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A Rare Case of Behçet's Disease Highlighting Uncommon Dilations: Case Report

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***Abstract—Introduction*: Behçet's disease (BD) is traditionally diagnosed based on clinical symptoms such as recurrent oral and genital ulcerations, eye and skin lesions, and, in some cases, a positive pathergy test. However, the clinical presentation of BD has evolved, with recent reports highlighting a broader spectrum of manifestations, including vascular involvement in nearly half of affected patients. Our report presents a unique case of a 42-year-old male with BD and atypical vascular involvement. This case is noteworthy due to the atypical vascular dilations, which extend beyond the vascular sites commonly observed in BD. It underscores the complex and diverse presentations of BD and highlights the importance of maintaining a high index of suspicion for atypical vascular involvement. The patient's symptoms were effectively managed with immunomodulatory therapy, demonstrating the need for individualised treatment strategies. This report calls for further research to improve the understanding and management of such rare BD presentations, contributing valuable insights to the existing literature.**

***Index Terms*— Atypical; Behçet's disease; Case Report; Thrombosis; Vascular Dilation.**

I. INTRODUCTION

Behçet's disease (BD) is a complex, systemic disorder that can affect multiple organ systems, with hallmark manifestations including recurrent oral and genital ulcers and uveitis. First described by Turkish dermatologist Hulusi Behçet [1], it has since been recognised for its broad range of clinical manifestations, including involvement of the cardio-vascular, central nervous, articular, and gastrointestinal systems. While the classic manifestations of BD are well-documented, vascular involvement such as venous thrombosis is a frequent and significant feature of the disease. However, such vascular manifestations are not limited to the commonly-observed conditions, and can sometimes present in atypical forms, challenging diagnosis and management. While the pathogenesis of BD remains incompletely understood, a combination of genetic, environmental, and immunological factors is believed to play a role [2]. The disease is characterised by an abnormal immune response, which leads to endothelial dysfunction, increased vascular permeability, and exaggerated inflammatory and coagulation states. The vascular changes often lead to thrombo-inflammatory events, which are significant complications associated with Behçet's Disease (BD). While vascular manifestations typically present as deep vein thrombosis or superficial venous thrombosis [3], atypical forms of vascular involvement such as extensive venous dilation are less common but important to recognise. This case study reports one such atypical vascular manifestation in a BD patient. Typically BD affects individuals in their third or fourth decade of life, with males often experiencing more severe symptoms [4], probably owing to an increased hypercoagulable state in men compared with women [5]. The disease is most prevalent in regions such as Turkey, the Mediterranean, and the Middle East [6]. Literature indicates that vascular manifestations occur in up to 50% of BD patients [7], underscoring the significant role of vascular involvement in the disease. Therefore, it is crucial to consider BD in patients presenting with atypical venous manifestations.

II. METHODS

Case presentation

A 42-year-old male smoker presented with complaints of weakness, fatigue, joint pain, and coughing. Initial examination indicated dilated

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veins in the upper and lower extremities, abdomen, chest, and scrotum. (Figure 1-4). The patient had a history of recurrent painful oral and genital ulcers over the previous few years. Additionally, he had recently undergone an atrial thrombectomy following the diagnosis of superior vena cava syndrome (SVCS) and the detection of a thrombus in the right atrium. At that time, he had been suffering haemoptysis, and his chest computerised tomography (CT) scan prior to that procedure had shown cavitary lesions covering almost the entire inferior lobe of the left lung, as well as partially increased wall thickness and volume loss in the left lung. Moreover, linear sequelae fibrotic changes were observed within the inferior lobe, accompanied by increased ventilation in the right lung. He was later diagnosed with an empyema infection upon admission with coughing and dyspnoea. Linezolid treatment was initiated as coagulase-negative staphylococcus was detected in cultures. After this first episode of empyema infection, recurrent episodes necessitated the placement of a thoracic drainage tube. Consequently, an iatrogenic cavitary lesion developed in his left lung parenchyma, due to which a partial pleurectomy was performed. In a thoracic surgery council, the patient's condition was evaluated as auto-pneumonectomy. In addition to the typical characteristics of BD, a positive pathergy test confirmed the diagnosis. The patient was hospitalised for a comprehensive evaluation and close follow-up of the vascular symptoms and associated complaints. Considering his multi-organ involvement and recurrent symptoms despite antibiotics and other guidelines-suggested

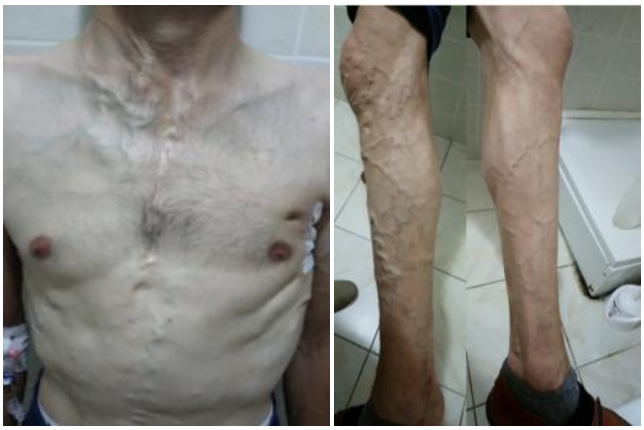

Figure 1 (left) shows dilated veins of the abdomen and chest, while **Figure 2** depicts dilated veins of the lower extremities.

medications such as prednisone, we decided to initiate intravenous immunoglobulin (IVIG) therapy, which produced a significant response, and the patient's symptoms improved.

III. DISCUSSION

Behçet's disease (BD) is a chronic, relapsing condition characterised by systemic inflammation

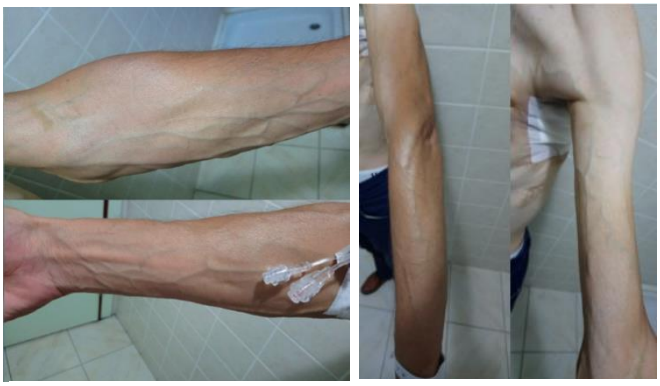

Figure 3 shows dilated veins of the upper extremities.

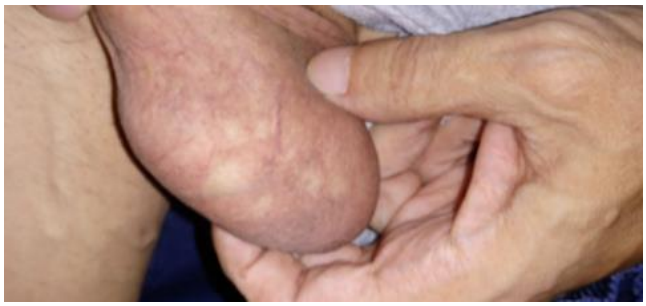

Figure 4 show dilated veins of the scrotum

and immune system dysfunction. Common manifestations include recurring oral and genital ulcers, skin lesions, uveitis, retinal vasculitis, neurological issues, venous thrombosis, and gastrointestinal disturbances. Since there is no single symptom specific to BD, diagnosis is based primarily on clinical presentation, as outlined by the International Study Group for Behçet's Disease [8]. The reported patient had already been diagnosed with BD based on his clinical symptoms. Additionally, a positive pathergy test was performed to further support the diagnosis. Patients with BD are also vulnerable to severe pulmonary complications, including haemoptysis and recurrent empyema. Haemoptysis is often linked to pulmonary artery aneurysms, a relatively common and potentially life-threatening manifestation of BD if they rupture [9]. Moreover, right atrial thrombosis, a potential complication of BD, can

result in superior vena cava syndrome (SVCS), which is marked by symptoms such as facial swelling, cyanosis, and haemoptysis due to impaired venous return and increased venous pressure. In our patient's case, the recurrent episodes of empyema were likely exacerbated by immunosuppressive treatment used to manage the inflammatory aspects of BD. While these therapies are essential for controlling the disease, they also increase the risk of infections [10].

Additionally, we suspect that the patient's male gender may have contributed to the severity of his condition. Research indicates that men with BD often experience more severe symptoms than women [11]. This could be due to the influence of testosterone, which might trigger an overreaction of neutrophils and Th1 cells, contributing to more aggressive disease progression [12]. Although chronic vascular inflammation is a hallmark of BD, our patient also developed a hypercoagulable state, eventually leading to the diagnosis of SVCS. This hypercoagulability required invasive intervention in the form of atrial thrombectomy to remove a thrombus in the right atrium. This event may be linked to the patient's atypical vascular dilation, a phenomenon previously associated with thrombotic events in BD, which leads to substantial vein wall remodelling [13]. In BD patients, vein wall thickness (VWT) tends to increase even in the absence of overt vascular involvement [14].

The patient's history of smoking is another crucial factor to consider. Exposure to nicotine and carbon monoxide can impair endothelial vasoreactivity, contributing to vascular dysfunction and possibly exacerbating the hypercoagulable state in BD patients [15].

Intravenous immunoglobulin (IVIG) is generally reserved for severe or refractory cases of BD, or when there are overlapping autoimmune conditions. It exerts immunomodulatory effects, reducing inflammation and modulating the immune response. However, its use should be considered on a case-by-case basis.

In conclusion, we believe that certain factors such as male gender and smoking may increase the risk of atypical venous complications in BD patients, necessitating closer monitoring and early intervention. This case highlights the importance of considering BD in patients presenting with atypical venous dilation, as vascular manifestations are observed in approximately 50% of BD cases.

IV. FUNDING

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V. CONFLICT OF INTEREST

The authors declare that they have nothing to disclose regarding funding or conflict of interest with respect to this manuscript.

VI. ACKNOWLEDGMENTS

None.

VII. PATIENT CONSENT

The patient's consent was obtained for the publication of this study, which was conducted in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

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