Radiology Case Reports 20 (2025) 625–628

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Neuro-Behçet leading to coma: A case report ☆

Soumia Kriouile, MD, Kaoutar Imrani, MD, Soumya El Graini, MD, Nabil Moatassim Billah, PhD, Ittimade Nassar, PhD*

Central Radiology Department, Ibn Sina University Hospital Center, Mohamed V University of Rabat, Rabat, Morocco

ARTICLE INFO

Article history:
Received 2 October 2024
Revised 15 October 2024
Accepted 16 October 2024

Keywords:
Behçet disease
Neuro-Behçet disease
Coma
Neuroradiology
MR

ABSTRACT

Neuro-Behçet disease is often difficult to diagnose due to its complex and severe clinical presentation. This article reports the case of a 35-year-old female patient with a history of Behçet's disease, admitted for a deep coma. Brain MRI performed upon admission revealed extended lesions in the basal ganglia, thalami, and midbrain, along with leptomeningeal contrast enhancement in the temporal region, suggesting meningoencephalitis compatible with parenchymal involvement of neuro-Behçet's disease. Following a comprehensive negative evaluation for infectious and malignant causes, combined with the patient's medical history, a diagnosis of neuro-Behçet disease was established.

© 2024 The Authors. Published by Elsevier Inc. on behalf of University of Washington.
This is an open access article under the CC BY-NC-ND license
(http://creativecommons.org/licenses/by-nc-nd/4.0/)

Introduction

Behçet's disease is a systemic vasculitis of unknown etiology, predominantly characterized by cutaneous, mucosal, and ocular lesions. Various systemic manifestations may occur, including neurological involvement, which constitutes neuro-Behçet disease (NBD). It should be considered in any young patient with Behçet's disease who presents a neurological symptoms, ranging from mild headaches to deep coma [1,2].

Case presentation

A 35-year-old female with a history of recurrent oral ulcers, and panuveitis was admitted to the emergency department with an acute loss of consciousness. The patient received azathioprine and cyclosporine for 5 years ago for Behçet disease. After that, the patient had been out of sight for 1 year prior to admission to the hospital.

Two weeks leading to the admission, the patient presented a mild fever recorded at 38°c with a sore throat that was treated with antibiotic. While the sore throat resulted, the fever became higher at 39°c, and over the 3 last days, she developed a photophobia, right body weakness and difficulty in walking.

A neurological evaluation showed an unconscious patient with a Glasgow Coma Scale score of 8/15 (eye opening: 2; verbal response: 3; motor response: 3). She had a bilateral vertical nystagmus with myoclonus of the right foot. Deep tendon reflexes were brisk with a bilateral Babinski sign. The examination of the other systems was intact. Large paraclinical tests were normal (hemogram, C-reactive protein (CRP) test,

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

* Corresponding author.
E-mail address: soumia.kr97@gmail.com (S. Kriouile).

https://doi.org/10.1016/j.radcr.2024.10.102

1930-0433/© 2024 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/)

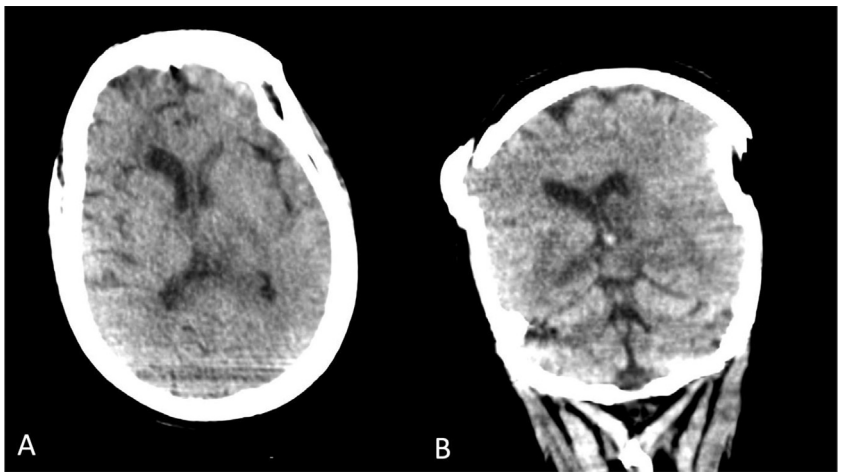

Fig. 1 – A 35-year-old female, with a history of Behçet's disease and a right body weakness, presented an acute loss of consciousness. Brain CT scan in axial view (A) and coronal view (B) at the level of the lateral ventricle horns showed a hypodense lesion centered in the left thalamus.

B12 and B9 seric levels, thyroid hormone, serum electrolytes, and renal and hepatic tests).

Computerized tomography (CT) of the brain was obtained which showed a hypodense lesion centered in the left thalamus, a contrast complement didn't show any pathological enhancement (Fig. 1).

Cerebral magnetic resonance imaging demonstrated bilateral and asymmetrical signal abnormality, more pronounced on the left side of the corpus callosum, internal capsules, left thalamus, meso-diencephalic junction, midbrain, and hippocampi, appearing T2 and fluid-attenuated inversion recovery (FLAIR) hyperintense and T1 hypointense. These lesions showed a heterogeneous enhancement after injection, and they were surrounded by a perilesional vasogenic edema causing a 5mm rightward midline shift. A leptomeningeal contrast enhancement at the right internal temporal region was also noted (Fig. 2).

MR angiography of the head and neck showed no arterial occlusion or stenosis, arteriovenous malformation, or aneurysm.

At this point, additional differential diagnoses were considered, in particularly an infectious or a neoplastic cause.

A lumbar puncture was done, and the cytochemical study of cerebrospinal fluid was normal, apart from high level of proteins. Other infectious workups included fungal (cryptococcus), parasitic (toxoplasmosis), bacterial (syphilis and tuberculosis) and viral (human immunodeficiency virus, hepatitis B, hepatitis C), studies, which were all negative. Given the concern for malignancy, a CT scan of the chest, abdomen, and pelvis was completed, but the results ruled out any disease process.

While the workup was ongoing, the patient received an intravenous methylprednisolone bolus (1 g/day for 5 days), she was also started on broad-spectrum antibiotics that were discontinued after the negative infectious workup. The treatment was then followed by prednisone 1 g/day for 4 weeks and intravenous cyclophosphamide (1 g/month). A 60 mg/day oral steroid tapering dosage was administered, with 8 weeks of gradual degression up to a minimum dose of 5 mg/day. Evolution was favorable, with the recovery of consciousness after 1 week of her admission. The patient had a mild residual right hemiparesis at the 3 months follow-up consultation.

Discussion

Behçet's disease (BD) is a multisystemic vasculitis of almost unknown origin. It affects both small and large blood vessels, involving veins and arteries, and is characterized by a nonspecific inflammatory process of the blood vessels [3].

Neurological involvement in Behçet's disease, known as neuro-Behçet's disease (NBD), is a significant cause of long-term morbidity and mortality, though it is rare. It is essential to recognize NBD and consider it in the differential diagnosis of inflammatory, infectious, and demyelinating diseases [4].

Two main phenotypes of NBD are described: the parenchymal and the nonparenchymal including dural sinus thrombosis, intracranial and extracranial aneurysm formation and arterial vasculitis [5]. Our patient presented with a typical parenchymal form of neuro-Behçet's disease, without associated thrombosis or aneurysm. Parenchymal involvement in neuro-Behçet's disease (NBD) occurs in about 80% of cases, mainly affecting the brainstem and basal ganglia. The clinical presentation of NBD is diverse, with symptoms varying based on the location and extent of brain involvement [6].

Our patient presented with a state of unresponsive unconsciousness. Consciousness is controlled by various structures of the extended reticular and thalamic activating system, which includes the thalamic nuclei and the reticular formation that extends throughout the brainstem. In our case, the parenchymal lesions were extensive, involving both structures, thus causing the coma. The patient also presented with right-sided hemiparesis prior to hospitalization, which can be explained by the involvement of the left thalamus [7].

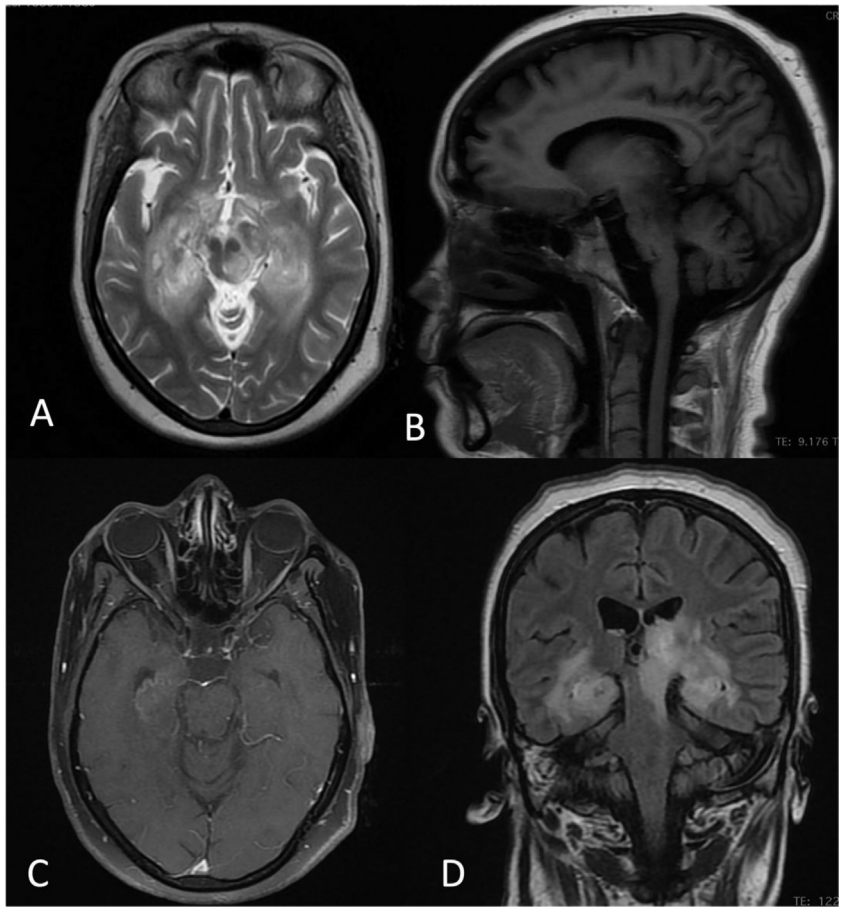

Fig. 2 – A 35-year-old female, with a history of Behçet's disease and a right body weakness, presented an acute loss of consciousness. Cerebral MRI showing signal abnormality. (A) T2-weighted image in axial view showing hypersignal of the corpus collosum and the midbrain. (B) T1-weighted image in sagittal view showing hyposignal of the thalami and the meso-diencephalic junction. (C) T1-weighted image with contrast in axial view showing a leptomeningeal enhancement at the right temporal region. (D) FLAIR sequence in coronal view showing bilateral and asymmetric hypersignal of the left thalamus, the left internal capsule, hippocampi, midbrain and meso-diencephalic junction surrounded by a vasogenic edema and a mild rightward midline shift: The cascade sign [9].

Regarding imaging modalities, MRI is the gold standard. In parenchymal NBD, MRI helps assess the nature, timing, location, and spread of lesions, distinguishing between acute, subacute, or chronic brain changes. Acute lesions are described to appear hypo- to iso-intense on T1-weighted images, hyper-intense on T2W and FLAIR images, and hyper-intense on diffusion-weighted images [8]. Lesions are frequently located in the mesodiencephalic junction (thalamus, midbrain, and internal capsule) appearing as the cascade sign in coronal views. This sign was nicely seen in our patient, in FLAIR sequence in coronal view. (Fig. 2, D) [9] Restricted ADC usually indicates vasogenic edema and vasculitis, while absent or reduced ADC suggests cytotoxic edema, commonly seen in subacute NBD [10]. The parenchymal distribution of lesions in NBD seems to support the hypothesis of small-vessel vasculitis. This lesion pattern may aid in distinguishing NBD from other types of vasculitis and inflammatory demyelinating diseases of the central nervous system [10].

In parenchymal NBD, cerebrospinal fluid (CSF) typically shows elevated protein levels, which was the case of our patient, frequent pleocytosis (lymphocytic or mixed), normal glucose levels, and absence of oligoclonal bands [11].

A nervous tissue biopsy is not required for diagnosis, but if performed, it may reveal vasculitis lesions and perivascular infiltration by lymphocytes and neutrophils. In later stages, axonal loss and gliosis may also be observed. In our patient, the biopsy was not indicated [12].

There are no established diagnostic criteria for NBD. The only available guidelines are the International Consensus Recommendation (ICR) criteria, which have been developed to aid in diagnosing the condition [10].

The ICR suggested 2 forms of clinical NBD: the definite and the probable form, detailed in Table 1.

Our patient met diagnostic criteria established by the International Study Group for Behçet's disease with a history of recurrent oral ulcers and panuveitis, in addition to that, she was admitted for a neurological syndrome, in particularly a

Table 1 – International consensus recommandation (ICR) criteria for NBD diagnosis [10].

Definite NBD meeting all of the following 3 criteria:

1. Satisfy the ISG[a] criteria for BD
2. Neurological syndrome recognized to be cause by BD and supported by relevant and characteristic abnormalities seen on either or both: Neuroimaging /CSF
3. No better explanation for the neurological findings

Probable NBD meeting one of the following 2 criteria in the absence of a better explanation for the neurological findings:

1. Neurological syndrome as in definite NBD, with systemic BD features but not satisfying the ISG criteria
2. A noncharacteristic neurological syndrome, occurring in the context of ISF criteria-supported BD

[a] ISG, International Study Group Criteria 1990 or any other accepted current of future criteria.

coma, justified by neuroimaging, and there were no other explanations for the neurological lesions found in the imaging, these criteria assist the diagnosis of a definite NBD [7].

Concerning the management of the disease, acute NBD is treated with high-dose intravenous methylprednisolone for 7-10 days, followed by a gradual tapering of oral doses over 3-6 months, based on relapse severity. This treatment has been effective, especially for brainstem lesions and the parenchymal form of NBD [13]. Long-term anti-inflammatory treatment has involved traditional immunosuppressive agents, including azathioprine, sulfasalazine, and other 5-aminosalicylic acid derivatives, as well as cyclosporine, to manage the ocular manifestations of BD [8]. Biological therapy, particularly monoclonal antibody treatments, has been extensively researched, with tumor necrosis factor (TNF) inhibitors like infliximab, etanercept, and adalimumab demonstrating beneficial effects [14]. In our case, following a high-dose steroid regimen, we initiated a prednisone taper along with cyclophosphamide therapy. For patients with cerebral veinous thrombosis, while the use of anticoagulation remains controversial, many experts still recommend it alongside immunosuppressive treatment [10].

Conclusion

Our case shows that NBD can manifest itself through a deep coma resulting in extensive brain steam lesions. It should be considered in cases of acute loss of consciousness especially in young patients who have a history of oral or genital ulcers.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

REFERENCES

[1] Hamid M, Cherradi S, Ait Berri M, Bourazza A. Neuro-Behçet's disease with coma. Cureus 2022;14:e31596.

[2] Saadoun D, W B. Manifestations neurologiques de la maladie de Behçet. Neurologie 2015; 17-169-A-10. doi:10.1016/S0246-0378(15)69055-8.

[3] Yurdakul S, Yazici H. Behçet's syndrome. Best Pract Res Clin Rheumatol 2008;22:793–809.

[4] Zouboulis CC, Keitel W. A historical review of early descriptions of adamantiades-Behçet's disease. J Investigat Dermatol 2002;119:201–5.

[5] Yazici Y, Yurdakul S, Yazici H. Behçet's syndrome. Curr Rheumatol Rep 2010;12:429–35.

[6] Serdaroğlu P. Behçet's disease and the nervous system. J Neurol 1998;245:197–205.

[7] Walter BL, Shaikh AG. Midbrain. In: Aminoff MJ, Daroff RB, editors. Encyclopedia of the neurological sciences. Oxford: Academic Press; 2014. p. 28–33. doi:10.1016/B978-0-12-385157-4.01161-1.

[8] Hatemi G, et al. Management of Behcet disease: a systematic literature review for the European League Against Rheumatism evidence-based recommendations for the management of Behcet disease. Ann Rheumat Dis 2009;68:1528–34.

[9] Cleaver J, et al. An important diagnostic clue for neuro-Behçet's disease: the 'cascade sign'. Rheumatology 2021;61:e130–1.

[10] Kalra S, et al. Diagnosis and management of Neuro-Behçet's disease: international consensus recommendations. J Neurol 2014;261:1662–76.

[11] Akman-Demir G, Serdaroglu P, Tasçi B. Clinical patterns of neurological involvement in Behçet's disease: evaluation of 200 patients. Brain 1999;122:2171–82.

[12] Hirohata S. Histopathology of central nervous system lesions in Behçet's disease. J Neurol Sci 2008;267:41–7.

[13] Siva A, et al. Behet's disease: diagnostic and prognostic aspects of neurological involvement. J Neurol 2001;248:95–103.

[14] Esatoglu SN, et al. Behçet’s syndrome: providing integrated care. JMDH 2017;10:309–19.