

Behcet's Disease (BD) Presenting as a Cerebral Venous Sinus Thrombosis (CVST)

S. Powell¹, B. Morris¹, J. Murray², E. O'Neill¹

1. Department of Ophthalmology, Mater Misericordiae University Hospital, Dublin.
2. Department of Radiology, Mater Misericordiae University Hospital, Dublin.

Abstract

Presentation

20 year old Caucasian male presented to eye casualty 4 weeks post initial diagnosis of bilateral acute anterior uveitis (AAU), with a three-week history of a progressively worsening headache associated with nausea and vomiting.

Diagnosis

Non-contrast Computed Topography of the head and Magnetic Resonance venogram revealed a cerebral venous sinus thrombosis (CVST). He had a long-standing history of intermittent oral ulceration, and was diagnosed with Neuro Behcet's Disease (NBD).

Treatment

The patient was commenced on a therapeutic dose of enoxaparin and prednisolone, and was discharged on enoxaparin, warfarin, tapering prednisolone and azathioprine.

Discussion/Conclusion

NBD is a rare, but serious manifestation of BD. BD is an important differential diagnosis in a young patient presenting with CVST or bilateral AAU.

Introduction

Behcets Disease (BD) is a multisystem, chronic, occlusive vasculitis of unknown aetiology that is relapsing in nature.¹ The classical triad of recurrent oral ulceration, recurrent genital ulceration and bilateral anterior uveitis was first described in 1937.² The diagnosis of BD remains a clinical one, and the gold standard International BD Study Group diagnostic criteria states that for a BD diagnosis to be made, a patient must have recurrent oral ulceration (at least 3 times in one 12 month period), plus two of the following symptoms: recurrent genital ulceration, eye lesions, skin lesions, or a positive pathergy test (read at 24-48h).³

Neurological BD (NBD) is a rare but serious complication of the disease occurring in 3%-9% of patients.^{1,4} NBD is associated with significant morbidity and mortality, primarily affecting the central nervous system (CNS). There are two primary forms of NBD: an inflammatory parenchymal pathology (involving the brainstem, spinal cord, thalamus and cranial nerves),¹ and a non-parenchymal/vascular form.⁵ Non-parenchymal BD causes cerebral venous sinus thrombosis (CVST), seen in 12-20% of BD patients with neurological disease.⁶

Case Report

A twenty-year old, previously healthy male was diagnosed with non-granulomatous bilateral acute anterior uveitis (AAU) after presenting to eye casualty with painful, red, photophobic eyes. Posterior segment examination was unremarkable. He was commenced on tapering topical steroids and cycloplegics. Baseline uveitis screening investigations were unremarkable, and the patient had a normal Chest X-Ray.

He represented to eye casualty four weeks later with a three week history of a progressively worsening headache associated with nausea and vomiting. He denied visual symptoms including diplopia, photophobia or decreased visual acuity. On examination, visual acuity was Snellen 6/6 in both eyes. The AAU had resolved and there was no retinal vasculitis, however both optic nerves appeared swollen with indistinct margins. The maculae and peripheral retinae were normal. He was hemodynamically stable and afebrile with no other neurological deficits. An urgent non-contrast Computed Tomography (CT) and Magnetic Resonance (MR) venogram confirmed the diagnosis of CVST (Figures 1 and 2). The patient had a history of recurrent oral ulceration, but denied other pertinent systemic symptoms such as genital ulceration or musculoskeletal symptoms. He had no history of deep vein thrombosis, and no known clotting disorders. A blood screen including full blood count, c-reactive protein, erythrocyte sedimentation rate, homocysteine levels, thrombophilia and coagulation screens including factor V Leiden, antiphospholipid antibodies, janus kinase 2 mutation autoimmune antibodies and human leukocyte antigen B*51 were all normal.

A diagnosis of CVST secondary to BD was made. He was commenced on a therapeutic dose of enoxaparin and prednisolone 40mg once daily (OD). On discharge he was commenced on azathioprine 100mg BD.

Differential diagnoses of CVST in a young person include stroke, systemic lupus erythematosus, multiple sclerosis, brainstem glioma and neurosarcoidosis.

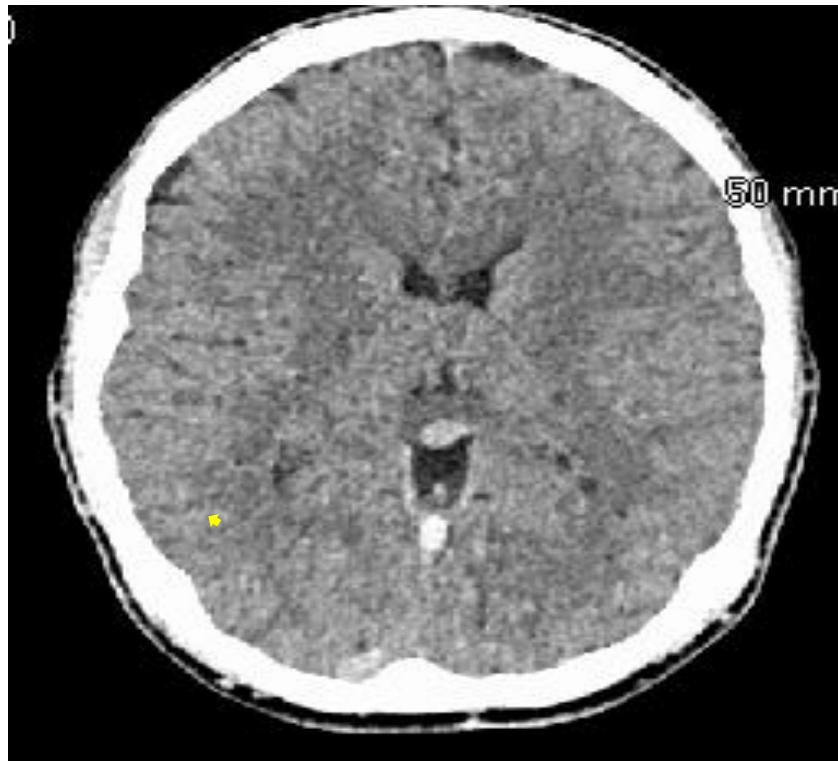


Figure 1. Non contrast CT brain revealing a hyperdense linear opacity in the straight sinus, concerning for CVST.

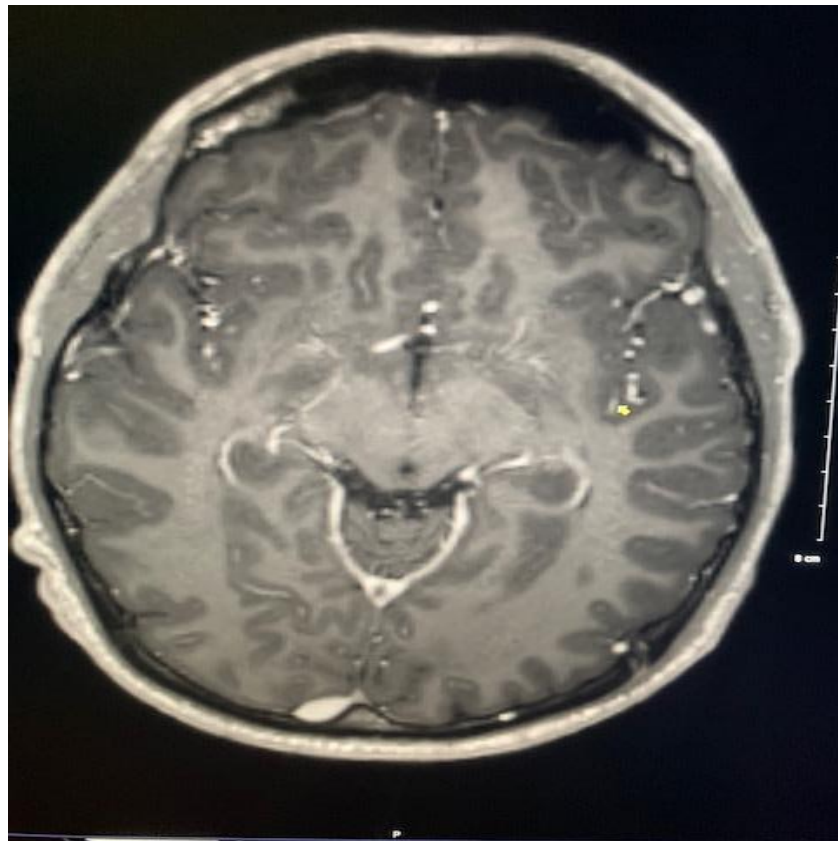


Figure 2. MR venogram (Gadolinium enhanced) demonstrating small focus low signal defect in straight sinus anteriorly in keeping with a CVST.

Discussion

Currently, CVST accounts for 18%-33.3% of presentations in non-parenchymal NBD, with prevalence higher in certain areas in the Middle East.⁷ NBD manifestations generally develop between 3-6 years after the initial onset of BD symptoms, however NBD can occur simultaneously to the onset of systemic symptoms.⁸ BD is a rare disease in the Caucasian Irish population and this presentation of CVST and NBD in a patient with undiagnosed BD is uncommon.

CVST secondary to NBD normally presents in a subacute clinical manner, whereby a severe, progressive headache over the course of a number of weeks is the most common clinical feature on presentation.^{1,6} CVST due to other causes is more likely to present as an acute onset of severe symptoms, often accompanied by focal neurological deficits including generalized seizures.⁹ Other common clinical signs associated with CVST secondary to NBD that were not present on examination of the patient in this case include both false localizing sixth and third nerve palsies.¹⁰

This case highlights the importance of accurate history taking, and the inclusion of BD in the differential diagnosis of bilateral AAU and CVST in a young male Caucasian patient.

Declaration of Conflicts of Interest:

There are no conflicts of interest to declare.

Corresponding Author:

Dr. Sarah Powell
Department of Ophthalmology,
Mater Misericordiae University Hospital,
Eccles Street,
Dublin 7.
E-Mail: powellsk@tcd.ie

References:

1. Al-Araji A, Kidd D. Neuro-Behçet's disease: epidemiology, clinical characteristics, and management. *The Lancet Neurology*. 2009;8(2):192-204.
2. Kalra S, Silman A, Akman-Demir G, Bohlega S, Borhani-Haghighi A, Constantinescu C et al. Diagnosis and management of Neuro-Behçet's disease: international consensus recommendations. *Journal of Neurology*. 2013;261(9):1662-1676.
3. INTERNATIONALSTUDYGROUPFORBEHC. Criteria for diagnosis of Behcet's disease. *The Lancet*. 1990;335(8697).

4. Kidd D. Neurological Complications of Behçet's Syndrome. *Current Neurology and Neuroscience Reports*. 2012;12(6):675-679.
5. Yazici H. Behçet's syndrome: An update. *Current Rheumatology Reports*. 2003;5(3):195-199.
6. Siva A, Kantarci O, Saip S, Altintas A, Hamuryudan V, Islak C et al. Behçet's disease: diagnostic and prognostic aspects of neurological involvement. *Journal of Neurology*. 2001;248(2):95-103.
7. Masheter H. Behcet's Syndrome. *BMJ*. 1965;1(5434):588-589.
8. Kalra S, Silman A, Akman-Demir G, Bohlega S, Borhani-Haghighi A, Constantinescu C et al. Diagnosis and management of Neuro-Behçet's disease: international consensus recommendations. *Journal of Neurology*. 2013;261(9):1662-1676.
9. Sorgun M. Cerebral Venous Thrombosis in Behçet's Disease Patients Compared to Other Causes of Cerebral Venous Thrombosis: a Retrospective Study. *Archives of Rheumatology*. 2016;31(3):248-253.
10. Yesilot N, Bahar S, Yilmazer S, Mutlu M, Kurtuncu M, Tuncay R et al. Cerebral venous thrombosis in Behçet's disease compared to those associated with other etiologies. *Journal of Neurology*. 2009;256(7):1134-1142.