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CASE REPORT

Behçet's Disease: Challenging recurrent thrombosis despite dual anticoagulation – A case report

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Author's Contribution

[1, 2] Write-up of manuscript

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A B S T R A C T

This case study investigates the intricate management challenges associated with Behçet's disease, with a specific focus on recurrent thrombotic events, including deep vein thrombosis and pulmonary embolism in a 41-year-old female patient. Despite adherence to recommended therapeutic management, the patient's persistent thrombotic recurrences highlight the nuanced nature of Behçet's disease and its resistance to conventional treatments. It highlights the complexity of Behçet's disease, necessitating a reevaluation of current guidelines and the need for tailored therapeutic approaches for effective management.

Keywords: *Behçet's disease; Recurrent thrombosis; Pulmonary embolism; Anti-coagulant therapy*

Introduction

Behçet's disease (BD) is a rare and inadequately recognized condition characterized by inflammation in blood vessels and tissues. It affects individuals of both genders during their middle years.[1] It is characterized by painful sores in the genital area and mouth, with potential symptoms affecting the eyes, nervous system, and gastrointestinal tract.[2] The disease is associated with pulmonary embolism, distinct from deep vein thrombosis but is connected to endothelial damage. The primary approach to treating venous involvement, including deep vein thrombosis, is the administration of corticosteroids and immunosuppressive medications such as azathioprine, Cyclosporine, or cyclophosphamide.

The efficacy of anticoagulants in deep vein thrombosis is under discussion. Refractory cases may be addressed with anti-TNF-alpha agents, either alone or combined with traditional Disease-modifying antirheumatic drugs or interferon-alpha, potentially with anticoagulants, considering bleeding risk and aneurysms. Extensive thrombosis, particularly in the vena cava, may necessitate combining anticoagulants with immunosuppressive therapy, with cyclophosphamide often being preferred. A triple combination of steroids, immunomodulating/immunosuppressive drugs, and anticoagulants is considered an effective approach.[3] It is a chronic, recurrent systemic inflammatory condition with unidentified underlying causes. It is hypothesized that a

combination of genetic, infectious, and immunological factors contribute to its etiology.[4]

Case report

A 41-year-old female with a previously remarkable medical history presented with a complicated dermatological condition. Seven years ago, the patient's health deteriorated as she started to have repeated fever episodes, loose stools, oral ulcers, and painful hemorrhagic nodules in her body. Later, she developed bilateral leg ulcers and pulmonary embolism (PE) secondary to right arm deep vein thrombosis (DVT). A comprehensive diagnostic workup led to the confirmation of Behçet's disease and the concurrent presence of pyoderma gangrenosum. She received a heparin infusion, steroids, anticoagulant therapy, and continued symptomatic treatment (Tables 1 and 2).

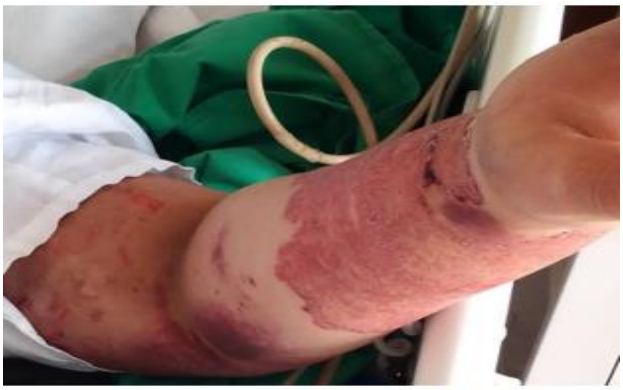

Figure 1: Clinical presentation of skin ulceration and erythematous patches

Recently, the patient was presented with the emergence of skin ulcerations, initially localized to her lower extremities, which gradually extended to involve her bilateral upper limbs and trunk over one year. She also experienced recurring thrombosis, specifically DVT and PE. The ulcer was initially presented as a 1-2 cm blister and later grew, reaching dimensions of approximately 3-4 cm.

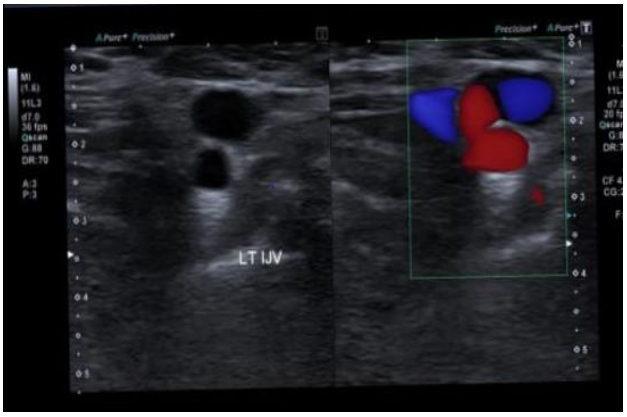

Figure 2: Venous Doppler of Left Upper Limb

***Note.** Partial thrombus in the distal segment of the internal jugular vein, and significant soft tissue edema

Additionally, it was accompanied by purulent discharge. During the clinical examination, the patient exhibited pallor, facial swelling, edematous neck, as well as aphthous ulcers in the mouth and over the body. The patient was taking Deltacortil 30 mg Hs, Tab Rivaroxaban 20 mg OD, Azainopine 50 mg OD, Rituximab 200mg OD, and Enoxaparin 80 mg BD as part of her routine treatment. However, the patient's demise occurred after a recent pulmonary embolism episode.

Table 1: Laboratory Investigations

Investigation	Value	Reference Range
Prothrombin Time, Plasma	11.8 sec	9.3-12.8
INR	1.1 ratio	0.9-1.2
APTT	28.1 sec	22.9-34.5
BUN	18 mg/dl	6-20
Serum Creatinine	0.8 mg/dl	0.6-1.1
eGFR	>60	>60 ml/min
Serum Sodium	137	136-145
Serum Potassium	3.8	3.5-5.1
Serum Chloride	94 mmol/l	98-107
Serum Bicarbonate	31.4 mmol/l	20-31
Serum Procalcitonin	0.13 ng/ml	<0.5 low risk of severe sepsis >2.0 High risk of severe sepsis
Hb	12.8 g/dl	11-14.5
HCT	39.7%	34.5-45.4
WBC	11.7*109/L	4.6-10.8
Monocytes	15.3%	3.9-10
Platelets	498*109/L	154-433

Table 2: Urine Detailed Report

Investigation	Value
Urine color	Yellow
Appearance	Clear
SP Gravity	1.015
pH	5
Protein	0.25 g/l (1+)
Glucose	Negative
Ketone	Negative
Urobilinogen	Negative
Bilirubin	Negative
Hb	25/ul (2+)
Nitrite	Negative
Leucocyte Esterase	100/ul (2+)
RBC	Occasional
Leucocytes	04/HPF

Discussion

The National Institute for Health and Care Excellence (NICE) guidelines recommend a comprehensive approach in administering anticoagulation medications for patients with Behçet's disease, for recurrent DVT and PE. They recommend initiating anticoagulation promptly, even before baseline investigations. The available anticoagulant options consist of apixaban or rivaroxaban, with additional alternatives such as low molecular weight heparin, dabigatran, or edoxaban. Individual considerations are crucial for patients with contraindications or specific conditions. The initial three-month duration of anticoagulation should be followed, with an extended period for those with heightened thrombotic risk. In cases of treatment failure, a systematic approach is recommended, including checking adherence, investigating hypercoagulability sources, and adjusting the anticoagulation regimen.[5]

The guidelines of the American Society of Hematology (ASH) recommend the use of direct oral anticoagulants (DOACs) as the first-line treatment for acute DVT or PE, emphasizing their preference over vitamin K antagonists (VKAs) in most cases. These guidelines also advocate for individualized approaches, taking into consideration factors such as bleeding risk and the presence of right ventricular dysfunction.[6] While anticoagulation stands as the established therapy for systemic venous thrombosis, its application in Behçet's disease (BD)-associated venous thrombosis is still a debated matter.[7] Despite dual anticoagulant therapy, the management of recurrent DVT and PE proved challenging in this case and the patient could not survive. The absence of an optimal management approach for BD patients with thrombosis leaves the specific role of long-term anticoagulation in these cases uncertain.

Conclusion

The primary focus of treating Behcet's disease, especially for vascular symptoms, lies in the use of immunosuppressive drugs, steroids, and anticoagulants. The case underscores the need for ongoing research and tailored therapeutic strategies for patients with Behcet's disease and associated recurrent thrombotic events, highlighting the limitations of existing guidelines and the persistent challenges in achieving optimal outcomes for this complex medical condition. There is a need to reevaluate the guidelines and tailor therapeutic strategies for treatment-resistant cases.

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