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Case Study

Psychiatric Features ushering Neuro-Behçet's Disease: A Case Report

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ABSTRACT

Nonspecific psychiatric manifestations can inaugurate Behçet's disease. Through a clinical observation and a review of the literature, we point up the clinical polymorphism of neuropsychiatric manifestations in Behçet's disease and take up main therapeutic management.

Keywords: *Behçet's disease; Neuro-Behçet's disease; Vasculitis; MRI; Depression*

Behçet's disease (BD), first described by the Turkish dermatologist «Hulusi Behçet» in 1937, is a systemic vasculitis, with a male predominance. Geographical distribution of this disease interests particularly populations originating from the Middle East, Japan and countries around the Mediterranean basin, including Tunisia [1]. The prevalence of BD is 0.1 to 7.5 per 100000 people in Europe and the United States. In North Africa, it is 110 per100000 people in Tunisia and it is frequent in Morocco as more than 900 cases were reported between 1977 and 2002.

The initial *symptoms occur in* young adults (20–30 years) with an average age of diagnosis between 32 and 36 years [2, 3]. BD is a chronic inflammatory disease, clinically characterized by oral or more often bipolar (oro-genital) aphtosis, associated with cutaneous, ocular, articular, neurological, cardiovascular and intestinal systemic symptoms [4].

BD diagnosis is clinical since there is no pathognomonic biological or pathological marker. The most widely accepted criteria for the diagnosis of Behçet's disease are the criteria of the American Colleges of Rheumatology (ACR) 2007 [5]

Neurological manifestations in Behçet's disease are rare and are one of the leading causes of morbidity and mortality [6]. The frequency of neurological manifestations varies from 2.3 to 44% depending on the inclusion criteria and the ethnic origin of the patients. They are more common in men than in women: The sex ratio in Tunisian patients is 2.7. [7].

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Several types of neurological manifestations can be seen during BD. Neuro-Behçet diagnosis is done on the identification of a small-caliber blood vessels vasculitis with venous predominance in the cerebral parenchyma. These lesions can spread in both hemispheres, brain stem, spinal cord, and meninges [7].

Psychiatric features are associated to the *neurologic manifestations of Behçet's disease*. They are for the most part secondary to the disease course and to the fatigue, functional deterioration and socio-occupational disability caused by it. There is rarely a direct organic lesion of the central nervous system [8,9]. Non-specific psychiatric manifestations may also inaugurate Behçet's disease.

Through a clinical observation, we report a rare case of neuro-Behçet with a mood disorder onset. We illustrate clinical polymorphism of psychiatric symptoms while reviewing therapeutic management.

Clinical case

MJ is a 30 years old Tunisian man, with no personal noteworthy medical history. His mother had a mood disorder. For four months after a romantic breakup, he has been depressed and having obsessive ideas. The situation *worsened* due *to* suicidal ideation and so he was hospitalized in a psychiatric department at Razi Hospital, Tunisia for depression with mixed features and undertook olanzapine and sodium valproate. He shortly developed a drug allergy including trunk rash and palpebral edema to chlorpromazine, and benzodiazepines (diazepam and clorazepate) initiated as an urgent sedative medication. During the first week of his hospitalization, MJ reported a weakness on the left sid**e** of his body. The neurological examination confirmed a left pyramidal syndrome.

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A brain MRI showed a thalamic lesion of the right brain stem and the pons in iso-signal intensity in T1 and a hyper-signal intensity in T2 with nodular enhancement (Figure). The physical examination revealed a pseudo-folliculitis over his back and a left scrotal aphtosis scarring. MJ also reported recurrent oral aphtoses (> 4 times / year). The pathergy test was positive. There was no evidence for uveitis or/and progressive vasculitis in the eye examination. The diagnosis of a neuro-Behçet has been *established* and the patient received *steroid* therapy.

DISCUSSION

We report a particularly interesting case of Neuro-Behçet. The distinctiveness of our clinical case is the reveling psychiatric symptoms. The reports of such cases are rare, and this is to our knowledge the first case reported in Tunisia.

The diagnosis of Behçet's disease is essentially clinical and has no biological criteria. The HLA system Grouping has only an epidemiological interest. In our case, the patient had four symptoms (Table) of the diagnostic criteria for Behçet's disease according to the American College of Rheumatology 2007 [7].

Table: Diagnostic criteria for Behçet's disease according to "the American College of Rheumatology (ACR) 2007 "

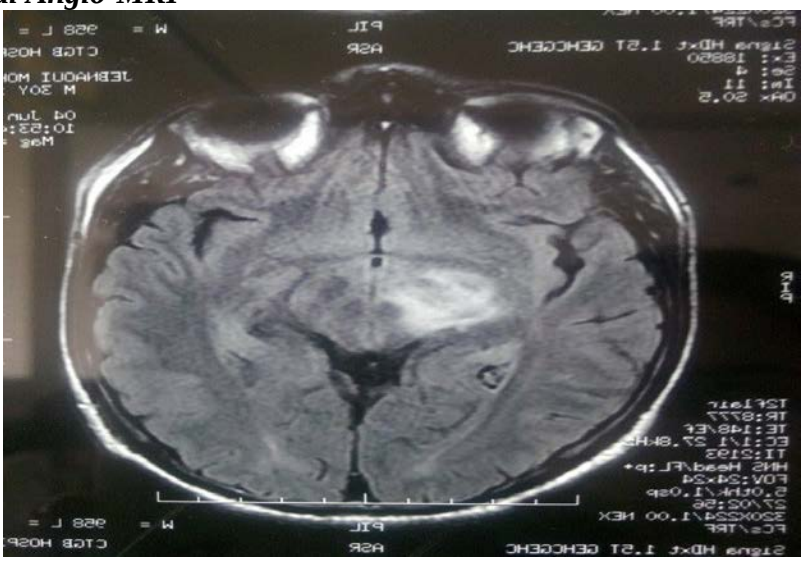
		symptoms Observed with the patient
Oral aphtosis	1 point (mandatory criterion)	Yes
Genital Aphtosis	2 points	Yes
Skin lesions	1 point	Yes
Ophtalmic symptoms (uveitis)	2 points	no
Positive pathergic test	1 point	Yes

Neurological manifestations specific to BD interest more the central nervous system (CNS) than the peripheral system (PNS). Localization in the CNS results from two main mechanisms:

- Angio-Behçet or Vasculo-Behçet: It affects large brain vessels causing cerebral venous thrombosis (CVT). It is the most common form.
- Neuro-Behçet, which consists of a brain parenchymal localization, in relation to a vascularitis of small-caliber vessels, predominantly venous. It may affect the hemispheres, brain stem, spinal cord, and meninges [7].

Our patient has a parenchymal neurobehçet, revealed by a left pyramid syndrome at the neurological examination and his Angio-Cerebral MRI showed a thalamic lesion of the right cerebral peduncle and pons in iso-signal T1 with a hyper-signal T2 with a nodular enhancement (image). Angio-MRI allows for the diagnosis and follow-up of the parenchymal lesions and the cerebral vascular mapping [7].

Image: Cerebral Angio-MRI



In the acute phase, the MRI *shows* a *range* of various types of lesions of the white substance in hyper-signal T2 and FLAIR, and in variable signal T1, often in iso-signal. Contrast injections show various modes of enhancement. This condition is most common in the brain stem and extends to the diencephalus. The temporal and frontal subcortical white substance, the inner capsule, thalamus and central grey nuclei are less frequently affected. Lesions are typically unilateral [10, 11].

Psychiatric and cognitive manifestations have been reported. Sudden personality changes were observed in patients with neuro-Behçet [12]. Anxiety and depression are the most common psychiatric symptoms, reaching up to 86% in some studies with prevalence higher than in other autoimmune disorders. Generally, they result from the disease and its impact on body image, quality of life and socio-professional functioning [13].

Depression and anxiety symptoms can also take place as a clinical feature that precedes the onset of typical BD symptoms, comparable to our patient. The role in managing stress in these patients may be relevant to the genesis of psychiatric features [14, 15].

In the literature, it seems relatively common for patients with BD to develop neurobehavioural syndrome characterized by euphoria, bipolar disorders and paranoid attitudes, disinhibition and indifference towards their disease, defined as «neuro-psycho Behçet». But the underlying pathogenic mechanism remains undetermined until this day. On the other hand, the prevalence of psychiatric symptoms during relapses or, in some cases, during the phases preceding the reactivation of the disease suggests that psychiatric disorders in BD might represent a sub-psychiatric unit or a clinical characteristic distinct from the disease [16]. An association with bipolar disorders was also described [17]. Currently, there is little data available on the therapeutic management of "neuro-psycho-Behçet", but in some reported cases, these symptoms have resisted conventional psychiatric treatment. A combination of mood-stabilizing drugs, such as sodium valproate, carbamazepine and olanzapine, combined with a specific treatment for BD may lead to some improvement in psychiatric symptoms.

CONCLUSION

Isolated psychiatric symptoms may inaugurate neuro-Behçet's disease but are more frequently described during relapses. Despite the scarcity of neuro-Behçet in psychiatric consultation in Tunisia, the practical value of an early and adequate diagnosis should be emphasized, given the resistance to usual therapies. As a result, the occurrence of atypical psychiatric manifestations in young man with no history of psychiatric disorders, should seek to interrogation about classic bipolar aphtosis of BD.

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Conflict of Interest
The authors carefully declare this paper to bear not conflict of interests

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