CASE REPORT

Mixed Neuro-Behçet: Exceptional Pattern of Neurological Involvement in Behçet's Disease

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ABSTRACT

Behçet's disease is a cosmopolitan systemic vasculitis, particularly common in countries located on the old "Silk Road." Neurological involvement in Behçet's disease (neuro-Behcet) is one of the most serious complications and condition the prognosis of the disease. It mainly affects the central nervous system and is subdivided into two types: Parenchymal and non-parenchymal involvement (cerebral venous thrombosis and cerebral arterial aneurysms). The concomitant occurrence of these two neuro-Behcet subtypes defines the mixed neuro-Behcet which remains exceptional and very challenging for clinicians. We report an original case of acute mixed neuro-Behcet in 42-year-old Tunisian man.

Key words: Behçet's disease, cerebral vein thrombosis, neuro-Behçet, non-parenchymal neuro-Behçet, parenchymal neuro-Behçet

INTRODUCTION

Behçet's disease was first described in 1937 by the Turkish dermatologist Prof Hulusi Behçet in front of the classic triad: Recurrent oral aphthous ulcers, genital ulcers, and anterior uveitis with hypopyon.[1] It was classified since 2012, according to the Chapel Hill consensus for the nomenclature and classification of systemic vasculitis, as a systemic vasculitis affecting the vessels of the body of any size (large, medium, and small) and of all types (arteries, capillaries, and veins).[2]

It is a cosmopolitan disease, particularly common in countries located on the old "Silk Road."[3,4]

The possible clinical manifestations of this vasculitis are very polymorphic; neurological (neuro-Behcet), cardiovascular (angioBehcet), ophthalmological (oculoBehçet), and digestive (enteroBehcet) disorders are the most serious and condition the prognosis of this disease.[4-7]

Neurological involvement in Behçet's disease mainly affects the central nervous system and is subdivided into two types: Parenchymal involvement and non-parenchymal involvement.[6,8] The concomitant occurrence of these two neuro-Behcet subtypes defines the mixed neuro-Behcet which remains exceptional and very challenging for clinicians.[8,9]

We report an original case of acute mixed neuro-Behcet in 42-year-old Tunisian man.

Observation

A 42-year-old Tunisian man diagnosed with Behçet's disease since the age of 18 year was admitted in our department for meningoencephalitis. The diagnosis of Behcet's disease was retained in front of the association of recurrent oral aphthous, genital ulcerations, pseudofolliculitis, positive pathergy test, and positive HLA B51. He was treated with colchicine and salicylate with good outcome.

Its current symptomatology dates back to 2 days with the onset of fever and headache, then agitation, mental confusion,

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and disorientation. His wife reported that he had stopped his treatment for a month.

The somatic examination noted a fever at 38.5°C, multiple oral and genital ulcerations, temporospatial disorientation, pyramidal syndrome, meningeal syndrome, and cerebellar syndrome. The hemodynamic and respiratory states were stable, and there were no focal deficits, skin lesions of vasculitis, lymphadenopathy, or organomegaly.

The basic biological tests showed erythrocyte sedimentation rate at 100mmH1, C-reactive protein at 18mg/l, and leukocytosis at 12,300/mm^3 with 80% of neutrophils. The other biological tests were within normal limits: Hemoglobin, platelets, creatinine, glycemia, calcemia, plasma ionogram, transaminases, lipid parameters, thyroid hormones, serum protein electrophoresis, and urinalysis. Procalcitonin was negative eliminating an infectious origin to this biological inflammatory syndrome.

The lumbar puncture showed aseptic lymphocytic meningitis. Direct examination and culture of cerebrospinal fluid were negative.

Cerebromedullary magnetic resonance imaging (MRI) objectified multiple punctate lesions of the deep white matter (particularly periventricular) with low signal in T1-weighted images [Figure 1], high signal in T2-weighted and fluid-attenuated inversion recovery images [Figures 2 and 3], and on the angio-RM a thrombosis of the superior longitudinal sinus [Figure 4].

The electrocardiogram, chest X-ray, thoracoabdominopelvic CT scan, and transthoracic ultrasound were without abnormalities.

Thus, the diagnosis retained was that of neurological involvement of Behcet's disease with mixed pattern (parenchymal neuro-Behcet and cerebral vein thrombosis).

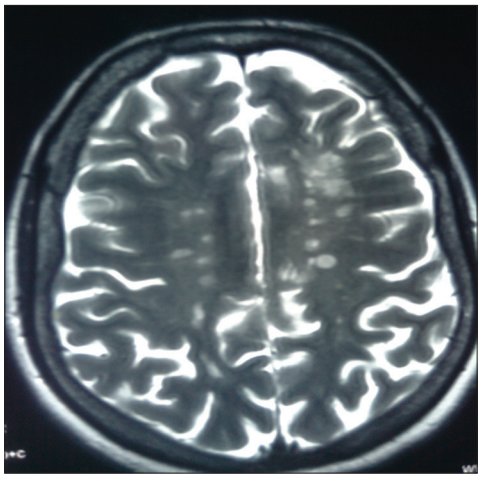

Figure 2: Axial T2-weighted cerebral magnetic resonance imaging: Multiple deep white matter lesions in hypersignal

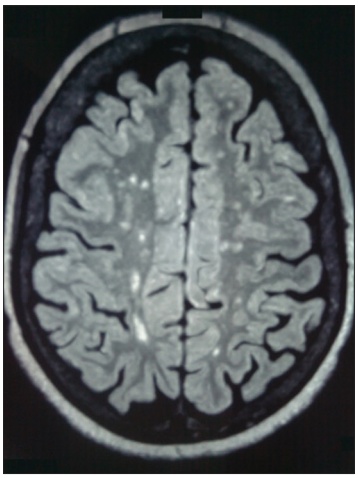

Figure 3: Axial fluid-attenuated inversion recovery-weighted cerebral magnetic resonance imaging: Multiple deep white matter lesions in hypersignal

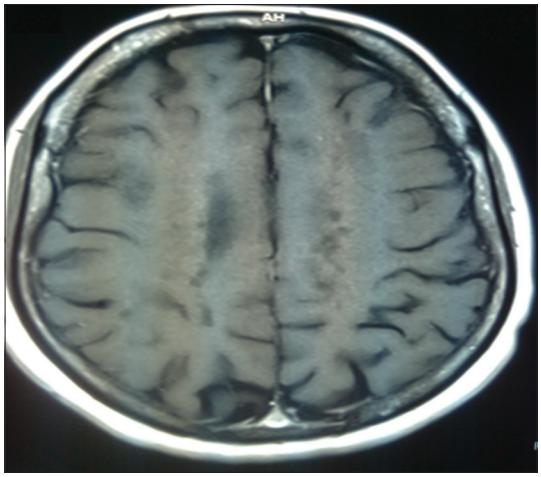

Figure 1: Axial T1-weighted cerebral magnetic resonance imaging: Multiple deep white matter lesions in hyposignal

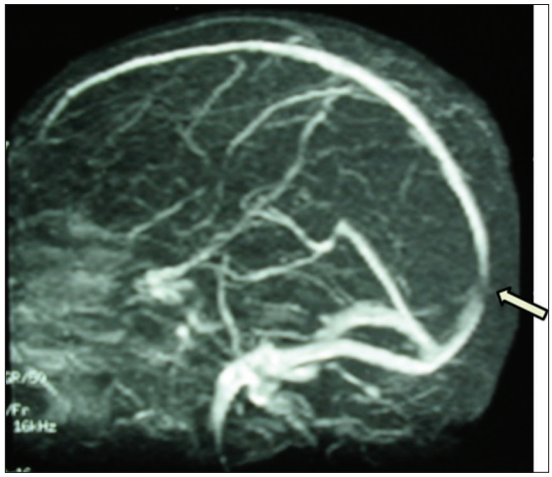

Figure 4: Angio-RM and lateral view: Thrombosis of the superior longitudinal sinus

The patient was treated with systemic glucocorticoids: Three boli of methylprednisolone at a dose of 1 g/d relayed by prednisone per os at the dose of 1 mg/kg/d, colchicine 1mg/d, intravenous cyclophosphamide at a dose of 15 mg/kg/month, and low-molecular-weight heparin in curative dose then relay with acenocoumarol at a dose of 4 mg/d.

The evolution was favorable with apyrexia and normalization of the state of consciousness from the 2nd day, the normalization of the biological tests of inflammation from the 7th day, and normalization of cerebral MRI and angio-MR at the 6th month.

DISCUSSION

Neurological involvement during Behcet's disease was described first by Knapp in 1941[10] and in 1961 Bienenstock and Margulis reported the first description of angiographically proven dural sinus thrombosis in a patient followed for this vasculitis.[11] Since then, the spectrum of neurological involvement of Behcet's disease has been widening.[9]

Neurological damage during Behçet's disease can affect the central nervous system (the most frequent and the most serious) or the peripheral nervous system (much rarer and with better prognosis).[6,8,9,12]

The overall prevalence of neuro-Behcet is variously estimated in the series: 2.2-49%.[13-15] This great disparity is explained by the nature of recruitment (internists, neurologists, or rheumatologists) and by the diagnostic criteria adapted for neuro-Behcet (some authors include isolated headaches in neuro-Behcet which considerably increases its prevalence).[13-16] The average overall prevalence of neuro-Behcet is estimated at 10%.[13-16]

Neurological involvement in Behçet's disease is divided in two major types: Parenchymal neuro-Behcet and non-parenchymal neuro-Behçet (also called extraparenchymal neuro-Behçet or cerebral angioBehçet: Cerebral venous thrombosis and cerebral arterial aneurysms).[13-16]

The concomitant occurrence (simultaneous parenchymal and non-parenchymal neuro-Behçet) is exceptional and not reported in the major series of neuro-Behçet.[13-16] Some authors estimated its prevalence at 5.8–6.5%.[17,18]

This mixed pattern of neuro-Behçet represents a real diagnostic challenge for clinicians[9] and can be a life threatening complication of the disease.[19]

The treatment of the mixed neuro-Behçet combines systemic steroids, immunosuppressants (cyclophosphamide or azathioprine), and effective anticoagulation. Acute forms usually respond favorably to this treatment.[17,18]

CONCLUSION

As rare as it is, the mixed clinical presentation of neuro-Behçet deserves to be known by healthcare professionals. Only an early diagnosis and appropriate and rapid management are the guarantees of a better prognosis. Medical imaging, and in particular MRI with angiographic sequences, has greatly facilitated the positive diagnosis of this exceptional complication of Behçet's disease.

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