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Neuro-Behçet in its Pseudo-Tumoral Form: A Case Report

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Abstract **Case Report**

Behçet's disease is a multi-systemic inflammatory pathology whose involvement of the central nervous system remains uncommon. It is characterized by white matter abnormalities like most inflammatory diseases. However, there are atypical forms such as the pseudo-tumor form which is rare. We report the case of a 43-year-old patient with a history of recurrent bipolar aphtosis with an episode of uveitis who presented with functional impotence of the right hemisphere with swallowing and speech disorders. The diagnostic approach led to the diagnosis of the pseudo-tumoral form of neuro-Behçet.

Keywords: Bipolar aphtosis, neuro-Behçet, pseudo tumor.

INTRODUCTION

Behçet's disease is a multi-systemic inflammatory disease characterized by oral and genital bipolar aphtosis and uveitis. Approximately 10-20% of patients has CNS involvement called neuro-Behçet and includes either parenchymal, meningeal or vascular involvement. Pseudotumor expression is rare to exceptional posing a differential diagnosis problem with brain tumors and characterized by marked deficit symptomatology and exceptionally reported as isolated observations.

OBSERVATION

We report the case of a 43-year-old patient with a history of recurrent bipolar aphtosis with episodes of uveitis with functional impotence of the right hemisphere one year ago resolved after three months. He presented with a functional impotence of the right hemisphere that had been evolving for 20 days with swallowing and speech disorders.

The clinical examination showed a spastic right pyramidal syndrome with dysarthria, all evolving in a context of apyrexia and conservation of the general state.

A brain MRI scan was performed, showing extensive left capsulo-thalamic lesions at the pontine and bulbo- Protuberan level, with damage to both cerebral peduncles, most markedly to the left of the red nucleus and the left superior and inferior colliculi and the vermis. These lesions are confluent and extensive in T2 hypersignal, Flair in, T1 iso signal with individualization of some foci in diffusion hypersignal with heterogeneously enhanced ADC restriction was clumped after gadolinium injection. There were also empty lesions on the T2 gradient echo sequence related to haemorrhagic stigmata. They are surrounded by a discrete peri-lesional oedema in Flair hypersignal, the whole creating a pseudo-mass responsible for a discrete mass effect at the supratentorial level on the ventricular crossroads with discrete deviation of the midline without any sign of involvement or upstream ventricular dilatation. There was no evidence of thrombosis on venous MR angiography (Figure 1). The radiological appearance was consistent with a pseudotumoural form of Neuro-Behcet's. The patient was put on corticosteroid therapy and the evolution was favorable.

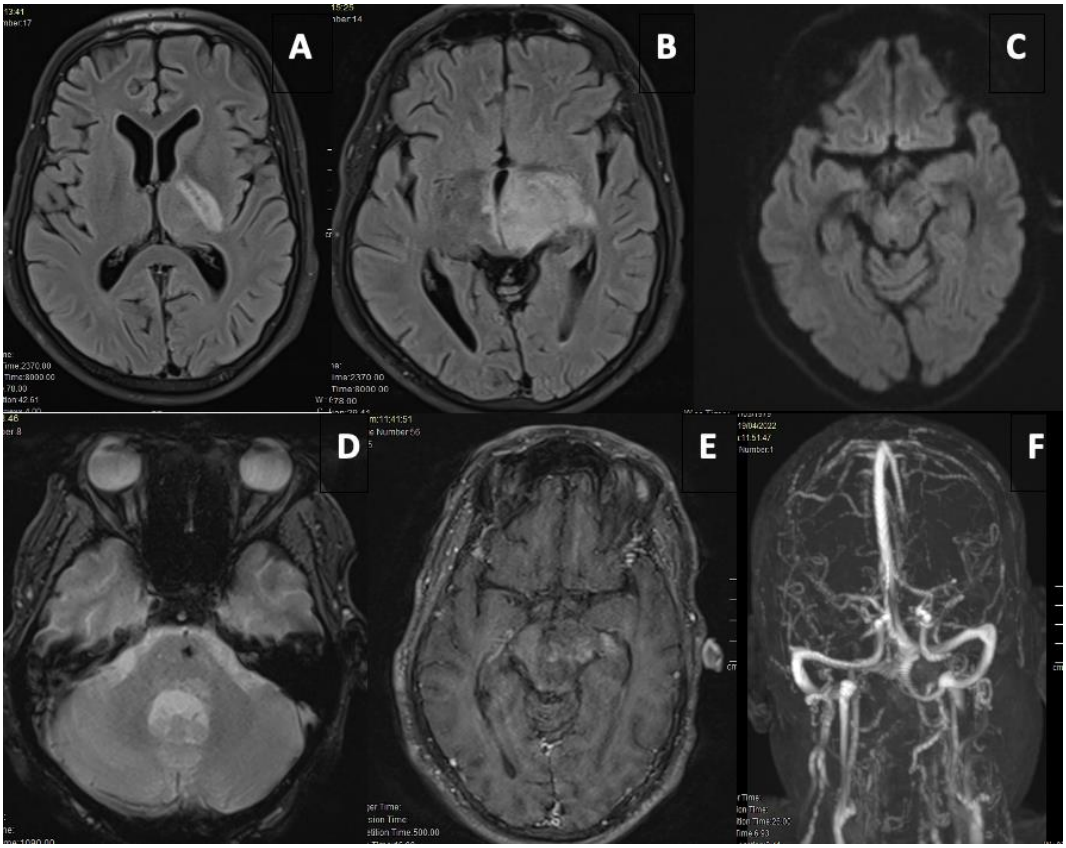

Figure 1

DISCUSSION

The International Consensus Recommendations (ICR) [1] defines neuro-Behcet's disease as all neurological impairments occurring in a patient who has suffered or is suffering from all other systemic symptoms of Behçet's disease. The pseudotumor form of neuro-Behçet has long been reported in isolated cases. According to Mohammed Guini *et al.*, 34 cases of the pseudotumor form were reported in the scientific literature in 2019 [2]. Pyramidal signs and hemiparesis seem to be more frequent than in classical parenchymal forms. It can be inaugural in one third of cases, thus posing a real problem of differential diagnosis with a brain tumor. In our case, it is an inaugural form because our patient was not known to have Behcet's disease.

On MRI, the lesions appear hypointense to iso intense in T1 sequence, with a hyper signal in T2 weighted sequence, variable in size, sometimes confluent and extensive, preferentially affecting the basal ganglia, the midbrain and the diencephalon, with possible extension to the internal capsules. Recent lesions may present a mass effect, with contrast giving a pseudotumor appearance [3]. The main differential diagnoses are glial tumors, lymphomas or infectious and granulomatous lesions [2].

The standard treatment for Neuro-Behçet's disease is high-dose methylprednisolone corticosteroid therapy for 7-10 days, followed by a gradual decrease in the oral doses over a period of 3 to 6 months. This treatment has been reported to be successful in brainstem lesions and in the parenchymal form [4]. Some authors hypothesis that this result is related to the oedema surrounding the lesions, which regresses after administration of corticosteroids.

CONCLUSION

Neuro-Behcet's is a rare vasculitis characterized by predominantly vascular and encephalic involvement. The tumor-like form is rare and poses a problem of differential diagnosis with tumor lesions. In these cases, the clinical context plays a very important role and is a key element of the diagnosis.

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