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CASE REPORT PEER REVIEWED | OPEN ACCESS

Neuro-Behçet's syndrome presented as multiple brain lesions

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ABSTRACT

Introduction: Behçet's syndrome is a multisystemic, chronic inflammatory disease characterized by recurrent oral and genital ulcers, ocular inflammation, arthritis, and skin lesions. Neurological impairment makes the severity of Behçet's disease, it most often occurs during its evolution and can be concomitant or inaugural. We describe a case of neuro-Behçet's syndrome presenting with features mimicking brain tumors.

Case Report: We report the case of a 38-year-old man admitted to the emergency department for progressive tetraparesia associated with vomiting and headache. Regarding the magnetic resonance imaging (MRI) aspect, the diagnosis of brain tumor was first suspected, but the diagnosis was rectified considering the history of Behçet's disease and the existence of bipolar aphtosis. The patient was transferred to the neurology department for management.

Conclusion: Neuro-Behçet's disease is rare, including the pseudo-tumoral form, and can exceptionally be its initial mode of presentation. It can be established in two different patterns: parenchymal and non-parenchymal. Nowadays, the prognosis of neuro-Behçet's disease is improved by early management and better therapeutic codification.

Keywords: Brain tumor, MRI, Neuro-Behçet's disease

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INTRODUCTION

Behçet's syndrome is a multisystemic, chronic inflammatory disease characterized by recurrent oral and genital ulcers, ocular inflammation, arthritis, and skin lesions [1]. Neurological impairment makes the severity of Behçet's disease, it most often occurs during its evolution and can be concomitant or inaugural [2]. We describe a case of neuro-Behçet's syndrome presenting with features mimicking brain tumors.

CASE REPORT

We report the case of a 38-year-old man, with a history of Behçet's disease with recurrent oro-genital ulceration, under colchicine, admitted to the emergency department for progressive tetraparesia associated with vomiting and headache related to intra-cranial hypertension.

At the clinical examination, the patient had weakness of arms and legs with dysarthria. The examination of the oral cavity and genital organs found bipolar aphtosis. The ophthalmological examination was normal showing no uveitis with a normal eye ground. Results of routine laboratory tests were within normal limits, with normal cerebrospinal fluid (CSF).

Brain magnetic resonance imaging (MRI) showed multiple intra-parenchymatous lesions, bilateral and diffuse, well circumscribed, of cortical and subcortical region, thalamic nuclei, and capsulo-lenticular bilateral producing a mass effect with a compression of the adjacent lateral ventricle. The lesions are hypointense in T1, hyperintense in fluid-attenuated inversion recovery (FLAIR) with nodular and ring enhancement (Figure 1).

Regarding the MRI aspect the diagnosis of brain tumor was first suspected, but the diagnosis was rectified considering the history of Behçet's disease and the existence of bipolar aphtosis. The patient was transferred to the neurology department. The initial treatment was based on methylprednisolone bolus with clinical and radiological monitoring.

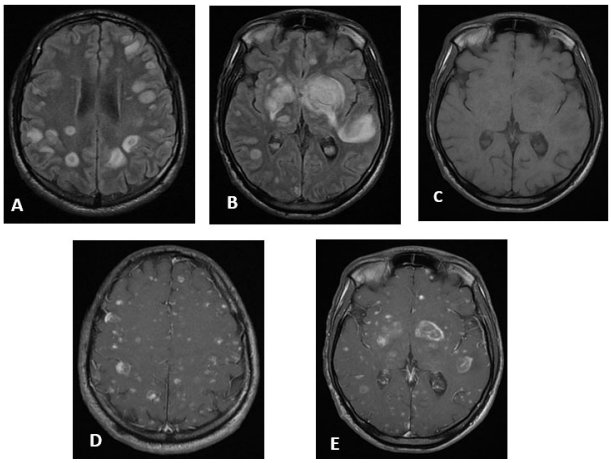

Figure 1: (A) to (E): Brain MRI exam showing in (A) and (B) axial FLAIR acquisition multiple hyperintense, diffuse, circumscribed parenchymal lesions, cortical and subcortical, in thalamus and capsule-lenticular, and bilateral. In (C), T1-weighted acquisition, larger lesions are hypointense, and after gadolinium in (D) and (E), it shows marked ring enhancement in larger lesions and nodular enhancement in small ones.

DISCUSSION

Behçet's disease is a rare systemic vasculitis, with unknown origin, first described in 1937, with predominance of skin-mucous and eye lesions [3].

Neurological impairment is highly polymorphic and observed in 10% to 30% of cases [1]. It occurs in the 2–4 years after the first signs, with a discrete male predominance in which the impairment seems more severe. However, it can be inaugural in 7.5% of cases, making a real differential diagnostic problem with a brain tumor [4].

It is frequently revealed by headache, motor deficit, dysarthria, and is often explained by pyramidal syndrome, cranial nerves involvement, and bulbar syndromes [1].

Neurological manifestations can rise from two main mechanisms: macrovascular or parenchymatous impairment [2].

Magnetic resonance imaging is the examination of choice for radiologic evaluation of neuro-Behçet's disease, especially at the brainstem level. It is superior to computed tomography (CT) for exploring the posterior cerebral fossa. Coupled with angio-MR, it allows vascular exploration with more accuracy [3, 4].

Parenchymatous lesions are the most common. They are nodular or even confluent, hyperintense in T2, iso- or hypointense in T1 with an inconstant enhancement of variable appearance. Hemorrhagic component can be seen in the acute phase as well as a mass effect. This disease predominates in the posterior fossa with pontine predilection and an evocative brain atrophy. It also affects with a decreasing frequency in the internal capsule, white substance, and central gray nuclei with no particular predilection for the periventricular regions. Thickening and meningeal enhancement can be seen. Vascular disease involves venous and arterial cerebral thrombosis. Spinal cord injury predominates in the cervical and thoracic levels, and it appears as a small and multifocal hypersignal T2 [2].

These features are not specific, and that makes differential diagnosis with other vasculitis, granulomatosis, or even a glial tumor lesion. The correct diagnosis is extremely important because it can avoid unnecessary neurosurgical approaches [4].

Treatment of the pseudo-tumor form of neuro-Behçet's disease must be initiated as soon as possible after diagnosis to improve the speed of recovery and to reduce symptoms to a greater extent [4]. It consists of intravenous bolus corticosteroids associated with immunosuppression, in different combinations [1].

Despite treatment, the natural progression of the disease is by attacks and remissions. However, Behçet's neurological disease has a high mortality rate, up to 25% within the first year after developing neurological manifestations [3].

CONCLUSION

Pseudo-tumor form of neuro-Behçet's disease is rare and can exceptionally be its initial mode of revelation. It can be established in two different patterns: parenchymal and non-parenchymal. Nowadays, the prognosis of neuro-Behçet's disease is improved by early management and better therapeutic codification.

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Author Contributions

Amalik Sanae – Conception of the work, Design of the work, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Imrani Kaoutar – Conception of the work, Analysis of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

El Menaoui Ouadie – Conception of the work, Design of the work, Acquisition of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Authors declare no conflict of interest.

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All relevant data are within the paper and its Supporting Information files.