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PSEUDOTUMORAL NEURO-BEHÇET'S DISEASE: A DIAGNOSTIC CHALLENGE AND TREATMENT APPROACH

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ABSTRACT

Neuro-Behçet's disease can present with various neurological manifestations resulting from parenchymal or vascular involvement. Magnetic resonance imaging is the gold standard for diagnosis, showing lesions with an inflammatory pattern. The pseudotumoral form of neuro-Behçet's disease, although rare, can appear as a large lesion mimicking a tumor.Early recognition and prompt initiation of treatment are crucial in the pseudotumoral form to prevent disease progression and improve outcomes. The neurological prognosis can be severe, with recurrent episodes and the potential for disability. Here we presente à demonstrative case of the pseudotumoral form of neuro-Behçet's disease in 30-yearold patient

KEYWORDS :

Pseudotumoral, Neuro-Behçet

MAIN ARTICLE

Clinical Presentation:

We present the case of a 30-year-old patient with no previous medical history who presented with rapidly progressive right hemiparesis and aphasia over the course of one week. The patient also reported symptoms of intracranial hypertension, including headache, vertigo, and sensory disturbances, particularly proprioceptive sensitivity. The patient's general condition remained stable throughout. Clinical examination revealed a conscious patient with right hemiparesis, aphasia, and oral and genital aphthosis. Ophthalmological examination showed no abnormalities, including the absence of uveitis and a normal eye fundus.

Investigation and Imaging Findings:

Laboratory investigations, including lumbar puncture, were within normal limits. The pathergic test was positive. Magnetic resonance imaging (MRI) of the brain revealed patchy lesions on the right side in the cerebellar peduncle and capsulotalamic area, as well as on the left side in the caudocapsulolenticular area, extending into the white matter of the temporal lobe. The lesions appeared isointense on T1-weighted images, hyperintense on T2-weighted images, and showed heterogeneous enhancement. The left-sided lesion exhibited weak and heterogeneous enhancement, while the right-sided lesion showed no contrast enhancement (Figure 1).

Treatment, Outcome, and Follow-up:

Initially, a low-grade glial tumor was suspected; however, after clinical correlation, particularly with the presence of bipolar aphthosis, the diagnosis of Neuro-Behçet's disease with neurological involvement was established. The patient was started on high-dose corticosteroid therapy with methylprednisolone, resulting in improvement of neurological and cutaneous-mucosal symptoms.

A follow-up MRI scan carried out after 4 months of treatment showed considerable regression of the cerebral lesions.

Discussion:

Neuro-Behçet's disease is a chronic, idiopathic, inflammatory multisystemic vasculitis characterized by orogenital ulcerations and recurrent uveitis [1].

The diagnosis is based on the criteria established by the International Study Group for Behçet's Disease, which have a sensitivity of 91% and specificity of 96% [2]. Our case fulfils these diagnostic criteria, presenting with oral and genital aphthosis and a positive pathergic test.

Neurological manifestations in Neuro-Behçet's disease are rare but can significantly contribute to morbidity and mortality. The frequency of neurological involvement ranges from 5% to 20% [1,3]. Neurological symptoms typically appear around the age of thirty, approximately five years after the onset of cutaneous and mucosal signs [3,]. Males are predominantly affected [2,4,5]. In 7.5% of cases, neurological involvement is the initial manifestation of Behçet's disease, and in 3% of cases, it can precede other signs by 1 to 10 years. Delayed diagnosis can occur due to failure to recognize the classic mucocutaneous signs preceding neurological involvement [3,5]. Neurological manifestations can result from parenchymal involvement or macrovascular venous or arterial involvement. Peripheral involvement is rare [3,5,7].

Regarding imaging, MRI is considered the gold standard. Neuro-Behçet often presents as patchy, nodular, or linear lesions with an inflammatory pattern showing hypo- or iso-intensity on T1-weighted images, hyperintensity on T2-weighted and FLAIR images, and variable heterogeneous enhancement. In some cases, the acute lesions may exhibit restricted diffusion. The lesions are predominantly located in the midbrain, pons, basal ganglia, thalamus, and white matter, without a specific preference for periventricular regions [1].

The pseudotumoral form is rare and typically appears as a single large lesion in the brainstem and basal ganglia, occasionally in the hemispheric white matter or cerebellum. On MRI, it appears hypointense on T1-weighted images and hyperintense on T2-weighted images. Perilesional brain edema is sometimes observed, usually of moderate extent, causing mild mass effect, along with variable contrast enhancement [2].

In our case, the lesion presented as a large bilateral mass in the brainstem, NGC, and left temporal lobe white matter, without a preferential periventricular location.

The main differential diagnoses include cerebral gliomatosis, acute disseminated encephalomyelitis (ADEM), primary CNS lymphoma, other vasculitides, particularly lupus, and multiple sclerosis.

There is no consensus on the treatment approach. The neurological prognosis is severe, with recurrent episodes and potential progression to disability [3].

For the pseudotumoral form of neuro-Behçet, prompt initiation of treatment is crucial due to the risk of disease progression. Long-term or even indefinite treatment is often necessary to achieve recovery, prevent relapses, and minimize long-term sequelae[2].

The primary goal of treatment is to control inflammatory lesions using high-dose

corticosteroids (1 mg/kg/day). Intravenous methylprednisolone boluses (typically 1 g for three consecutive days) may be administered initially. To prevent relapses, which commonly occur during corticosteroid tapering, immunosuppressive therapy such as azathioprine (2.5 mg/kg/day) or intravenous cyclophosphamide (1 g every 4 weeks) may be added. Interferon-alpha can also be considered. The duration of treatment is not well defined. Immunosuppressive medications are recommended for a minimum of 2 years [3].

Conclusion :

The pseudotumoral form is rare and presents a diagnostic challenge due to its similarity to tumors, especially when cutaneous and mucosal signs are absent or unrecognized. This can lead to delays in appropriate management and poorer prognosis.

Learning points :

• Neurological involvement in behçet's disease occurs in up to 20% of cases.

• Sometimes neurological symptoms may inaugurate the clinical picture.

• The pseudotumour form of neurobehçet is rare and poses a diagnostic challenge given the similar differential diagnoses and the need for early treatment.

• Neurobehcet's should be considered in young adults with lesions of the brain stem and basal ganglia .

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