Case Report / Olgu Sunumu

Neuro-Behçet Disease Presenting with Sinus Venous Thrombosis: Case Report

Sinus Ven Trombozu ile Prezente Olan Nöro-Behçet Hastalığı: Olgu Sunumu

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Abstract

Behçet's disease (BD) is a rare, chronic, autoimmune, autoinflammatory disorder of unknown origin. Mucocutaneous lesions and panuveitis constitute the hallmark of Behçet's. The disease is mostly seen at the third decade and disease course is dramatically more severe for males than females. Vascular involvement can occur in up to 40% of the cases. Although veins are mostly involved, all sizes and types of vessels can be affected. The most common clinical presentation of disease is lower extremity vein thrombosis. Vena cava thrombosis, pulmonary artery aneurysms, Budd-Chiari syndrome, peripheral artery aneurysms, dural sinus thrombosis and abdominal aorta aneurysms are also frequent at the disease's course. Prevalence of cerebral venous thrombosis is 8%. In this article, we presented a 28-year-old man with sinus venous thrombosis due to BD.

Keywords
Behçet disease, sinus venous thrombosis, headache

Öz

Behçet hastalığı (BH) etiyolojisi bilinmeyen, kronik, relapslarla seyreden, multisistem tutulumu yapabilen enflamatuvar bir hastalıktır. Tekrarlayıcı deri mukoza lezyonları ve panüveit hastalığın ana belirtileridir. Başlangıç yaşı genellikle 3. dekaddır ve erkekler kadınlara göre daha ciddi etkilenir. Vasküler tutulum olguların %40'ında görülür. Bütün çap ve tipteki damarlar etkilenmekle birlikte venleri arterlere göre daha sık etkiler. Alt ekstremite ven trombozu hastalığın en sık klinik bulgusu olup bunu vena kava trombozu, pulmoner arter anevrizması, Budd-Chiari sendromu, periferik arter anevrizması, dural sinüs trombozu ve abdominal aorta anevrizması izler. Serebral venöz trombozu %8 oranında görülür. Biz bu yazıda BH'ye bağlı sinus ven trombozu gelişen 28 yaşındaki erkek olguyu sunduk.

Anahtar Kelimeler
Behçet hastalığı, sinus ven tromboz, baş ağrısı

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Introduction

Behçet's disease (BD) is a chronic inflammatory disease that occurs with relapses and can cause multisystem involvement, and its etiology is not known (1). The most important histopathological attribute is vasculitis, which includes arteries and veins with different diameters. Various neurological findings can be observed in BD (2). The rate of neurological symptoms is 5-6% in affected patients (3). Prevalence of cerebral venous thrombosis is 8% (4) and it consists of 18% neuro-

Behçet's disease (5). In this report, a patient with BD and sinus venous thrombosis is presented.

Case Report

A 28-year-old male patient referred to hospital due to severe headaches that did not disappear after use of analgesics. In his background, BD was diagnosed one year ago. The temperature of patients was 36.5 °C, arterial blood pressure was 110/80 mmHg and heart rate was 80 beats/min. He had oral aphthous and genital ulcers. In his neurological examination, he did not have any pathology other than papilledema and hypoactive deep tendon reflexes. Filling defects were observed in the sinus rectus, both transverse and sigmoid sinuses and jugular vein tracing in his brain magnetic rezonans imaging (MRI) venography, and he was diagnosed with sinus vein thrombosis (Figure 1a, 1b). The hemogram, biochemistry, thyroid function tests, vitamin B12, protein C, protein S, antithrombin 3, and active protein C resistance were normal in his laboratory analyses. The results of factor 5 Leiden mutation, MTHFR and PAI-1 mutations and tests for etiology of vasculitis were negative. There were no abnormal values other than an elevated sedimentation value, which was 54 mm/hour (0-20), elevated C-reactive protein value, which was 23.4 mg/dL (0-0.8), elevated homocysteine value, which was 15.9 µmol/L (0-12), and decreased folic acid value, which was 3.67 ng/mL (4.6-18.7). Lumbar puncture was performed since the patient had papilledema. The opening pressure of cerebrospinal fluid (CSF) was 380 mmH_2O and closing pressure was 180 mmH_2O. The appearance of CSF was clear. A total of 10/mm^3 lymphocytes was observed. CSF glucose value was 75 mg/dL, concurrent blood glucose value was 97 mg/dL and the protein value was 30.2 mg/dL (15-45). No uveitis was observed in the patient's eye examination. He had oral aphthous and genital ulcers in his dermatological examination. There was no erythema nodosum. The results of the pathergy test were positive. The patient was diagnosed with sinus venous thrombosis related with BD according to the clinical, radiological and laboratory findings. A thoracic tomography angiography was performed for pulmonary aneurysm and the results were normal. The results of the bilateral lower extremity venous Doppler and abdominopelvic ultrasonography, which were performed to exclude thrombosis and aneurysm, were normal. 1 mg/kg/day oral corticosteroid, 25 mg/day azathioprine, warfarin tailored to international normalized ratio target, 3 tablets/day acetazolamide and 1 tablet/day potassium were started for the patient. It was planned to gradually decrease the

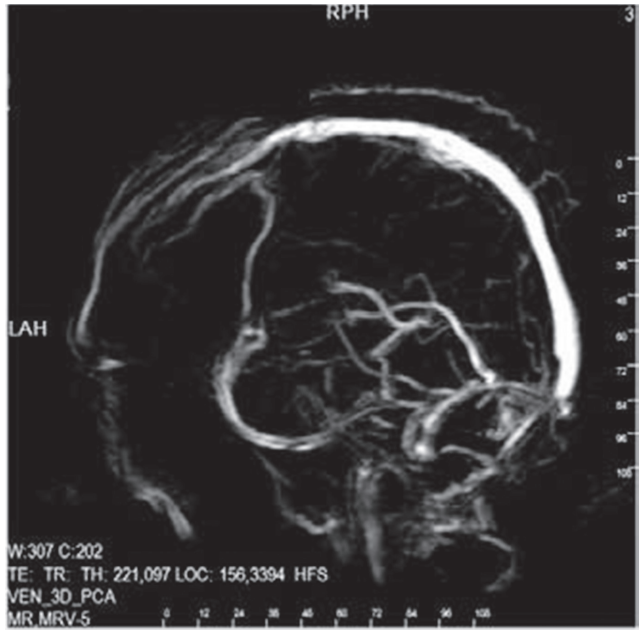

Figure 1a. Brain magnetic rezonans imaging venography: Filling defect in the sinus rectus, transverse and sigmoid sinus and jugular vein

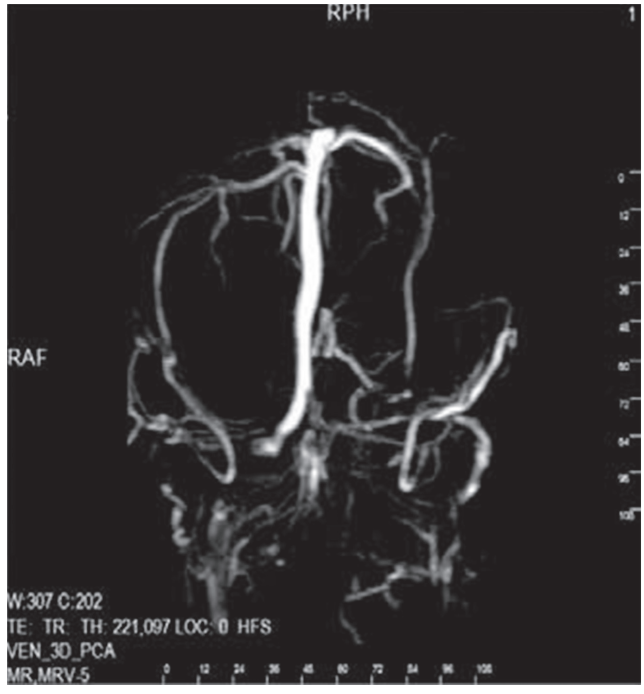

Figure 1b. Filling defects in both transverse and sigmoid sinuses and bilateral jugular vein tracing

steroid treatment by 4 mg weekly and discontinue it after one month, and increase the dose of azathioprine treatment to 2 mg/kg/day according to the follow-up of weekly hemogram and liver function tests. It was observed that the patient's edema decreased and his headache clinically disappeared in the check-up performed after one month. The patient's treatment is currently ongoing.

Discussion

Neurological involvement in BD occurs on average five years after the systemic findings are observed; it is observed 3-4 times higher in men than in women and the most common clinical presentation is sinus venous thrombosis at central nervous system (CNS) involvement; peripheral nerve or muscle involvement is very rare. It is possible to examine the CNS involvement in BD in two main groups: parenchymal CNS involvement (80%) and non-parenchymal CNS involvement (20%) (6).

The most typical parenchymal lesions seen at brainstem. Spinal cord involvement and hemispheric involvement are rare. Pyramidal findings, hemiparesis, behavioral and cognitive changes, sphincter disorders and/or impotence are the main clinical findings. Psychiatric problems can occur in some patients. Non-parenchymal involvement is observed with the findings of an increase in intracranial pressure that includes severe headache, papilledema and oculomotor nerve palsy. Dural nerve thrombosis has a good prognosis (7). It affects men at a higher rate than women and it is observed at an early age. Clinical course is various in many cases but chronic variant is rarely seen. Intracranial hypertension, focal neurological signs, seizure and/or impaired consciousness can occur in patients (8). Despite the fact that neurological involvement usually occurs years after systemic involvement, a case of a patient who had oral and genital ulcers one week after diagnosis of sinus vein thrombosis and who was diagnosed with BD was presented (9).

The most frequently observed incident is superior sagittal sinus thrombosis, and it is followed by transverse sinus, deep cerebral veins and cavernous sinuses (10). Loss of flow phenomena can be observed in the occluded dural sinus in the brain MRI, and other than this, the MRI is usually normal. MRI venography or cerebral angiography is helpful for the diagnosis. CSF is almost always normal except for the elevated pressure (6).

Steroids and anticoagulant medication can be used for the treatment of sinus thrombosis associated with BD. The most frequently used immunosuppressive drugs are azathioprine and methotrexate (11).

Our patient admitted to our clinic due to the complaint of a headache. The patient had papilledema in his examination and sinus vein thrombosis was observed in his MRI venography. An etiology of sinus vein thrombosis was investigated. The etiology was associated with BD as the patient had BD in his background. Since pulmonary artery aneurysm can be observed in BD, pulmonary artery aneurysm was eliminated after thoracic tomography angiography and anticoagulant medication was administered to the patient. Steroid treatment was started and azathioprine was started in terms of immunosuppressive therapy. Clinical improvement was observed in the patient. As a result, supraventricular tachycardia should be considered in patients with a headache that has started recently or that has changed character, radiological investigation should be performed quickly and BD must be investigated as the etiology, as BD is commonly observed in our country.

Ethics

Informed Consent: It was taken.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: H.B., O.D., Concept: O.D., H.B., Design: H.B., H.D.K., Data Collection or Processing: G.V., Ş.T., Analysis or Interpretation: Ş.T., G.V., Literature Search: H.D.K., H.B., Writing: H.B., O.D.

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