DIAGNOSIS OF BEHCET DISEASE DUE TO VENA CAVA SUPERIOR THROMBOSIS

Case Report

VENA CAVA SÜPERİOR TROMBOZUNDAN BEHÇET HASTALIĞI TANISINA: OLGU SUNUMU

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ABSTRACT

Behcet's disease is a chronic multisystemic vasculitis that is characterized by recurrent oral apthous ulcers, genital ulcers and uveitis and its etiology remains still unknown. Although, It may affect any organ system, also vascular involvement can be seen. Venous involvement is more common than the arterial one but pathologies like as thrombosis and aneurysm formation can be seen in arterial system. Vena cava superior thrombosis can be seen in course of Behcet's disease as the years pass. We aimed to present a 29 years old male patient with thrombosis of vena cava superior that is diagnosed as Behcet's Disease in this study.

Key words: *Behcet Syndrome; Superior Vena Cava Syndrome; vasculitis.*

ÖZET

Behçet Hastalığı; nedeni bilinmeyen, tekrarlayan oral aftöz ülserler, genital ülserler ve üveit tablosu ile karakterize, kronik multisistemik bir vaskülittir. Birçok organ sistemlerini etkileyebilmekle birlikte vasküler tutulum da gözlenebilir. Venöz tutulum daha sık olmakla birlikte arteriyel sistemde de tromboz ve anevrizma formasyonu gibi patolojilere yol açabilir. Vena cava süperior trombozu Behçet Hastalığınınseyrinde yıllar içerisinde gözlenebilir. VCS trombozu kliniği ile başvuran ve Behçet Hastalığı tanısı alan 29 yaşında erkek hastamızı bu çalışmamızda sunmak istedik.

Anahtar Kelimeler: *Behçet sendromu; Süperior vena kava tıkanıklığı; Vaskülit.*

INTRODUCTION

Behcet's disease that is characterized by triple complex of recurrent oral aphthous ulcers, genital ulcers and uveitis was first described in 1937 by Dr. Hulusi Behçet, a well known dermatologist in Turkey (1). This disease is particularly common in between the Mediterranean, the Middle

East and the Far East, corresponding to the Old Silk Road, an ancient trading route stretching.

Although, incidence may change widely according to geographical regions, the prevalence is approximately 20-421/100.000 in adults in our country (2). Its etiology remains unknown.

HLA-B51 antigen is mostly associated with Behcet's disease that its incidence was found 9,4 fold higher in patients who were carried in investigation about genetic origins of disease (3). On the contrary,It was reported that HLA-DR1 and HLA-DQw1 antigen may have protective effects on development of the disease. It was claimed that viral-bacterial agents, immune complex cumulations, sex hormones and also dietary habits might be involved in etiology of disease in various studies but the etiopathology couldn't be elicited yet. However, immunologic disorders, genetic and environmental factors, bacterial and viral agents are being in charge of emergence of disease (4). International Study Group's criteria which was issued in 1990 is used for diagnosis of the disease (5). The existence of oral ulcers with two more organ involvement is required for the diagnosis.

Vena cava superior thrombosis can be seen in course of Behcet disease as the years pass. Although, the lower extremity venous thrombosis is the most common vascular involvement in the course of the disease, superior or inferior vena cava thrombosis may also be seen. We aimed to present a 29 years old male patient with thrombosis of vena cava superior that is diagnosed as Behcet's disease in this study.

CASE REPORT

A 29 years old male, who had complaints of dyspnea, recurrent aphthous ulcers, fast fatigue, swelling on the face, neck, shoulders and chest was admitted to the hospital. He had complaints about 4 months and it has been increasing in the last month. The patient had recurrent genital ulcers and uveitis.

The physical examination showed that mild cyanotic appearance of the face, bilateral periorbital edema, edema on anterior and posterior chest wall and shoulders, cervical venous filling, common venous collateral vessels and as a diagnosis vena cava superior syndrome (SVC) was suspected because of the signs and symptoms noted above. When the patient has been searched for diagnosis of Behcet's disease, recurrent genital ulcers and uveitis have also been detected. Intravenous contrast computerized tomography (CT) was performed. Thrombosis of SVC, common venous collaterals and bilateral pleural effusion was detected by CT (**Figure 1**).

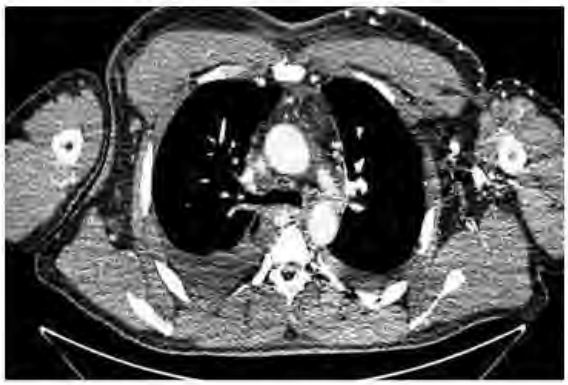

Figure 1: *Thrombosis, bilateral pleural effusions and common venous collaterals in vena cava superior syndrome.*

He had no history of systemic disease, trauma or central venous intervention. Thoracic and mediastinal masses or findings of metastasis were not detected in the physical examination. Herein, we thought that SVC thrombosis may be due to Behcet's disease. After the rheumatology consultation and investigation, diagnosis of Behcet's disease was made. The patient had negative laboratory results about systemic

lupus erythematosus and antiphospholipid syndrome.

Informed consent was taken and patient was included in the study. Low dose anticoagulation treatment, intravenous corticosteroid and diuretic treatment was started. Surgical intervention was not considered because the symptoms of patient had relieved partially by medical treatment.

DISCUSSION

Behcet's disease is a chronic multisystemic vasculitis that affects mucocutaneous, locomotor cardiovacular, pulmonary and central nervous systems. Although, It may affect any organ system, vascular involvement can also be seen. Especially involvement of cardiovascular and central nervous system are important causes of mortality. However, venous involvement is more common than the arterial one, pathologies like as thrombosis and aneurysm formation can be seen in arterial system. In arterial system, aneurysm formation is the most frequently seen and less frequently arterial thrombosis is seen. In venous system, venous thrombosis is predominant. Thrombophilic factors which are the leading cause of thrombosis weren't detected in Behcet's disease. Whereas, there are suspects on defects of fibrinolytic system (6).

It was believed that development of thrombosis was caused by vasculitis related with inflammation of vascular wall. Thromboembolic complications are rarely seen because thrombus is tightly adherent to the vessel wall. Venous involvement frequently occurs in young males at first year of the course of the disease. After the first episode, recurrent venous involvements may be seen.

The incidence of lower extremity deep and superficial venous thrombosis, superior and inferior vena cava thrombosis, cerebral venous thrombosis and hepatic vein thrombosis are in a decreasing order. The incidence of vena cava inferior thrombosis is reported as % 0,4 to7,4 in various studies (7).

Vena cava superior syndrome due to thrombosis is more frequent than inferior vena cava and may have similar symptoms and may be confused with the other causes of the disease. Some of these are dyspnea, cyanosis of face, edema on neck, upper extremity and chest, vasodilatation, cough, headache…etc. Lung cancer is the most frequent disease and less frequently thoracic and mediastinal malignancy, pacemaker and central catheter interventions, infection related fibrosis (tuberculosis, sphilisis …etc.), post-radiation vascular fibrosis, Behcet's disease and planjon guatr take place in the etiology of SVC syndrome (8).

Head and neck edema, cyanosis, distention of internal jugular vein, common venous collaterals, pupillary edema, pleural effusion and chylothorax can be *detected* in patient's physical examination. Computerized Tomography, Magnetic Resonance Imaging and Transesophageal Echocardiography can be used as diagnostic tools in vena cava superior thrombosis. Role of antiocoagulation treatment in Behcet's disease related thrombosis is still contraversial. However, most doctors give anticoagulation treatment for Behcet's disease related venous thrombosis in practical applications.

On the other hand, some authors have offered that thrombosis in Behcet's disease is due to vasculitis related intimal injury and because of this, anticoagulation treatment is useless. Palliative treatments like; head elevation, periodic oxygen support, restriction of fluids, and medical treatment with corticosteroids, diuretics and immunosuppresive agents (azothioprine) can be used. Fibrinolytic treatment can be used in relapses but an effective result can't be achieved always.

Surgical intervention may be a treatment of choise in patients with occlusion of vascular and intracardiac structures who have unstabile hemodynamic condition, advanced symptoms and insufficient vascular collaterals. Medical treatment which is a component of perioperative treatment shouldn't be neglected to preclude recurrence of disease in long term.

Vena cava superior thrombosis may be seen in course of Behcet's disease. However, Behcet's disease is rarely diagnosed during investigation of the etiology of VCS syndrome in literature [9]. Behcet's disease should be held in mind while investigating the etiology of vena cava superior thrombosis in young and middle aged patients without history of malignancy.

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