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A Different Clinical Manifestation: Behçet's Disease Diagnosed with Superior Vena Cava Syndrome

Farklı Bir Klinik Tezahür: Süperior Vena Kava Sendromu İle Tanı Konulan Behçet Hastalığı

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ÖZET

Behçet Hastalığı (BH) birçok klinik durumda ortaya çıkan, nadir görülen bir otoimmün vaskülitik hastalıktır. Oral ve genital ülserler en sık klinik prezentasyon iken, superior vena kava tutulumu yaygın değildir. Otuz sekiz yaşında erkek hasta yüz ve boyunda birkaç gündür artan şişlik şikayeti ile acil servise başvurdu. Doppler ultrasonografi ile bilateral internal juguler ven trombozu tespit edildi. Tekrarlayan oral aft dışında bilinen bir hastalık öyküsü olmayan hasta, Lemierre Sendromu düşünülerek kardiyovasküler cerrahiye sevk edildi. Takiplerinde genital ülser gelişmesi üzere BH tanısı konuldu. Literatür incelememizde benzer olgu sayısı az olduğu için sunduk. Klinik prezentasyonu nadir bir bölgede tromboz ile başvuran hastaların ayırıcı tanısında kollajen doku hastalıklarının düşünülmesi için bu olgunun önemli bir örnek olacağını düşündük.

Anahtar Kelimeler: Behçet hastalığı, süperior vena kava sendromu, venöz tromboz

ABSTRACT

Behçet's Disease (BD) is a rare autoimmune vasculitic disease that presents in many clinical conditions. While oral and genital ulcers are the most common clinical presentation, superior vena cava involvement is not common. A 38-year-old male patient was admitted to the emergency department with swelling in the face and neck that had been increasing for a few days. Bilateral internal jugular vein thrombosis was detected with Doppler ultrasonography. The patient, who had no known disease history except recurrent oral aphthae, was transferred to cardiovascular surgery due to considered Lemierre Syndrome. The patient, who developed genital ulcer during follow-up, was diagnosed with BD. We presented it because there are only a few similar cases in our literature review. We thought that this case would be an important example for considering connective tissue diseases in the differential diagnosis of patients whose clinical presentation is presented with thrombosis in an uncommon region.

Key words: Behçet's disease, superior vena cava syndrome, venous thrombosis

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INTRODUCTION

Behçet's disease (BD) is a rare vasculitic disease defined by Hulusi Behçet, a Turkish dermatologist, in 1937. BD presents with oral and genital ulcers progresses with relapses and has a multisystemic chronic inflammatory feature. BD is generally seen in countries on the silk path, it may show many findings including eyes (uveitis, retinal vasculitis), skin (pseudofolliculitis, eritema nodosum), gastrointestinal (diarrhea, perforation, pain, hemorrhage), articular (arthralgia, arthritis, ankylosing spondylitis), vascular (arterial and venous thrombosis, aneurysm) and neurological (many manifestations, mostly meningoencephalitis) (1,2). Vascular involvement affects arteries and veins of all diameters. However, involvement of the veins is more common than arteries. Vascular involvement occurs mostly in the lower extremity veins and the etiology is still unknown (3–5). In the multicenter VENOST study, 108 of 1144 patients with cerebral venous sinus thrombosis (CVST) had BD (6). In a cohort study with 820 Behçet patients, the CVST ratio was in 7.8% (7). Although CVST is seen in BD, it is rarely seen as the first sign. In this article, the diagnosis, treatment and clinical course of the patient, who has no previously known systemic disease, presented with superior vena cava syndrome and diagnosed with BD, will be discussed.

CASE REPORT

A 38-year-old male patient with no known systemic disease and family history. He admitted to the emergency department with swelling and redness of the face and neck. His signs started a week ago and progressed gradually. On physical examination, there was extensive redness and swelling that included the neck and face and he had mild tachycardia (120/min). He had no fever. On neurological examination, bilateral papillary edema was detected and the other neurological examinations were normal. Doppler ultrasonography showed thrombus in the bilateral internal jugular vein. Computed tomography angiography of neck performed for detailed scanning of the patient showed thrombus in the bilateral internal jugular vein and superior vena cava (Figure 1). In blood tests, C-reactive protein (CRP) (49.7 mg/L, normal range: 0.1-5 mg/L) and white blood cells (WBC) (14.600/mm^3, normal range: 4.000-10.000/mm^3) were high. Considering the preliminary diagnosis of Lemierre syndrome, characterized by bacteremia and jugular vein thrombophlebitis, the patient was anticoagulated and antibiotherapy was added to the treatment. Antinuclear antibody (ANA) for differential diagnosis of vasculitis were negative; protein C, protein S, antithrombin III for the differential diagnosis of thrombophilia were in the normal range and anticardiolipin IgG and IgM were negative. During the hospitalization, the patient complained of ulcers in the genital area for the first time in his life. He also had recurrent oral ulcers for 2 years. His pathergy test was negative. According to The International Criteria for Behcet's Disease (ICBD) he was scored five (oral ulcer: 2, genital ulcer: 2, vascular involvement: 1) and diagnosed with definitive BD (Table 1)(8). Pulse methylprednisolone (240 mg/day) was given for 5 days. The positive HLA B51 confirmed the diagnosis. He was discharged with 1.5 mg/day colchicine, 8 mg methylprednisolone and 100 mg/day azathioprine

Table 1. International Criteria for Behçet's Disease-point score system: Scoring ≥4 indicates Behçet's diagnosis

Sign/Symptom	Points
Oculer lesions	2
Genital aphthosis	2
Oral aphtosis	2
Skin lesions	1
Neurological manifestations	1
Vasculer manifestations	1
Pozitif pathergy test*	1

*Pathergy test is optional and primary scoring system does not include pathergy testing. However, where pathergy testing is conducted one extra point may be assigned for a positive result.

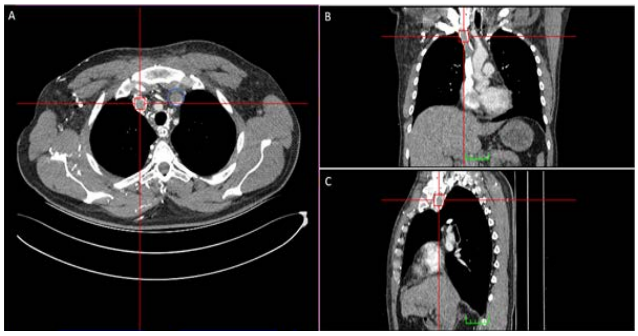


Figure 1. A) Thrombus in the bilateral internal jugular vein (right jugular vein marked with square, left jugular vein marked with circle) B) Thrombus formation extending from the right jugular vein to the superior vena cava C) Sagittal view

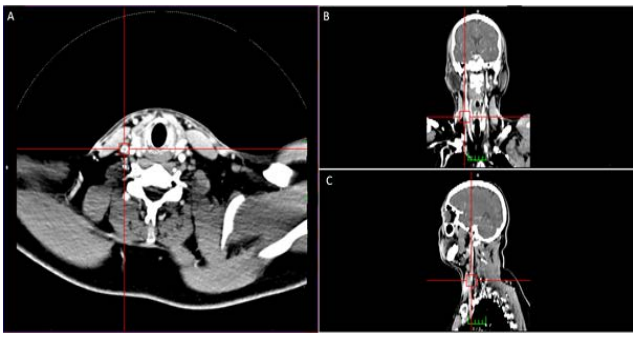
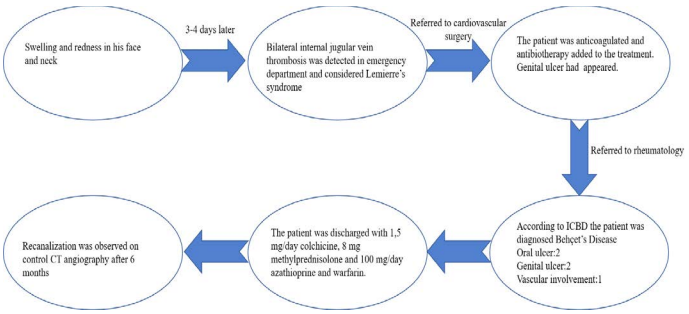


Figure 2. A)Recanalized thrombus in the right jugular vein B) Coronal view C) Lateral view



Flow chart: Patient's flow chart

and warfarin. Recanalization was observed on control CT angiography after 6 months (Figure 2, flow chart).

DISCUSSION

Behçet's disease is a chronic inflammatory vasculitic disease that can show eye, skin, vascular, gastrointestinal and neurological involvement, mostly progresses with oral and genital ulcers, and progresses with relapses (1,2). ICBD is used as a diagnostic criterion for Behçet's disease. According to these criteria; Our patient was among the possible BD diagnostic criteria due to oral aphthae and vascular involvement at the time of admission to the emergency department. The patient's first presentation was evaluated as Lemierre syndrome due to bilateral internal jugular vein thrombosis and accompanying high CRP and WBC. Lemierre syndrome is a disease with jugular vein thrombophlebitis and septic embolism caused by gram-negative bacteria (9). Lemierre syndrome was excluded because the patient did not have septic symptoms such as fever and blood culture was negative. During the follow-up, genital ulcer was detected in the patient, and the patient with an ICBD score of 5 was diagnosed with definitive BD. Although oral ulcers are seen in almost all patients with Behçet's disease, its specificity is not high since it can also be seen in many other diseases (10). Our patient was previously evaluated for oral aphthae and was evaluated as recurrent aphthous stomatitis.

BD can affect a wide variety of vascular structures such as lower extremity veins, superior vena cava, and pulmonary artery (11). Vascular involvement is seen at rates of up to 40% and is an important indicator of morbidity and mortality (12). Cerebral vein involvement was found to be 8% (7). Carotid artery involvement is rare in BD. Aguiar de Sousa et al. in the multicenter VENOST study, 4 out of 230 patients and one case of SVST in 108 patients were shown (6,13). There is no consensus on the treatment of Behçet's patients followed up with CVST (14). When anticoagulation and immunosuppression therapy were compared in Behçet's patients with deep vein thrombosis (DVT), recurrence was seen in patients given anticoagulation alone; There was no significant difference in terms of recurrence and clinical progression between patients receiving immunosuppression alone and those given immunosuppression and anticoagulation (15). The use of anticoagulants in patients with venous thrombosis due to Behçet's disease is still controversial because of potentially fatal complications in the presence of pulmonary artery aneurysm. However, there are studies showing that the use of anticoagulants contributes to clinical improvement when the presence of aneurysm is excluded and in the case of resistant venous thrombosis. Immunosuppressive therapy has been tried alone in many studies and has shown the same effect as other treatment methods (14).

In conclusion, Behçet's disease should be included in the differential diagnosis of all forms of vascular disease, especially in young and middle-aged patients, and immunomodulatory treatments should be planned with multidisciplinary clinical approaches.

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