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Case Report Open Access

Cerebral Venous Thrombosis as First Manifestation of Behçet's Disease

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Abstract

Behçet's disease is a chronic inflammatory multisystem disorder which can involve the central nervous system. Cerebral venous thrombosis is one of its major neurological manifestations. We report cerebral venous thrombosis as the initial presentation of Behçet's disease in a 26-year-old female. Our patient presented with a two week history of severe headache and vomiting. Fundoscopic examination disclosed a bilateral papilledema. She developed VI cranial nerve palsy in the right eye 3 days later. Magnetic Resonance Venography of the brain showed left transverse sinus venous thrombosis. A diagnosis of BD was made based on clinical criteria. Occurrence of CVT as a first presentation is quite exceptional in BD.

Keywords: Behcet disease; Venous sinus thrombosis; Headache; Transverse sinus thrombosis

Introduction

BD is a chronic, multisystemic, relapsing inflammatory disorder with an unknown cause which affects eyes, skin and mucosa, joints, vascular system (mainly the veins), lungs, gastrointestinal tract and nervous system. The predominant histopathological feature of BD is a vasculitis that characteristically involves veins and arteries of distinct sizes. Neurologic symptoms are the presenting manifestation of BD in 5–6% of the cases. Occurrence of CVT as a first presentation is quite exceptional in BD [1,2].

Case Report

A 26 year-old female patient was admitted to Department of Neurology, Gazi University, for headache and vomiting. She had a constant headache for 2 weeks. In the beginning, daily squeezing band like headache and feeling of pressure on the head lasted several hours and it was improving a little with analgesics. Then it became generalized and dull. The headache was present even when she was sleeping. During the past 2 weeks, she had several attacks with vomiting and the headache became severe and longer in duration. The last week she developed sixth cranial nerve palsy in the right eye and large painful oral-genital ulcers that she never had before. Physical examination was normal. Neurological examination revealed bilateral papilledema and right sixth cranial nerve palsy. Magnetic Resonance Imaging (MRI) of the brain was normal (Figure 1).

Magnetic resonance venography (MRV) of the brain showed left transvers sinus venous thrombosis (Figure 2).

Initial laboratory tests haemoglobin, haematocrit, white blood cell (WBC), platelets, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), rheumatoid factor (RF) and routine biochemical laboratory studies were normal. Serology for viral hepatitis B and C was negative.

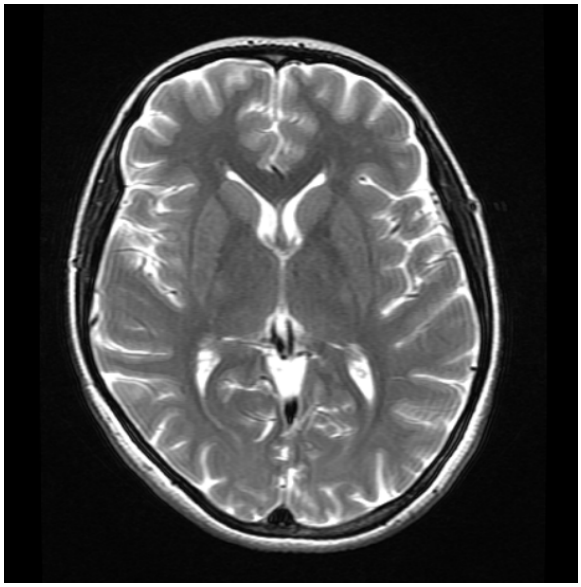

Figure 1: T2 weighted axial sequences of brain MRI was normal.

The patient had undergone detailed medical interviews and routine physical examination by ophthalmologist and rheumatologist. Ophthalmologic examination disclosed evidence of sequelae uveitis and bilateral papilledema in both eyes. Thrombophilia screening was undertaken and the patient was evaluated for prothrombotic disorders. The values of PT, aPTT, lipid profiles, haemoglobin electrophoresis, protein-C, anti-thrombin III were normal. Mutations of factor V Leiden, prothrombin 20210-A, MTHFR and ANA, anti-dsDNA, lupus anticoagulant, ANCA, anti-cardiolipin antibodies were negative. Protein S: 52 was low. Levels of Ig G, A, M, D and E measured and were normal. Also she had HLA-B51 positivity. Pathergy skin

hypersensitivity test was positive. Echocardiography was normal. Detailed past medical history revealed that she had small aphthous ulcers on her tongue and palate once in two or three months for a few years but she had never been consulted to a physician with this complaint. In the light of this information, the patient was diagnosed with Behçet's disease according to the International Study Group guidelines [3,4].

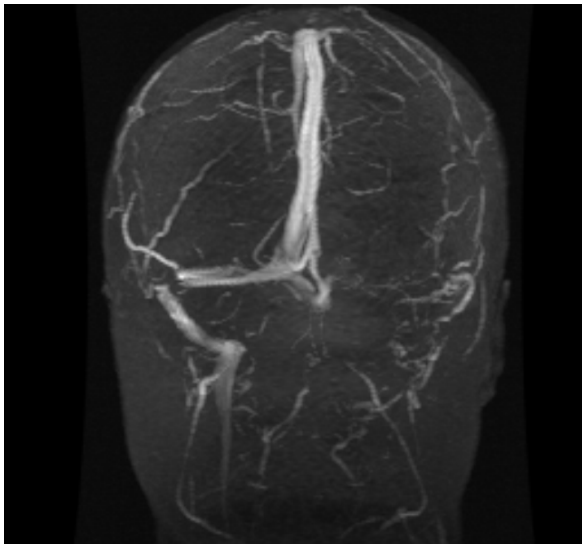

Figure 2: Time-of-flight (TOF) MR venography showed left transverse sinus thrombosis.

On the 7th day of her hospitalization in the neurology inpatient clinic she was started on IV 1 g/day methylprednisolone and after 5 days this was changed to oral prednisolone 1 mg/kg/day. The anticoagulant treatment was then added with oral azathioprine at 2 g/kg/day. Our patient responded well to the treatment with significant clinical improvement.

Discussion

BD is multisystem disease characterized by recurrent oral and genital ulcers, relapsing uveitis and mucocutaneus, articular, urogenital, intestinal, vascular and neurological manifestations. The CNS involvement in BD is named as neuro-Behcet's disease (NBD) and can be categorized in two groups. The first group is parenchymal involvement due to inflammation and the second group is non-parenchymal involvement due to vascular complications. MRI shows high signal intensity particularly in brainstem and capsula interna in T2-weighted images. Non-parenchymal involvement is described as large artery occlusion, thrombosis of large venous sinuses, aneurysm, and haemorrhage [5,6]. In the literature the prevalence of NBD in BD is known to be approximately 5% in large series and CVT occurs in 10-20% of patients with NBD [6-8]. In our case, there was not any signal change of white and grey matter in cranial MRI, however, MRV of the brain showed left transverse sinus thrombosis. For this reason, we determined the case as non-parenchymal NBD. Although our patient has a history of oral aphthae for several years before the main clinical picture of disease, they were indistinguishable from recurrent oral aphthous ulcers. When she presented with CVT, major oral and genital ulcers showed exacerbation. The presentation of full-blown picture enables us to diagnose.

There are few reports about first presentation of BD with CVT [9-15]. Cerebral vein and dural sinus thrombosis due to BD appear to be rare, but their identification has increased in recent years, due to the increased use of MRI, and MRV [16,17]. MRV can serve as a useful diagnostic study in situations where confirmation or exclusion of sinus thrombosis is required.

Evidence based data related with the treatment of neurological complications in BD are insufficient. Corticosteroids, anticoagulant therapy, other immunosuppressant, and colchicine are tried in various studies. Corticosteroids is often helpful, but usually the other immunosuppressant drugs, such as, azathioprine, methotrexate, cyclosporine A, and cyclophosphamide are necessary. Because of the vasculitic process, systemic corticosteroids combined with azathioprine for long-term immunosuppression is the most recommended therapy when there is central nervous system involvement in BD. Recently, interferon-α-2a and infliximab have been used with success even in patients seemingly resistant to other immunosuppressant. Treatment of cerebral vein thrombosis in BD is not standardized [2,18].

This case report emphasizes the importance of headache related to CVT followed by major oral and genital ulcers as first and unusual manifestation of BD. Furthermore, it can increase awareness among physicians of the neurological manifestations of this disease.

Conclusion

CVT can have multiple presentations and occasionally may be limited to a chief complaint of headache. We emphasize, therefore, the systematic application of MRV in patients with unclear persistent headaches. Lastly, this case report reinforces the importance of initial presentation of Behcet's disease as a cause of cerebral venous thrombosis.

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