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CASE REPORT PEER REVIEWED | OPEN ACCESS

Neuro-Behçet's disease revealed by diencephalo-mesencephalic impairment

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ABSTRACT

Introduction: Behçet's disease is a systemic variable vessel vasculitis with unknown cause. Neurologic involvement known as neuro-Behcet's disease (NBD) is often diagnosed in patients who present neurological symptoms and radiological lesions of the central nervous system.

Case Report: A 42-year-old woman with a history of orogenital ulceration presented with behavioral disorders and heaviness of left hemibody. Brain magnetic resonance imaging (MRI) was performed and showed a pseudo-tumor diencephalo-mesencephalic area with T2 and Flair hypersignal, a slight hypersignal in diffusion sequence, and no contrast uptake.

Conclusion: Pseudotumoral NBD is a rare but severe manifestation of Behçet's disease. Cerebral MRI is the most efficient method to explore, detect, and monitor parenchymal lesions.

Keywords: Brain MRI, Cerebral pseudo-tumor, Neuro-Behçet

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INTRODUCTION

Behçet's disease is a systemic variable vessel vasculitis with unknown cause whose main symptoms include skin-mucosa lesions and recurrent oral and genital ulceration [1].

Neurologic involvement known as Neuro-Behcet's disease (NBD) is uncommon with a highly variable prevalence (5–59%) [1, 2]. It is often diagnosed in patients with known Behçet's disease who present neurological symptoms and radiological lesions of the central nervous system. We present herein a case of pseudotumoral form of NBD.

CASE REPORT

A 42-year-old woman with a history of orogenital ulceration presented with behavioral disorders and heaviness of left hemibody, and headache. Physical exam showed pyramidal syndrome with left hemiparesis.

Brain MRI was performed in flair (Figure 1), diffusion (Figure 2), T1, T1 injected sequences (Figure 3), T2 (Figure 4), Gradient Echo, and 3D TOF T1, showing a diencephalo-mesencephalic lesion with hypersignal in T2 and flair, slight hypersignal in diffusion sequence and no contrast uptake. No venous thrombosis or arterial abnormality was detected.

DISCUSSION

Behçet's disease is a systemic vasculitis whose neurological involvement, sometimes severe, is found in approximately 15% of cases [1, 2]. Neuro-Behçet's disease pseudotumoral involvement has its own clinical

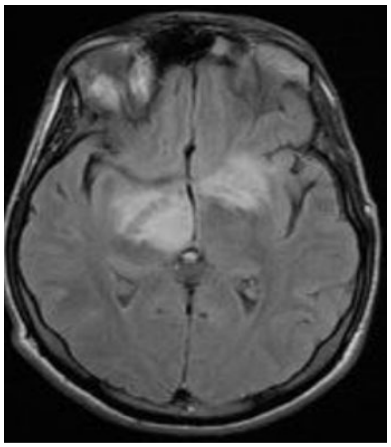

Figure 1: Axial scan in flair sequence: bilateral diencephalo-mesencephalic hypersignal predominant on the right.

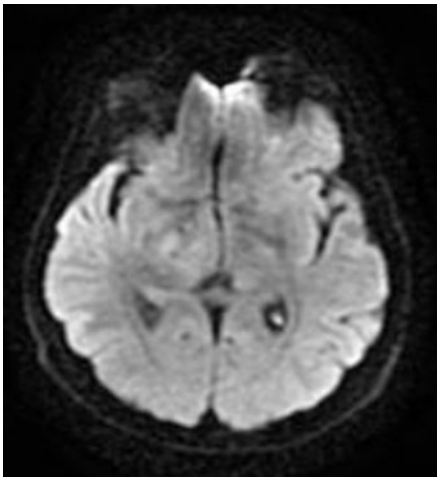

Figure 2: Diffusion sequence: no water restriction on the diffusion sequences.

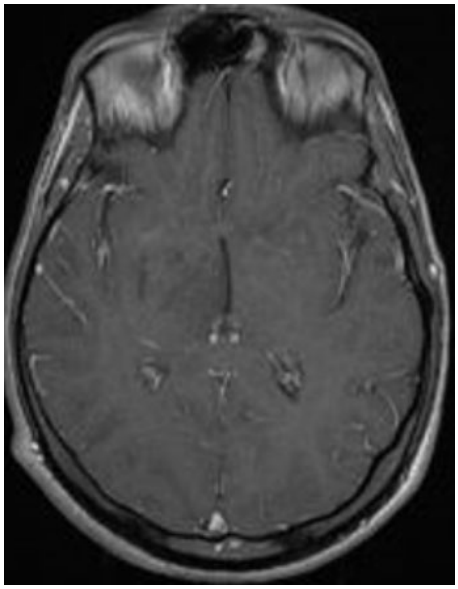

Figure 3: Axial scan in T1 after injection: there is no contrast enhancement.

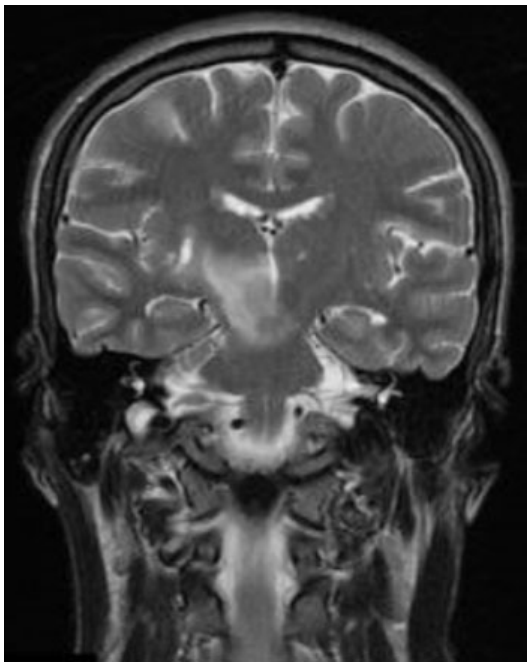

Figure 4: Coronal scan in T2 sequence: right mesencephalo-thalamic hypersignal.

characteristics: pyramidal signs and hemiparesis seem more frequent than in classic parenchymal forms [3].

The most common and most suggestive brain lesion is diencephalo-mesencephalic involvement, which is found half the time, followed by bulboprotuberant involvement in 40% of cases [4]. This lesional topography reflects a vasculitis affecting the small vessels predominating in the venous compartment. Mesencephalo-diencephalic lesions are often bilateral and asymmetrical, with contrast uptake in the initial phase [4, 5]. The differential diagnoses being mainly tumoral (glioblastomas or lymphomas) or infectious (tuberculous abscess) [5, 6].

In the context of brainstem involvement, cervical cord involvement by contiguity can be observed. Conversely, isolated myelitis is rarer, mainly seen as extensive lesions in height. It is most frequently a multifocal transverse myelitis, located in the cervical and dorsal regions, and which can be a factor of poor prognosis [7, 8].

The useful imaging examinations for the diagnosis of neuro-Behçet are head computed tomography (CT) with injection, and angio-MRI, with morphological sequences: T1, T2, gradient echo, diffusion sequences with ADC mapping [9–12].

The lesions are asymmetrical and mainly distributed in the meso-diencephalic junction (46%), the pontobulbar (40%), hypo-thalamo-thalamic (23%), and the central gray nuclei (18.5%). The telencephalon is more rarely affected (7.7%) as well as the marrow (4.6%) [11, 12]. Several areas can be affected in the same patient. It should be noted, however, that the imaging may be normal in approximately 10–20% of cases, particularly in the acute forms, where the abnormalities may appear secondarily

to the MRI [13]. Conversely, pathological imaging with neurologically asymptomatic Behçet disease is known as subclinical neuro-Behçet.

The lesions are hypointense in T1 sequence and hyperintense in T2 sequence, with an edematous area around the wound. In the acute phase, they take on contrast, with typical characteristics of inflammatory damage [9–14].

The predilection for meso-diencephalic involvement could be explained by the venous anatomy. Indeed, telencephalic venous drainage occurs in two directions, through the existence of multiple anastomoses between the superficial, pial veins, and the basilar vein of Rosenthal. In the brainstem, these anastomoses are rarer [6, 9, 10].

In the pseudotumor forms, the lesions appear in hyposignal in T1 and in hypersignal in T2 with significant vasogenic edema and peripheral contrast uptake after injection of gadolinium [6, 15].

Di and telencephalic lesions are most often located in the white matter, deep or subcortical. They appear in T2 hypersignal and do not enhance. There are very few lesions of periventricular white matter, unlike demyelinating pathologies such as multiple sclerosis [15, 16].

CONCLUSION

Pseudotumoral NBD is a rare but severe manifestation of Behçet's disease. It should be brought up in front of cerebral mass as a differential diagnosis. Cerebral MRI is the most efficient method to explore, detect, and monitor parenchymal lesions of the neuro-Behçet and that of the vessels.

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Author Contributions

Jamal Aouifi – Conception of the work, Design of the work, Acquisition of data, Analysis 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

Houda Bouchama – Conception of 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

Hajar Eljouadi – Acquisition of data, 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

Ahmed Hanine – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation 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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Conflict of Interest
Authors declare no conflict of interest.

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