



# Deep Venous Thrombosis in Behcet's Syndrome: is Anticoagulation Necessary?

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## Abstract

Deep vein thrombosis (DVT) in the lower extremities is a common medical presentation. Anticoagulation is the cornerstone of management. However, not all DVTs require anticoagulation. We report a case of DVT in a patient with Behcet's Syndrome where venous inflammation is the primary pathology and anti-inflammatory therapy is primary and the role of anti-coagulation is moot.

## Introduction

Behcet's syndrome is a chronic, multisystem, inflammatory disease of unknown cause, classified as a variable vessel vasculitis using the Chapel Hill consensus criteria [1]. It is rare in Australia with highest prevalence found in Turkey and other countries that line the Silk Road [2]. It is usually defined by recurrent mucocutaneous ulceration with ocular involvement or other systemic manifestations as described in the International Criteria for Behcet's Disease [3] (Table 1). Non-erosive arthritis is seen in almost half of the cases. This is usually transient and primarily involves the large joints [4].

Vascular inflammation is not rare. It occurs in up to 40% of patients, with a male predominance. This occurs early in the disease with a median of 5 years after onset [5]. Venous disease is more frequent, usually presenting as lower extremity venous thrombosis while arterial involvement is associated with increased mortality, a common example being pulmonary artery aneurysm.

The present management of lower limb venous thrombosis in Behcet's Disease remains based on expert opinions [6].

Table 1: International Criteria for Behcet's Disease [3].

Symptoms	Points <sup>a</sup>
Oral Aphthosis	2
Genital Aphthosis	2
Ocular Lesion	2
Skin Lesion	1
Neurological Manifestation	1
Vascular Manifestation	1
Positive Pathergy Test	1 <sup>b</sup>

<sup>a</sup>Point score System: Scoring  $\geq 4$  indicates Behcet's diagnosis [3].

<sup>b</sup>Pathergy test is optional and the primary scoring system does not include pathergy testing. However, where pathergy testing is conducted one extra point may be assigned for a positive result [3].

Immunosuppression is the mainstay; however, the necessity of anticoagulation remains debatable.

## Case

A 38-year-old Caucasian male presented with cramping right leg pain while walking. It started a week previously as an 'arthritic' pain in his right knee that was worse in the morning. He denied prolonged immobility, leg trauma or history of previous thrombosis. He is known to have recurrent painful oral and scrotal ulcerations, intermittent right knee arthritis, as well as anterior uveitis for which he was taking paracetamol 1 g QID, Celecoxib 200 mg daily, and colchicine 500 mcg twice daily.

On examination his right knee has no clinical signs of inflammation with full range of motion. His right calf was tender, erythematous, and mildly swollen. Complete blood exam, renal function, and liver function were within normal range. His C-reactive protein was 70 mg/L (n 0-8 mg/l). A right leg ultrasound revealed deep venous thrombosis from the popliteal vein to femoral and external iliac veins. The common iliac vein was clear. On the basis of his history and lack of alternate cause on investigation he was considered to have Behcet's syndrome complicated by right leg DVT. He was given one dose of anticoagulant (enoxaparin 100 mg SC) upon presentation at the emergency department. He was subsequently managed with 1 mg/kg of prednisolone and Azathioprine after consultation with his rheumatologist and further anti-coagulation was not given.

## Discussion

Deep venous thrombosis is a common medical problem. It is due to combinations of at least two of the following: 1) vessel wall damage, 2) alteration of blood flow and, 3) abnormality in the coagulation cascade leading to hypercoagulability [7]. The mainstay of management is anticoagulation. The primary aim is to prevent pulmonary embolism, recurrence, and postphlebotic syndrome [8]. However, not all DVT needs anticoagulation. DVT as a manifestation of Behcet's syndrome is an exemption to the rule.

In Behcet's Syndrome pulmonary embolism is rare. It is commonly thought that thrombus is firmly adherent to the inflamed vascular wall [5,9,10] and most cases of pulmonary occlusion is due to local thrombus from underlying vasculitis rather than emboli from the leg [9].

The recurrence rate of DVT in Behcet's patient is 23.0% at 2 years and 38.4% at 5 years [11]. In the retrospective study done by Anh, et al. [10] comparing immunosuppressant and anticoagulant for prevention of DVT recurrence in patient with Behcet's syndrome. It showed that there is 75% recurrence rate in those managed with anticoagulant although the numbers were small (n = 4) compared with immunosuppressant alone (n = 16) and combination (n = 17) in which the recurrence rate was 12.5% and 5.9% respectively. Further clinical research with larger numbers is required to satisfactorily settle this question.

The pathogenesis of thrombosis in Behcet's syndrome is not fully understood. It has been postulated that endothelial cell dysfunction plays a major role in triggering an inflammatory cascade [12]. In the study done by Silingardi, et al. [13], it was concluded that there is no association found between factor V Leiden or prothrombin gene G20210A mutation and DVT in patients with Behcet's disease. However, it does not mean that we should not screen patient with Behcet's who presented with venous thrombosis for the first time for an inherited prothrombotic condition as they can co-exist, and if identified should be managed as a classic venous thrombosis with anticoagulation [14]. Anti-coagulation can pose a risk to patients with Behcet's as these patients are at risk of developing pulmonary arterial aneurysms [9]. Managing these patients with anticoagulant can be detrimental and regular screening for PAA is warranted.

## Conclusion

The primary purpose of anticoagulant therapy in classic DVT is prevention of thrombus progression and pulmonary embolism. In Behcet's Syndrome where the primary pathology is venous inflammation and the risk of thrombus progression and embolization is much less the use of immunosuppressant therapy is indicated. As such, there continues to be no proven role of anticoagulation in DVT as a manifestation of Behcet's disease in patients without inherited prothrombotic conditions. The risks would appear to out-weigh the benefits.

## References

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