

Case Report

May-Thurner Syndrome as The Presenting Symptoms of Behcet's Disease: A Rare Case Report

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Abstract**Introduction**

May-Thurner syndrome as a presenting symptom of Behcet's disease is rare in the literature. The aim of the current study is to report a rare case of Behcet's disease which has been presented with May-Thurner syndrome.

Case presentation

A 26-year-old woman presented with left leg pain for a one-year duration. On examination, there were dilated superficial veins and edema in the calf. A computed tomography scan of the pelvis showed a markedly narrowed left common iliac vein which was compressed by the right common iliac artery, suggesting May-Thurner syndrome. After a period of not responding to medications, she had been referred to a rheumatologist. The patient was diagnosed with Behcet's disease. She received immunosuppressant agents and glucocorticoids. The patient's symptoms improved after two months of follow-up.

Conclusion

May-Thurner syndrome could be an early manifestation of Behcet's disease, which may further complicate the clinical picture of Behcet's disease.

1. Introduction

Behcet's disease (BD) is a chronic, multisystemic autoinflammatory condition characterized by recurrent oral aphthous ulcers, genital ulcers, skin lesions, eye lesions and other abnormalities [1]. It is an inflammatory disease of the vascular system, and has been described as variable-vessel vasculitis in the Chapel Hill classification [2]. The BD's epidemiology is distinctively spread across the ancient Silk

Road from Mediterranean nations, such as Turkey, which has the greatest prevalence rate (80 to 370 cases per 100,000), to Middle Eastern and East Asian countries, but it is less frequent in northern Europe, North America, Australia, and Africa [3,4,5]. Vascular involvement in BD occurs in up to 40% of all patients [6]. Venous wall inflammation, manifested as superficial and deep venous thrombosis (DVT), is more common than arterial involvement [6,7]. May-Thurner

syndrome as a presenting symptom of BD is extremely uncommon [8].

The aim of the current study was to report an extremely rare case of BD presenting with May-Thurner syndrome.

2. Case Presentation

2.1. Patient information and Clinical findings

A 26-year-old married woman presented with chronic left leg cramp-like pain for one year duration, associated with swelling but no paresthesia. She had a negative past medical and surgical history. The pain had been present almost all the time during the last year without any known relieving or exacerbating factors.

2.2. Diagnostic assessment

On examination, the patient was aware and active, with no fever, cyanosis or pallor. There had been dilated superficial veins and edema in the calf, no ankle or knee swelling, no coldness and bilateral distal pulses were positive. She had visited many physicians for this condition.

2.3. Therapeutic intervention

All hematological investigations were normal. Pelvic ultrasound showed marked varicosity and dilated venules on the left side of the pelvis, leading to pelvic congestion. A computed tomography (CT) scan of the pelvis showed a markedly narrowed left common iliac vein, which was compressed by the right common iliac artery (Figure 1), suggesting May-Thurner syndrome. Finally, she was diagnosed as a case of May-Thurner syndrome. She was managed with aspirin and rivaroxaban. The patient was referred to a rheumatologist and still complained of leg pain and arthralgia. The patient reported that she had recurrent oral and genital ulcerations. There was pelvic venous thrombosis (VT) by ultrasound. Based on these findings, the patient was diagnosed with BD. She had been put on colchicine (0.6 mgx1), prednisolone (20 mgx2), and azathioprine (100 mgx2).

2.4. Follow-up

After two months of follow-up, all of the patient's symptoms improved.

4. Discussion

Behcet's disease is a life-threatening chronic inflammatory disease with the involvement of multiple organs [4]. According to a recent study, psoriasis, psoriatic arthritis, BD and spondyloarthropathies share the same immunopathogenic basis [9]. Vascular BD affects vessels of all sizes, both in the arterial and venous systems, and the incidence varies in different countries [5,10]. Vascular thrombosis in BD is a serious condition that primarily affects the lower extremities but can also affect the superior vena cava, inferior vena cava, suprahepatic arteries, pulmonary artery and heart [7,11]. Lower extremity VT in BD often develops within 5 years of disease

onset and affects the proximal venous system of the lower extremity, as found in the general population [12]. Men predominate among people with vascular BD, and these thromboses tend to have a more scattered pattern than idiopathic DVT [5]. The incidence of major vessel involvement is 5–10%. In BD, arterial manifestations, which occur in 5% of all patients, are among the leading causes of morbidity and mortality [10].

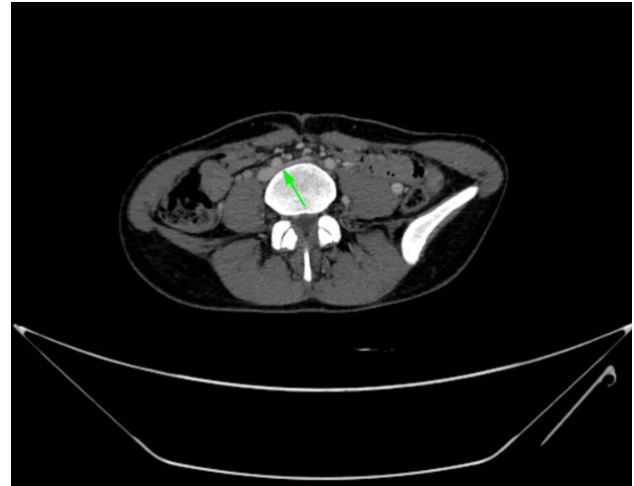


Figure 1: Axial section of contrast enhanced CT of pelvis shows compressed left common iliac vein "green arrow" by right common iliac artery.

The pathophysiology of thrombosis in BD is yet unknown. Because no consistent underlying defect of the coagulation or fibrinolytic system has been found, it is unlikely that thrombophilia plays a role in the pathophysiology of thrombosis in the majority of individuals with BD [7]. A common theory holds that active BD causes vasculitic endothelial dysfunction, which increases the risk of thrombosis. However, other vasculitis syndromes, such as Polyarteritis Nodosa and Giant Cell Arteritis, affect the endothelium but do not increase the risk of thrombosis as much as BD [13]. The human leukocyte antigen (HLA) complex on chromosome 6p21 is the most extensively studied genetic locus in BD. HLA-B*51 has consistently been associated with disease susceptibility. A recent meta-analysis found that HLA-B*51 carriers were much more likely than non-carriers to acquire BD across several geographic regions [14].

The onset of symptoms is most common in young people aged 20 to 40 years, but it is also found in children on rare occasions [15]. The diagnosis of BD is challenging, especially in low-prevalence countries. The International Study Group (ISG) Criteria, which are widely accepted as a diagnostic tool, have poor sensitivity, particularly in early instances when a significant organ involvement, such as uveitis or DVT, appears without other prominent signs [15]. Furthermore, in recent years, incomplete BD has increased in far eastern countries such as Japan and Korea [16]. The International Criteria for Behcet's Disease provided a new criterion based on multinational data that has substantially higher sensitivity than the ISG criteria while maintaining reasonable specificity [17]. The current case initially had symptoms of May-Thurner syndrome; however, she subsequently developed recurrent oral and vaginal ulcers as well as lower extremity VT.

New treatments with various target molecules are increasingly being investigated in people with BD [18]. The disease's heterogeneous presentations necessitate a personalized therapy plan based on the organs involved [19]. Each manifestation has its own "mild versus severe" forms, therefore, a careful assessment of the risk of long-term injury is required to establish the duration of therapy, the choice of immunosuppressive agents, and proper patient protection against medication toxicities [20]. Despite inconclusive results from controlled studies, colchicine, either alone or in combination with short-term topical glucocorticoids, is regarded as the first-line therapy for oral and genital ulcers and acne-like lesions in daily practice due to its safety and tolerability [20].

Glucocorticoids, azathioprine and cyclophosphamide are first-line therapies for vascular BD. However, growing evidence suggests that tumor necrosis factor (TNF) inhibitors may be possible options for the treatment of resistant vascular BD in everyday practice. Anticoagulant use for vascular BD is also debatable because of inadequate data from retrospective investigations [21]. Hamuryudan et al., found that vascular and neurological involvement was less common among individuals who had received azathioprine treatment during a 7-year follow-up [22]. A retrospective case study also demonstrated azathioprine's therapeutic effects on vascular involvement [23].

Although immunosuppressive therapy is the mainstay of vascular BD management, there is no agreement on anticoagulation [21]. Although anticoagulation with venous stenting and pharmaco-mechanical thrombectomy is the mainstay of treatment for acute DVT in the setting of May-Thurner syndrome, immunosuppression is highly recommended in DVT guidelines in BD [8]. A retrospective study evaluating several vascular BD treatment modalities in several centers was conducted. Patients who received immunosuppressive therapy alone or immunosuppressive therapy combined with anticoagulants had the same risk of recurrence [24]. Another retrospective study found no beneficial effect of anticoagulants on the development of post-thrombotic syndrome following DVT [25]. However, approximately half of the vascular BD patients in a multi-center study had been using anticoagulants at some time throughout their disease course, indicating that physicians still prefer to utilize anticoagulation in vascular BD on an expert-based approach [21]. The current case was first treated with anticoagulation but was unresponsive; after being diagnosed with BD, she was managed with immunosuppressives, and glucocorticoids, and clinical symptoms improved significantly.

Interventional approaches for BD-related DVT are seldom reported. Han et al., described a successful percutaneous transluminal angioplasty (PTA) of a short-segment inferior vena cava occlusion that causes Budd-Chiari syndrome [26]. Tekbas et al., described five patients with persistent lower extremity DVT who were managed with catheter-directed thrombolysis, PTA, and stenting. The success rate for the recanalization of chronic lesions was roughly 50% [27]. Relapses are frequent despite excellent immunosuppressive therapy, and disease-related damage develops in a considerable subset of patients, particularly those with visual, vascular, and neurological involvement [20].

5. Conclusion

May-Thurner syndrome could be considered a possible early manifestation of BD in young people who are resistant to therapy and have no known risk factors for vascular thrombosis. May-Thurner syndrome may further complicate the clinical picture of BD. These individuals should be given therapeutic immunosuppression in addition to anticoagulation.

Declarations

Conflicts of interest: There is no conflict to be declared.

Ethical approval: Not applicable.

Patient consent (participation and publication): Consent has been taken from the patients and the family of the patients for publication of this report.

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Authors' contributions: SAM The physician that managed the case. MNH and MNH conceived and designed the study. FHK wrote initial and final draft of article, and provided logistic support. SHT The radiologist performing the assessments. BAA, DAH and SMA conducted research, provided research materials, and collected and organized data. All authors have critically reviewed and approved the final draft and are responsible for the content and similarity index of the manuscript.

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