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Case Report

Behcet's Disease Presenting with Focal Fibrosing Mediastinitis and SVC Thrombosis: A Case Report with Review of Literature

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Abstract

Introduction

Behcet's disease is a rare, multisystemic disease marked by vasculitis and a triad of recurrent oral and genital mucosa ulcers, as well as relapsing uveitis and thrombophlebitis. Mediastinal fibrosis is extremely rare in Behcet's disease.

Case presentation

We present ultrasound and computed tomography (CT scan) in a 26-year male with mediastinal fibrosis and superior vena cava (SVC) thrombosis, with thrombus in left internal jugular veins (IJV) associated with Behcet's disease.

Conclusion

Behcet's disease can cause mediastinal fibrosis in rare cases. CT and MRI are efficient methods for detecting mediastinal fibrosis and superior vena cava thrombosis.

1. Introduction

Behcet's disease (BD) is a chronic, multisystemic autoinflammatory disorder characterized by recurring oral aphthous ulcers, genital ulcers, skin lesions, eye lesions, and other abnormalities. [1]. It was first described by Dr Hulusi Behcet, a Turkish ophthalmologist, in 1937. Recurrent oral and genital ulcerations, as well as uveitis, comprise the classic triad. Other systemic manifestations of Behcet's disease have been reported, including skin lesions, arthritis, thrombophlebitis, neurologic and pulmonary involvement [2].

The disease progresses more rapidly in men than in women and in those under the age of 25 at the time of onset. Behcet's disease

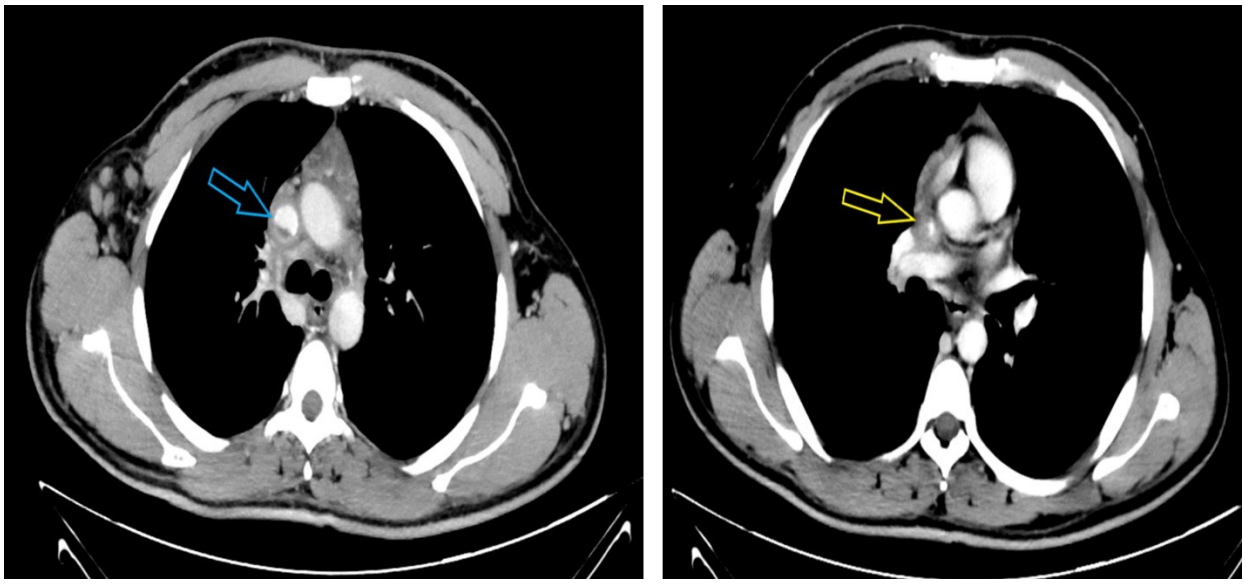

Figure 1. Contrast-enhanced CT, axial section, shows an ill-defined enhanced area around SVC and narrowing its lumen, obliterating fat planes, obliterating azygos vein (yellow arrow), and a thrombus in SVC (blue arrow)

is most common in the Mediterranean and the Middle East [1]. Pleuropulmonary involvement is rare, occurring in 1-8% of patients, and is typically more severe in men. The presence of mediastinal fibrosis in Behcet's disease is very rare [3]. Superior vena cava (SVC) syndrome can be caused by extrinsic compression, conjugation of external compression, and SVC thrombosis, a rare but well-recognized manifestation of Behcet's disease [2,4].

This study aims to report a case of Behcet's disease complicated by SVC thrombosis due to extrinsic compression by mediastinal fibrosis.

2. Case Presentation

2.1. Patient information and Clinical findings

A 26-year-old male presented with a history of headache and neck engorgement for a one-week duration. These symptoms were generalized body weakness.

Physical examination revealed aphthous ulcers in the mouth, which were tender and regarded by GP as ordinary aphthous ulcers, also one ulcer in the genitalia in thorough examination

2.2. Diagnostic assessment

Ultrasound of the neck revealed engorged both IJVs, even in sitting position, with thrombus in left IJV. CT chest with IV contrast revealed dilated both IJVs with thrombus in left IJV (Figure 1), dilated upper part of SVC, irregular enhanced area obliteration fat planes around SVC, narrowing its lumen, and a thrombus in SVC but not completely occluding it, obliteration azygos vein proximal to its drainage to SVC (Figure 1 and Figure 2). C-reactive protein was elevated, CBC was normal, and renal function was normal.

2.3. Therapeutic intervention

The patient management conservatively. He was given Rivaroxaban 15 mg x2, Prednisolone tab 5mg x1 and Ibuprofen tab 400 mg x2.

2.4. Follow-up

The patient was followed up for 4 months, he responded partially to the treatment.

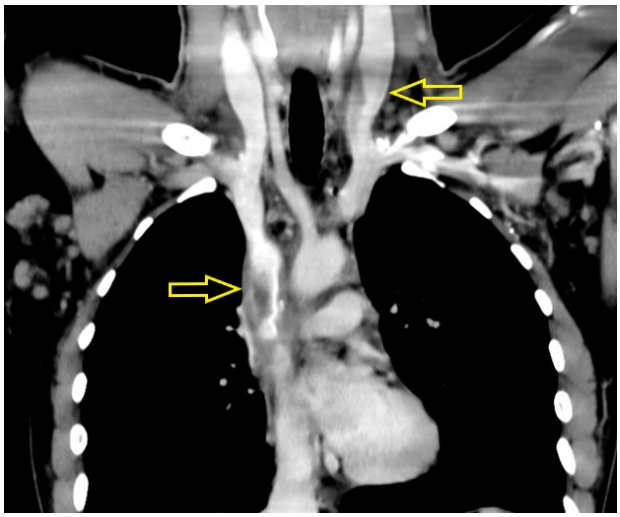

Figure 2. Contrast-enhanced CT, coronal section, shows thrombus in left IJV and SVC (yellow arrows) with focal mediastinitis around SVC

4. Discussion

Fibrosing mediastinitis (FM) is a rare benign disorder caused by proliferation of cellular collagen and fibrous tissue within the mediastinum. The pathogenesis is hypothesized to be a severe fibroinflammatory host response to various antigens, such as Histoplasma, Mycobacterium, or Treponema, and autoimmune conditions, such as granulomatosis with polyangiitis. Behcet's

disease and IgG4 disease have also been associated with FM [5]. Whereas mediastinal fibrosis associated with Behcet's disease has been reported very rarely [2]. FM is classified into two types. The granulomatous type, also known as the focal subtype, is the most common in the United States, accounting for approximately 90% of cases, and is primarily caused by infection (histoplasmosis, tuberculosis, mucormycosis, and blastomycosis) with histoplasmosis being the most common pathogen. The non-granulomatous or diffuse subtype is idiopathic, rare, and associated with various autoimmune diseases (SLE, rheumatoid arthritis, Behcet's disease, IgG4), radiation, and certain drugs such as methysergide in certain types of lymphoma [5]. Patients with FM may remain asymptomatic for months or years before significant mediastinal structure obstruction occurs. Common symptoms include shortness of breath, hemoptysis, dysphagia, headaches, facial swelling, syncope, and chest pain [6].

Behcet's disease is a chronic relapsing systemic vasculitis in which orogenital ulceration is a prominent feature. Because Behcet's disease affects multiple systems and causes hypercoagulability, it should be considered in the differential diagnosis of SVC thrombosis caused by mediastinal fibrosis, even if the cardinal findings of Behcet's disease are absent [2]. Vascular manifestations are seen in 10-40% of cases of Behcet's disease, with venous manifestations accounting for 80–90% of cases. Involvement of the venous system is most frequently seen in the form of thrombophlebitis. According to Rodriguez-Carballeira et al., vascular thrombosis is common in 19.7% percent of the Spanish registry, and men have a higher prevalence of ocular involvement and venous thrombosis [7]. SVC thrombosis can be either primary or secondary to axillary or subclavian vein thrombosis. It usually happens after the disease's mucocutaneous manifestations have faded. It is rarely the first manifestation preceding the other events [6]. Clinically, it may be latent, well-tolerated, with a silent evolution, or it may have a rather aggressive manifestation characterized by headache, upper extremity and facial swelling, vision changes, and cerebral edema [7]. The current case was a 26-year-old male with Behcet's disease who presented with a one-week history of headache and neck engorgement. Our case had aphthous ulcers in the mouth and one ulcer in the genitalia.

According to the International Study Group for Behcet's disease diagnostic criteria, the detection of oral ulcers and at least two of the following criteria is used to make the diagnosis: recurrent genital ulcers, ocular lesions such as uveitis and retinal vasculitis, and skin lesions like folliculitis and erythema nodosum [1]. The role of imaging in diagnosis is excellent when SVC thrombosis is suspected. This includes chest radiographs but especially computed tomography and magnetic resonance imaging [7]. The radiological appearance of fibrosing mediastinitis varies depending on the type (diffuse or focal) and pathogenesis (granulomatous and non-granulomatous). Granulomatous type is mainly located in paratracheal, subcarinal, or pulmonary hilum, and it contains stipple or coarse calcification, usually causing SVC syndrome. Non-granulomatous type affects multiple compartments, and calcification is uncommon [8].

The CT scan in our case revealed an ill-defined enhanced area around the SVC, narrowing its lumen, obliterating the azygos vein and fat planes, and a thrombus can be seen in the SVC and the left IJV. While de Paiva and colleagues demonstrated the case with Behcet's disease, the chest scan revealed a narrowing of the SVC lumen due to vessel wall thickening but no evidence of vena cava thrombosis [4]. Treatment of FM is extremely challenging. Patients with Behcet's disease respond well to standard steroid and colchicine therapy, as well as anticoagulant therapy for vascular thrombosis [2].

5. Conclusion

Behcet’s disease should be suspect in young patients with the rare condition of diffuse mediastinal fibrosis associated with a thrombus of SVC. CT and MRI are effective tools for detecting mediastinal fibrosis and thrombosis of the superior vena cava.

Declarations

Conflicts of interest: The author(s) have no conflicts of interest to disclose.

Ethical approval: Ethical approval is not required for this study in accordance with local or national guidelines.

Patient consent (participation and publication)**:** Written informed consent was taken from the patient for the publication of any related information and images or illustrations.

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Authors' contributions: SHT The radiologist who performed the assessment, major contribution to the idea and revision of the manuscript. FHK, DMH Literature review, writing the manuscript, major contribution to the conception of the study. SAM, KMS and AMS were involved in data interpretation and in the writing of the first draft of the manuscript. RJR, SHM, and HMH were involved in obtaining patient data, and critically revise of the manuscript. SHT and AMS confirm the authenticity of all the raw data. All authors read and approved the final manuscript.

Data availability statement: The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

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