CASE REPORT OF NEURO-BEHÇET'S DISEASE

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ABSTRACT

The neurological disease of Behçet's is a manifestation of Behçet's Disease, a rare condition related to autoimmune vasculitis. It causes recurrent inflammation in various parts of the body, including mouth, genital organs, skin, eyes, and in some cases, neurological manifestations. Neurological symptoms vary and can include encephalitis, meningitis, cerebral venous thrombosis, hemorrhages and cerebral aneurysms. Here's a case report of a 59-year-old male patient presenting clinic and radiologic presentation of Limbic Encephalitis with excellent response to imussupression treatment.

KEYWORDS: BEHÇET'S DISEASE; LIMBIC ENCEPHALITIS

INTRODUCTION

Behçet's Disease (BD) is an inflammatory condition characterized by involvement of large, medium, and small blood vessels. Disseminated vasculitis is believed to be the main trigger for systemic manifestations, including recurrent oral ulcers associated with ocular, urogenital, neurological, cutaneous, gastrointestinal, and articular symptoms [1].

When a patient with confirmed Behçet's Disease develops neurological symptoms, it is diagnosed as Neuro-Behçet's Disease. Involvement of the central nervous system is observed in about 9% of individuals with BD, more commonly in males [2].

It can affect both the brain parenchyma and the vascular structures of the central nervous system (CNS) in its non-parenchymal form. Known consequences of Neuro-Behçet's disease include recurrent meningoencephalitis, cranial nerve paralysis, epilepsy, cerebral venous thrombosis, and episodes of diencephalic and brainstem dysfunction that may mimic strokes [2].

The presentation can be acute, with meningoencephalitis or cerebral venous thrombosis, or progressive, with dementia, ataxia, and dysarthria. Cognitive impairment can occur independently of neurological manifestations [3].

CASE REPORT

A male patient, 59 years old, was referred to the Neurology Institute of Goiânia due to abdominal pain associated with headaches and lesions on the tongue. Treatment with antibiotics (Amoxicillin and Clavulanic Acid) was initiated. The patient presented acute memory impairment, with frequent repetitions due to difficulty in retaining new information, starting two days before. In a symptomatological interview, a history of long-standing recurrent headaches was evident, as well as frequent facial pustules and oral aphthae (tongue and buccal mucosa) – in the last two years, he had monthly ulcerative lesions. The patient reported a subacute episode of imbalance, diplopia, and paraparesis five years before, diagnosed at the time with ischemic pontine stroke. He underwent extensive investigation, including tests for vasculitis, but no definitive etiology was found. Retrospective evaluation of cranial images revealed bilateral lesions in the pons that did not respect vascular territories, with a tumefactive effect, vasogenic edema, and contrast enhancement within the lesion suggestive of an inflammatory pattern. Additionally, he had a diagnosis of dyslipidemia and was taking pantoprazole, simvastatin, and aspirin. On physical examination, there was a slight alteration in memory, especially episodic memory, without other focal neurological deficits. The patient reported a fall in the kitchen four days before. Ectoscopy of the left elbow showed prominent inflammatory signs, with significant swelling and redness. During hospitalization, imaging and laboratory tests were requested to clarify the presented pathology. On the first day of hospitalization, Magnetic Resonance Imaging (MRI) of the skull and a neoplastic/infectious screening with Computed Tomography (CT) of the Chest and Abdomen were performed. Cranial MRI showed a lesion with hypersignal and significant edema in the left hippocampal formation, sparing the amygdala, with heterogeneous and irregular leptomeningeal enhancement from the ependymal plexus of the left lateral ventricle and diffusion restriction. There was

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also a band-like lesion in the central region of the pons, with hyposignal in T1, hypersignal in T2, without contrast enhancement or diffusion restriction (Figure 1). This involvement characterizes a clinical and radiological picture compatible with Limbic Encephalitis.

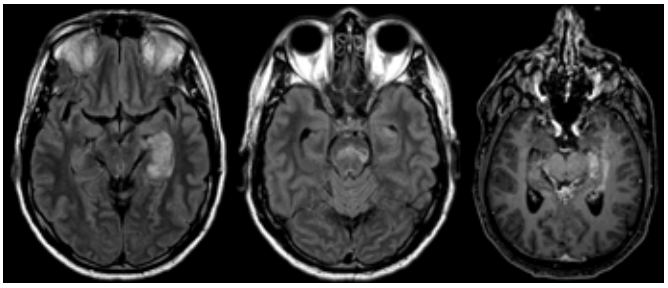

Figura 1: Cranial magnetic resonance imaging. (A) Axial FLAIR, showing vasogenic edema and tumefactive effect involving the left hippocampal formation; (B) Axial FLAIR, demonstrating signal alteration in the pons, bilaterally transversely (band-like), with cavitations, suggestive of a sequela; (C) Axial post-contrast T1, highlighting leptomeningeal and cortical enhancement of the hippocampal formation and contiguous enhancement of the left choroid plexus.

Due to significant inflammation in the left upper limb, the patient underwent an elbow MRI, which revealed olecranon bursitis associated with signs of infectious/inflammatory process around it and a focus of bone edema in the olecranon. Drainage of the collection was performed, and antibiotic therapy was escalated to Ceftriaxone. Abdominal CT showed supernumerary mesenteric lymph nodes. Chest CT showed no alterations. Lumbar puncture (LP) was performed, and the results showed significantly increased cellularity – 348 cells (55 lymphocytes, 15 plasma cells, and 23 monocytes); elevated protein levels (160); and increased lactate of 3.6 mg/dL.

On the second day of hospitalization, additional investigative measures included a normal Electroencephalogram (EEG). Additional laboratory tests performed included: anti-nuclear antibody (ANA) with a fine dense speckled pattern 1:160, non-reactive Anti-DNA, Anti-Sm < 0.7 U/mL, C4 38 mg/dL, Anti-Ro < 0.3 U/mL, C3 228 mg/dL, rheumatoid factor < 20 IU/mL, non-reactive ANCA, and Angiotensin-Converting Enzyme (ACE) 35 U/L, all within normal ranges, except for a significantly elevated erythrocyte sedimentation rate (ESR) with a value of 82.

On the third day, the Pathergy test was performed – the insertion of a 5 mm needle into the forearm, with readings at 24 and 48 hours. The result was strongly positive, with the appearance of a 6 mm papule. With this, the diagnosis of Neuro-Behçet's disease was established, considering the following diagnostic criteria (recurrent oral aphthous ulcers, pseudofolliculitis, and papulo-pustular lesions, positive Pathergy test). Immunosuppressive therapy was then initiated, consisting of methylprednisolone pulse therapy for three days, followed by a course of Cyclophosphamide.

During the 10-day hospitalization, the patient's ESR (erythrocyte sedimentation rate) decreased from 82 on the first day to 23 on the 9th day, and the PCR (C-reactive protein) was 8.7 at discharge. The patient was discharged without headaches, with complete improvement of oral aphthae, resolution of inflammation in the left elbow, and the amnestic picture.

DISCUSSION

Behçet's neurological disease is defined as a set of neurological signs and symptoms in patients with confirmed Behçet's disease (BD). It includes a variety of presentations, primarily involving the central nervous system and, more rarely, the peripheral system. The latter is characterized by neuropathies and myopathies, often in a subclinical manner and confirmed by electrophysiological studies [2].

Manifestations related to the central nervous system (CNS) can be divided into parenchymal and non-parenchymal, with the former being more common and likely affecting the patient in question. Truncal, hemispheric, spinal syndromes, and meningoencephalitis can be clinical manifestations of this subtype. Non-parenchymal involvement includes arterial disorders and venous sinus thrombosis. [4].

Patients with Behçet's disease have a higher risk of experiencing cardiovascular events, such as heart attack and stroke, with the latter being considered a differential diagnosis, especially in patients over 50 years of age [5]. Symptoms such as headache and polyneuropathy can occur secondary to Behçet's disease itself and the medications used to control the disease. In most cases, they represent an underlying uncontrolled inflammatory condition [2].

The diagnosis of Behçet's Disease is based on diagnostic criteria according to the International Study Group (ISG) diagnostic criteria published in 1990 [6], with a new proposal in 2014 that included an expansion of diagnostic criteria to improve sensitivity and specificity [7]. The Pathergy test, a non-mandatory criterion but scoring in favor of the diagnosis, involves skin hyperreactivity followed by trauma with a needle. This test has high specificity, although there are reservations regarding its inconsistent reproducibility and variable sensitivity. The Pathergy phenomenon may be evidence of the endothelial dysfunction characteristic of vasculitis, a group that includes BD [8].

It is important to differentially diagnose the condition from Multiple Sclerosis, Systemic Lupus Erythematosus, Sarcoidosis, CNS infections, and other causes of inflammatory pathologies. Encephalitis, encephalomyelitis, and meningoencephalitis share symptoms with BD, such as headaches, reduced level of consciousness, behavioral changes, and epileptic seizures. Nonetheless, it poses a challenge for neurologists as it relies on the summation of diagnostic criteria [4].

The case report in question not only presents a rare dis-

ease but also brings an extremely unusual clinical and radiological presentation for BD. Limbic encephalitis does not represent a described presentation in the disease, especially with exclusive hippocampal involvement associated with leptomeningeal enhancement with venous pattern restriction, making this report essential for medical literature.

The treatment of the neurological manifestation of BD aims to control the patient's inflammatory condition. The main approach involves the use of intravenous methylprednisolone, followed by oral corticosteroid therapy. Cyclophosphamide, Azathioprine, Methotrexate, and immunobiologics such as Infliximab may be used as indicated, usually required as maintenance therapy [7].

CONCLUSION

The case of the presented patient illustrates the complexity and diversity of DB manifestations, especially when there is an atypical presentation such as limbic encephalitis. Early identification of Neuro-Behçet's Disease and differentiation between its parenchymal and non-parenchymal manifestations are crucial for proper treatment. In this specific case, the patient's favorable response to pulse therapy and subsequent treatment is encouraging and highlights the importance of early diagnosis and appropriate management of the condition. However, long-term follow-up is essential since DB is chronic and requires continuous monitoring to prevent relapses and complications. Ultimately, the case underscores the importance of awareness and education about DB, especially its neurological manifestations, to ensure early diagnosis and effective treatment, thereby improving the quality of life for patients affected by this rare and challenging condition.

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