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Case report
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A case of Behcet's disease with upper extremity thrombosis

Üst Ekstremite Trombozlu Behçet Hastalığı Olgusu

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Abstract

Behcet's disease is a multi-system vasculitis in which recurrent oral and genital ulcers, uveitis as well as musculoskeletal, neurologic and gastrointestinal system involvement are observed. In previous studies, vascular involvement rate ranges between 1-38%. Most common vascular complication is superficial vein thrombosis of the extremities, followed by deep vein thrombosis. In this study, we will present a case of Behcet's disease with upper extremity thrombosis.

Keywords: Behcet's disease, Upper extremity, Superficial vein thrombosis

Öz

Behçet hastalığı tekrarlayan oral, genital ülserler, göz bulgularının yanısıra kas-iskelet, nörolojik ve gastrointestinal sistem (GİS) tutulumları ile seyreden sistemik bir vaskülittir. BH'da vasküler tutulum oranı serilere göre %1-38 arasında değişmektedir. En sık görülen vasküler komplikasyon ekstremitelerin yüzeyel ven trombozu iken bunu derin ven trombozu (DVT) izler. Bu yazıda, üst ekstremite trombozu olan bir Behçet hastalığı olgusundan bahsedilecektir.

Anahtar kelimeler: Behçet hastalığı, Üst ekstremite, Yüzeyel ven trombozu

Introduction

Behcet's Disease (BD) was first defined by the Turkish dermatologist Hulusi Behcet in patients with recurrent oral and genital ulcers and hypopyon iridocyclitis [1]. Other studies showed articular, pulmonary, gastrointestinal, urogenital, cardiac, vascular and neurologic system involvement accompanied these three symptoms [2]. The prevalence of BD in Turkey is 20-421/100,000 [3]. Symptoms, severity and clinical course of the disease differ among patients. The prevalence and severity of the disease are higher in men than in women [4]. The rate of vascular involvement in BD is 1-38% in some studies [5]. The most commonly seen vascular complication is superficial vein thrombosis followed by deep vein thrombosis of the extremities [6]. Superficial or deep vein thromboses of the lower extremities are more commonly seen than upper extremity thrombosis [7].

In this case report we present the case of a patient who admitted to the Department of Emergency Medicine at Umraniye Training and Research Hospital with a swollen arm and was diagnosed with upper extremity superficial vein thrombosis. The patient was diagnosed with BD two years ago and hadn't been taking his medication regularly.

Case presentation

A 32-year-old male patient who had been followed for two years for BD was admitted to the Department of Emergency Medicine at Umraniye Training and Research Hospital with a swollen and painful arm. The patient had a history of one time pulmonary emboli and two times orchitis. He was admitted to an outpatient clinic two years ago due to his recurrent oral apthous ulcers and was diagnosed with BD with a positive pathergy test. He had no history of genital ulcers and uveitis. He did not take his medicine regularly because he thought the treatment didn't help him. He is currently on day 100 mg acetylsalicylic acid treatment. There was no significant family medical history.

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The patient had been suffering from right leg pain and admitted to the Emergency Department four days ago. He had a normal physical examination. Deep vein thrombosis of the right leg was suspected. Complete blood count, ALT, AST, creatinine, serum glucose, Na, K, D-dimer levels and color doppler ultrasonography result were normal. He was referred to an outpatient clinic but not admitted.

In his left antecubital fossa where venous puncture was made four days ago, there was a seven cm diameter, edematous, painful, warm and hyperemic lesion. Five cm diameter difference was detected between left and right elbows of him. On his lower extremities multiple erythema nodosum lesions were detected. There was no diameter difference between his left and right legs. A lot of papulopustular lesions with residual scars were seen on his back. He denied oral or genital ulcers and uveitis.

Left upper extremity color doppler ultrasonography was performed with the preliminary diagnosis of venous thrombophlebitis. The ultrasound was reported as focal inflammation around soft tissue of the left elbow and thrombosis in collateral veins in the same region, other main branches of deep and superficial veins were normal. The patient was referred to Department of Cardiothoracic and Vascular Surgery. Subcutaneous bemiparin sodium 7500 IU/day and oral amoxicillin/clavulanic acid (1000 mg twice a day) treatments was given to the patient.

Four days later the patient was admitted to the Family Medicine outpatient clinic because of increased pain in his legs. In his physical examination, it was noticed that edema and hyperemia of the left antecubital fossa lessened (figure 1a) and the diameter difference in the upper extremities decreased two cm.

The number of erythema nodosum lesions on his legs increased and their color became darker (Figure 1 b-d). There was no diameter difference between lower extremities of him. Bilateral lower extremity doppler ultrasonography was performed and reported as "minimal intimal irregularities and thickening accompanied by multifocal calcific deposits in all the examined arteries, plaque and thrombus formation were not observed in venous system".

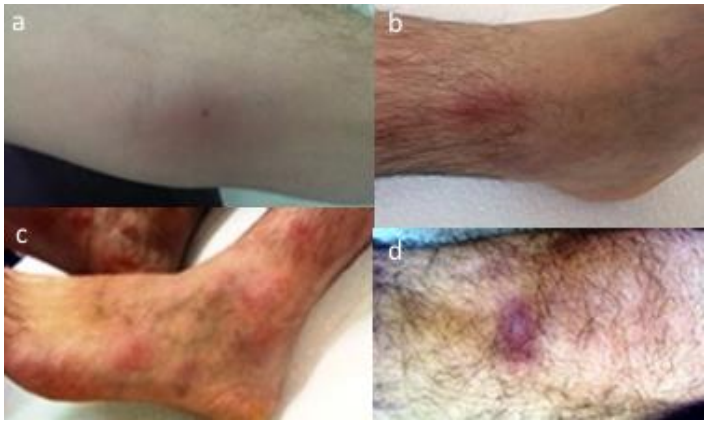

Figure 1: a: Antecubital fossa after four days treatment, b-d: The number of erythema nodosum lesions on his legs increased and their color became darker

Discussion

We report the case of a patient diagnosed with upper extremity venous thrombosis, a rare complication of BD. The patient had been followed for BD for two years, and hadn't been taking his medication regularly.

BD is a systemic vasculitis, which affects the veins and arteries of all sizes. Perivascular infiltration by neutrophils and monocytes and immune-mediated vasculitis cause endothelial dysfunction, and increase the risk of thrombosis. Endovascular and perivascular inflammation may lead to stenosis, thrombi and aneurism [6].

Venous involvement is more common than arterial involvement in BD. Although superficial vein thrombosis and deep vein thrombosis are commonly seen in lower extremities, upper extremity venous thrombosis is rarely seen [7].

In a study conducted by Kuzu et al. [8] involving 1200 patients, venous and arterial involvement were seen in 173 (14.4%) and 19 (1.6%) of the patients respectively. Of the patients with venous involvement, 154 (12.8%) were venous thrombosis, 17 (1.4%) were superior vena cava syndrome, five (0.4%) were inferior vena cava syndrome, five (0.4%) were varicose veins, two were upper extremity thrombosis, one was cavernous sinus thrombosis, one was internal jugular vein thrombosis and one was hepatic vein thrombosis.

The risk of venous thrombosis is increased in patients who were treated by heparin infusion or underwent venous blood sampling [9]. The venous punction performed four days ago led to a further increase in the risk of thrombosis in our patient. Therefore, higher tendency to develop thrombosis should be kept in mind, when an invasive procedure is planned in patients with BD.

Treatment of BD varies according to age, gender, clinical signs and the severity of the disease. Colchicine is the most commonly used drug for treatment of BD. It is preferred, especially for the patients with mucocutaneous symptoms [10]. Topical corticosteroids are effective treatment modalities for oral and genital ulcers. They decrease inflammation if especially used in the first five days of inflammation when the inflammation is severe. Azathioprine suppresses humoral and cellular immunity. For retinal vasculitis, it is used in combination with prednisolone at a dose of 1-2 mg/kg/day. Cyclosporin that is used for the treatment of uveitis inhibits T lymphocytes selectively. Infliximab and etanercept are tumor necrosis factor alpha inhibitors commonly used for the treatment of BD recently [11].

Vascular involvement is commonly seen in the first five years of the disease [12]. Our patient was diagnosed with pulmonary emboli a year after the diagnosis of BD, and upper extremity vein thrombosis the next year. This could be related with severe prognosis of the disease in our case patient or poor patient adherence to the treatment regimen.

For the good prognosis of the disease, regular follow-up is very important. Inadequate number of rheumatologists in Turkey leads to irregular follow-up and lower compliance of the patients with treatment. In this respect, primary care physicians should take an active role in frequent follow-up visits of the patients with BD and close monitoring of the treatment, enhance treatment compliance of the patients and refer patients with exacerbation to the Rheumatology clinic to receive immune-suppressive treatment as soon as possible.

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