of the inferior olivary nucleus, consistent with hypertrophic olivary degeneration (fig. 2).

Discussion

A progressive pseudobulbar and pyramidal syndrome as well as his hiccup attacks were symptomatic of BD. An extensive brainstem lesion, mainly located in the ventral pons, was responsible for the patient's symptoms. It is known that BD has tropism for venules. The shortage of colateral venous circulation in the brainstem seems to explain BD predilection for this location [2, 4].

Following the rostrocaudal axis, a more frequent location of BD is the mesodiencephalic union. Typically, the lesions spread from the midbrain upwards to the thalamus and the basal ganglia [1, 3, 5]. The second most common location for BD lesions is the pons [6], which was where our patient's lesions were found. The comparison between figures 1 and 2 with an improvement of the pons lesion, but with an increase of the inferior olivary nucleus, is

consistent with hypertrophic olivary degeneration (HOD). This HOD may be due to pons lesions with an involvement of the Guillain-Mollaret triangle. Having been described in multiple disorders, HOD has never been reported in BD. Hiccup attacks, a medulla oblongata symptom usually reported for NMO disorders [7] but seldomly described in neuro-Behcet, could be related to HOD.

Following the ventrodorsal axis, BD shows a tendency for the anterior brainstem. Multiple sclerosis, for example, has a tendency for the posterior brainstem. This anterior predilection would explain why BD patients often present with a pyramidal syndrome [8, 9].

There are no clinical trials to support a definite treatment in BD. In this case report, the patient had a clinical and radiological improvement after intense immunosuppressive therapy, including intravenous methylprednisolone, followed by monthly pulses of cyclophosphamide.

Disclosure Statement

The authors state that they have no conflicts of interest.

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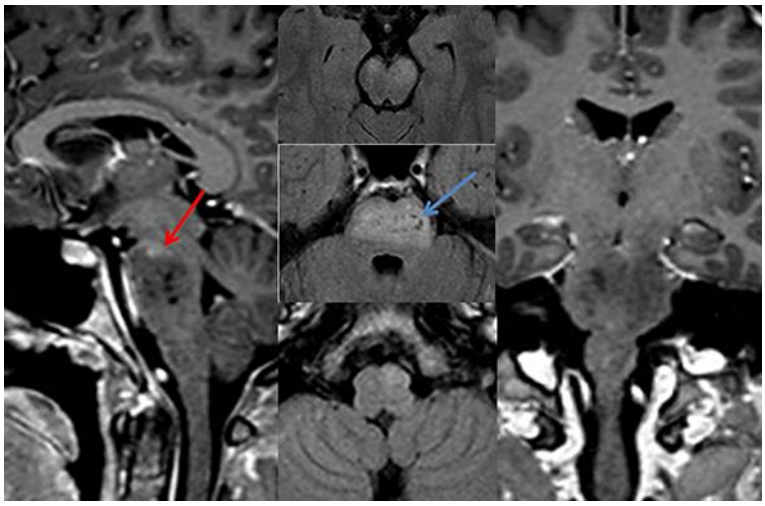

Fig. 1. Sagittal (left column) and coronal (right column) post-contrast T1WI show an extensive hypointense lesion located in both cerebral peduncles and ventral pons with a faint enhancement centered at the top of the pons (red arrow). Axial-FLAIR images in the middle column (from top to bottom: midbrain, pons and medulla oblongata) show an extensive hyperintense lesion mainly located in the ventral pons. There is also a hyperintense image at the left inferior olivary nucleus. The main lesion shows a mild expansion and cystic changes (blue arrow).

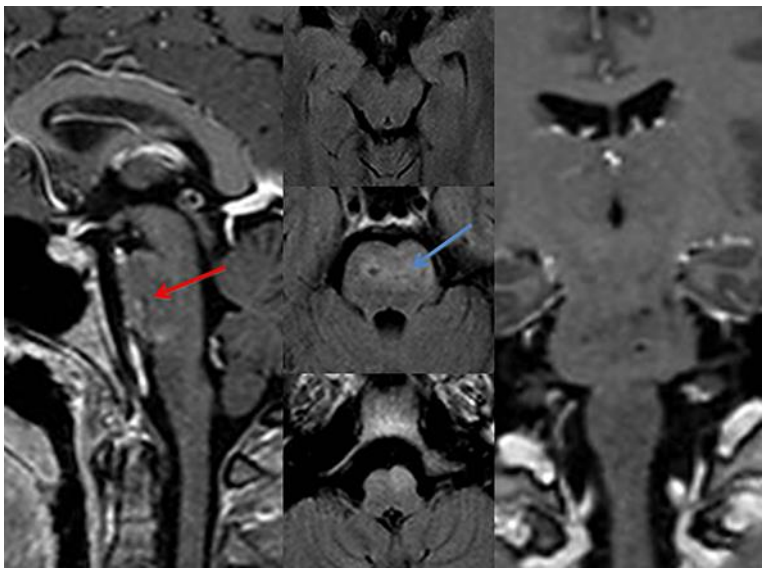

Fig. 2. Axial-FLAIR images in the middle column (from top to bottom: midbrain, pons and medulla oblongata) show a radiological improvement with a lower mass effect and the size of the lesion, yet still showing a cystic change (blue arrow). On the other hand, there was an increase of the inferior olivary nucleus. Sagittal (left column) and coronal (right column) post-contrast T1WI show the lesion with dotted enhancement in the pons (red arrow).