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AN UNCOMMON AND ELUSIVE CAUSE OF CEREBRAL VENOUS THROMBOSIS

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ABSTRACT

We present a case of of young 27year male who was admitted with history of headache and vomiting for one week. He was diagnosed as having dural sinus thrombosis of superior sagittal, right lateral, right sigmoid sinus. There was previous history of uveitis 6 months prior to it. All thrombophilia workup was negative except homocysteine levels were moderately high. He was also found to be having recurring genital and oral ulcers. Pathergy test was negative. His HLA B 51/5 testing came back positive. Final diagnosis of Bechet's disease was made and responded well to long term steroids and Colchicine. A multisystem vasculitis like Bechet's disease should always be thought in patients with venous thrombosis with negative thrombophilia screening and recurrent oral and genital ulcers.

KEY WORDS:

Behcet's disease, Cerebral venous thrombosis, Dural sinus thrombosis

INTRODUCTION:

Behçet disease (BD) is a rare vasculitic disorder that is characterized by a triple-symptom complex of recurrent oral aphthous ulcers, genital ulcers, and uveitis.1Neurologic manifestations (neuro-BD) are relatively rare2. The frequency of neurologic manifestations varies, ranging from 5% to 30% of patients. CVT represents approximately 30% of all CNS lesions of BD2.

Case report

A 27 years old male was admitted with history of headache for last one week which was gradual onset, severe and generalised. Headache was associated with multiple episodes of vomiting. There was vague history of low grade fever on and off for last 20 days. No history of fits or loss of consciousness. There was previous history of visual impairment and redness left eye about a year ago and was diagnosed as having uveitis and macular edema and treated with topical and intraocular steroids. There was history of testicular swelling couple of months ago which resolved with anti-inflammatory and antibiotics. There was history of recurrent oral and genital ulcers and multiple OPD visits with them.

At presentation his GCS was 15/15. He had a pulse of 82/minute, regular, temperature of 99F.There were no signs were meningeal irritation. Motor and sensory system examination was normal. His initial CT scan brain was normal. Blood cultures were negative. His ESR was 70mm and CRP was 17. CSF R/E showed 15 cells, out of which 60% were neutrophils. CSF proteins were 28mg/dl. CSF glucose was 55mg/dl. He was started on Injection ceftriaxone 2gmIV BD along with vancomycin. However, he did not improve, headache did not settle. On 7th post admission day he developed diplopia right lateral gaze and severe headache with vomiting. His MRI brain showed cerebral venous thrombosis involving superior sagittal, right lateral, right sigmoid and confluence of sinuses. There was partial thrombosis of left lateral sinus as well.

He was started on LMWH along with antibiotics were switched to meropenum in meningitic dose. Meanwhile he was also worked up for Brucella, HIV, syphilis. HIV serology was negative. Brucella antibodies were

negative. VDRL/RPR serology was also negative. During hospital he again developed oral ulcers along with scrotal ulcers with cervical and inguinal lymphadenopathy. Lymph node biopsy was not possible as size of lymph nodes was very small.

His thrombophilia screening also came back negative except serum homocysteine levels which were moderately high (22.8, desirable< 15umol/L). His vasculitis screen including ANA, Anti DsDNA, anticardiolipin antibodies and anti ENA antibodies were negative. Pathergy test on him was negative. After six months of anticoagulation MRV was repeated which revealed partial recanalization of right transverse and sigmoid sinus, rest of the MRV findings were unchanged.

Few months later he again presented with high grade intermittent fever, not responding to antibiotics. Blood cultures were negative. CRP was 82mg/L, ESR was 110mm . All workup for fever was repeated but no source was found. Patient was put on steroids and fever settled

His HLA B 51/5 testing was also done which came back positive.

Our patient satisfied the International Study Group criteria for the diagnosis of Behcet's disease (table 1)3. He was also started on colchicine thinking of Behcet disease.

TABLE 1: International Study Group criteria for the diagnosis of Behcet's disease

Recurrent oral ulceration
Minor aphthous, major aphthous or herpetiform ulceration
observed by physician or patient, which recurred at least 3 times
in a 12-month period
And two of the following
Recurrent genital ulceration
Aphthous ulceration or scarring, observed by physician or patient
Eye lesions
Anterior uveitis, posterior uveitis, or cells in vitreous on slit lamp
examination, or retinal vasculitis observed by ophthalmologist
Skin lesions
Erythema nodosum observed by physician or patient, pseudofolliculitis
or papulopustular lesions, or acneiform nodules observed by the
physician in post-adolescent patients not on corticosteroid treatment
Positive pathergy test
Read by physician at 24–48 hours

Discussion

Behçet's disease is a multisystem inflammatory condition most commonly seen in those of Mediterranean and Eastern origin4. It is observed commonly among populations living along the historic Silk Road5. The exact cause is unknown. However, combination of genetic and environmental factors is likely to play a role5

Neurologic manifestations (neuro-BD) are relatively rare, but they must be thoroughly investigated due to their severe prognosis2

Vascular involvement is not limited to any vessel size, affects veins more than arteries and, when present, can be associated with constitutional symptoms. Patients might develop venous thrombosis affecting the lower limbs, vena cava or hepatic veins. Occasionally, haemoptysis as a result of pulmonary artery aneurysms occurs, and is associated with worse outcomes4.

Behçet disease is most common in persons aged 20-40 years. The mean age at onset is 25-30 years. Cases that develop before age 25 years are more likely to involve eye disease and active clinical disease6.

The characteristic ocular feature is relapsing uveitis as anterior, posterior, or panuveitis, and retinal vasculitis. Venous sinus thrombosis is the most frequent vascular manifestation in nonparenchymal disease followed by cortical cerebral veins thrombosis6.

Orchitis and epididymitis can also occur in patients with BD6

Saadoun et al found that cerebral venous thrombosis (CVT) was present in 7.8% of a large cohort of patients with Behçet disease. The main complication of CVT was severe visual loss due to optic atrophy. Papilledema and concurrent prothrombotic risk factors were independently associated with the occurrence of sequelae; peripheral venous thrombosis and concurrent prothrombotic risk factors were associated with relapse of thrombosis. Anticoagulant therapy proved safe and effective in up to 90% of patients2 However, there are controversial recommendations about anticoagulation. According to EULAR recommenations, beneficial role of anticoagulation is unclear, and can lead to adverse events in patients with coexistent pulmonary aneurysms7. We however treated our patients with anticoagulation for one year and he tolerated it well with no complication.

Hyperhomocysteinemia was found in up to 12% of patients2. Hyperhomocysteinenmia was found in our patient as concomitant prothrombotic risk factor and was appropriately treated.

Venous thrombosis in Behçet's disease is caused by phlebitis and not thrombophilia: hence the thrombus remains attached to the inflamed vessel wall and tends not to metastasise. Venous thrombosis responds well to treatment with immunosuppression8.

The first-line treatment for mucocutaneous manifestation of Behcet's Disease is colchicine (1 mg/day)5. Immunosuppressive therapy may cause overwhelming infections and patients should be counselled about this before the initiation of treatment

This case emphasizes the importance of identifying the cause of unprovoked venous thromboses and that clinicians should consider Behçet's disease in appropriate ethnic groups, particularly in patients with a history of recurrent oral or genital ulceration. Increasing

awareness among physicians can increase the diagnosis of Behcet's Disease and reduce morbidity.

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Farheen Niazi; concept, data collection, data analysis, manuscript writing, manuscript review
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