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Case Report

Deep Cerebral Vein Thrombosis Revealing Behçet's Disease: Case Report

Izi Zineb*, Laridi Aya, Abourak Chaimaa, Oubaddi Tlaite, Jaheddine Fadwa, Enouali Hassan and Elfenni Jamal

Radiology Department, Hôpital Militaire d'Instruction Mohamed V, Mohammed V University, Rabat, Morocco

***Corresponding Author:** Izi Zineb, Radiology Department, Hôpital Militaire d'Instruction Mohamed V, Mohammed V University, Rabat, Morocco.

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Abstract

Behçet's disease (BD) can lead to deep cerebral venous thrombosis (DCVT), a rare but serious neurological complication. We report a 28-year-old male presenting with headache, confusion, and vomiting. MRI was essential in confirming DCVT, revealing internal cerebral vein thrombosis. Clinical features supported the diagnosis of BD. Early imaging enabled prompt treatment with corticosteroids and anticoagulation, resulting in rapid improvement. This case underscores the vital role of MRI and MRV in diagnosing neuro-Behçet complications.

Keywords: *Behçet's Disease; Cerebral Venous Thrombosis; Neuro-Behçet Syndrome*

Abbreviations

BD: Behçet's Disease; DCVT: Deep Cerebral Vein Thrombosis; MRI: Magnetic Resonance Imaging; MRV: Magnetic Resonance Venography; ISG: International Study Group; CVT: Cerebral Venous Thrombosis; DWI: Diffusion-Weighted Imaging; FLAIR: Fluid-Attenuated Inversion Recovery; SWI: Susceptibility-Weighted Imaging; CT: Computed Tomography; CTV: CT Venography

Introduction

Behçet's disease (BD) is a rare, chronic inflammatory condition of unclear cause that results in inflammation of blood vessels throughout the body, causing symptoms such as mouth and genital ulcers, skin rashes, and eye inflammation. Neurological involvement is among the most serious complications of the disease. It can also lead to thrombosis due to immune system dysfunction. Cerebral venous thrombosis occurs in 10% to 30% of individuals with neurological symptoms, it can be superficial and/or deep [1]. The diagnosis of deep cerebral vein thrombosis (DCVT) in Behçet's disease is typically confirmed through advanced imaging methods like MRI and MR venography.

Case Report

A 28-year-old male with no notable medical history arrived at the emergency department after experiencing a severe headache, confusion, and vomiting for three days. Neurological evaluation identified papilledema and mild weakness on the right side. His vital signs were stable, and lab tests did not reveal any signs of infection. Due to the altered mental state, an urgent brain MRI was conducted.

Imaging revealed hyperintense lesions in the deep cerebral veins on T1-weighted sequences and Flair, with thrombus formation. MR venography (MRV) showed occlusion of the internal cerebral veins, consistent with deep cerebral vein thrombosis (DCVT) (Figure 1). Further clinical investigation revealed recurrent oral and genital ulcers and a positive pathergy test, fulfilling the International Study Group (ISG) criteria for Behçet's disease.

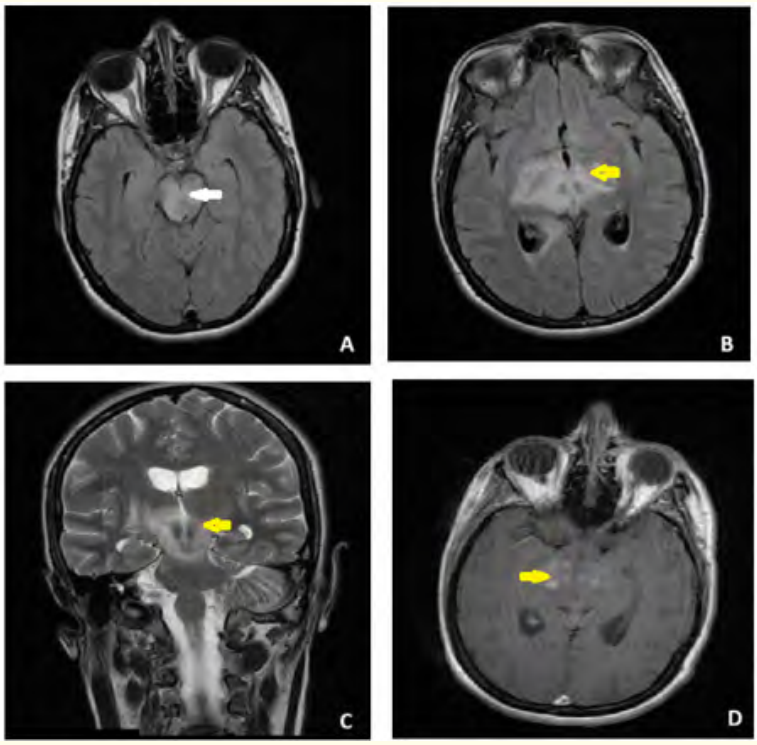

Figure 1: *Axial FLAIR sequences (A and B) and coronal T2-weighted (C) imaging demonstrate hyperintensity of the basal ganglia and midbrain (white and yellow arrows). Post-contrast axial T1-weighted imaging (D) shows enhancement, consistent with deep cerebral venous thrombosis.*

The patient was started on high-dose corticosteroids and anticoagulation therapy, resulting in significant neurological improvement within one week. Azathioprine was added to maintain long-term disease control. Follow-up MRV demonstrated partial recanalization of the affected veins.

Discussion

Behçet's disease (BD) is a chronic, relapsing, multisystem inflammatory disorder of unknown origin. Although it occurs worldwide, its prevalence is highest along the historic Silk Road [2].

This disease affects both sexes, with higher frequency and severity in men. The onset of the disease most often occurs between the ages of 25 and 30 [3].

Behçet's disease was first described by Hulusi Behçet in 1937, who identified a diagnostic triad of recurrent oral ulcers, genital ulcers, and uveitis [3]. In 1990, the International Study Group (ISG) for Behçet's Disease established diagnostic criteria, requiring recurrent oral aphthosis and at least two of the following: genital aphthosis, uveitis, pseudo-folliculitis, and a positive "pathergy test". The criteria established by the ISG remain the most widely used for diagnosis, although a key limitation is the lack of significant organ involvement, such as vascular, neurological, and gastrointestinal manifestations [4].

Behçet disease serves as a natural example of inflammation-triggered thrombosis in humans, with thrombosis arising from an abnormal immune-inflammatory response rather than traditional cardiovascular risk factors. Neutrophil hyperactivation and their mechanisms of damage are central, as they directly lead to endothelial dysfunction, platelet activation, and the development of thrombosis [4,5].

Neurological involvement occurs in 5% of patients with Behçet's disease (BD) and is classified into two main forms: parenchymal and vascular. Parenchymal neuro-BD causes inflammation in the brainstem, diencephalon, and basal ganglia, leading to symptoms like hemiparesis, bilateral pyramidal signs, behavioral changes, and headaches. In contrast, vascular neuro-BD involves cerebral veinous thrombosis, presenting primarily with headaches and papilledema. This form is the most common manifestation of vasculo-Behçet's disease and may be superficial and/or deep localization. It is also often associated with venous thrombosis in the lower extremities and generally has a more favorable prognosis compared to parenchymal neuro-BD [6-8].

In the literature, the superior sagittal sinus and transverse sinus are reported as the most and second most frequent sites of CVT in patients with BD. In contrast, thrombosis of the cerebral deep venous system is a less common occurrence [9,10].

The clinical manifestations of DCVT are nonspecific, and patients may exhibit a brief history of symptoms such as declining consciousness, confusion, aphasia, seizures, nausea, vomiting, coma, or even death. Distinguishing DCVT from other common causes of altered mental status, such as metabolic encephalopathy or meningoencephalitis, can be challenging based on clinical presentation alone [11].

Imaging plays a crucial role in diagnosing DCVT in Behçet's disease, with MRI and MR venography (MRV) being the most commonly used modalities. MRI is particularly sensitive in detecting the presence of venous thrombosis and associated parenchymal changes. In DCVT, MRI typically reveals hyperintense lesions on T1-weighted images due to the presence of thrombus and associated hemorrhage in the acute to subacute phase. T2-weighted and FLAIR sequences are also helpful in identifying venous infarcts and surrounding edema. Susceptibility-weighted imaging (SWI) is highly effective in detecting deoxygenated blood products in the thrombosed veins. Diffusion-weighted imaging (DWI) may show restricted diffusion in areas affected by venous infarction.

MR venography (MRV) is an essential tool for directly visualizing venous occlusions, making it a key diagnostic step after abnormal findings on MRI. MRV can confirm the lack of blood flow in the affected cerebral veins, providing detailed anatomical information about the location and extent of the thrombosis. This is crucial for determining the appropriate therapeutic approach [6,10].

Therefore, MRI and MRV are considered as the gold standard for diagnosing DCVT in BD, enabling the detection of both thrombotic changes and secondary complications such as venous infarcts. These modalities also help clinicians differentiate DCVT from other causes of venous thrombosis [11].

CT, though less sensitive than MRI, is sometimes used, especially when MRI is unavailable. It can show hyperdensity in thrombosed veins in the acute phase and reveal venous infarctions. CT venography (CTV) provides similar information to MRV, offering a visual assessment of venous occlusions, though it lacks the same level of sensitivity for detecting subtle thrombotic changes.

Conventional angiography is rarely used but can be helpful in complex cases.

The management of DCVT in BD typically involves a combination of high-dose corticosteroids, such as prednisolone, to address inflammation. In some cases, anticoagulation therapy with heparin followed by warfarin may be used, but this must be carefully monitored due to the risk of bleeding, particularly in patients with systemic aneurysms [12]. Immunosuppressive agents like azathioprine and colchicine are also considered to prevent recurrence of DCVT and manage the underlying inflammatory disease [13]. Acetazolamide can be used to treat intracranial hypertension when present.

The prognosis for DCVT in BD is generally favorable with early and appropriate treatment, though some patients may experience long-term sequelae such as cognitive impairments, visual disturbances, or persistent headaches. DCVT, particularly involving the internal cerebral veins, is associated with a worse prognosis compared to dural venous sinus thrombosis [14]. Recurrence of CVT is possible, requiring long-term follow-up to manage and prevent further complications. Early intervention is critical to prevent severe neurological damage, and treatment response tends to be good in the majority of cases, with the potential for recovery depending on the extent of venous occlusion and associated infarcts.

Conclusion

In cases of unexplained altered sensorium, early MRI of the brain is recommended. If MRI findings indicate DCVT, MRV should be performed to confirm the diagnosis. Clinical assessment also plays a crucial role in supporting the suspicion of Behçet's disease as the underlying cause of DCVT.

Conflict of Interest

Declare if any financial interest or any conflict of interest exists.

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