

Superficial venous thrombosis in Behçet's disease: a case report and a review of the literature

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Abstract: Behçet's disease is a relapsing–remitting vasculitis that affects both arterial and venous vessels of various sizes. A 48-year-old man with Behçet's disease was seen at the outpatient clinic of our hospital for painful lesions on his left leg. Physical examination showed two tender erythematous linear indurations on the medial side of the left thigh and calf, of about 3 cm and 10 cm in length, respectively. Duplex ultrasonography revealed echogenic material in the lumen of collateral superficial veins, with no extension to the deep venous system. The patient was started on once-daily subcutaneous low-molecular-weight heparin in combination with elastic compression stockings, with spectacular improvement. Superficial venous thrombosis is a common manifestation of Behçet's disease, and the diagnosis is based on clinical features. Superficial venous thrombosis has a dual prognostic value: on one hand, it should raise the suspicion of concomitant deep venous and/or arterial involvement, and on the other hand, it may predict the development of future severe visceral manifestations.

Keywords: Behçet's disease, vasculitis, vascular involvement, venous thrombosis, superficial vein thrombosis

Introduction

Behçet's disease (BD) is a relapsing–remitting vasculitis that affects both arterial and venous vessels of various sizes. Recurrent bipolar ulcers and uveitis are the hallmark signs of the disease. However, other parts of the body may be affected, such as joints, vessels, brain, and gastrointestinal tract.¹ Herein, we present a case of multiple superficial venous thrombosis (SVT) as a relapsing form of BD. We then provide a review of the literature, which was performed to summarize the present state of knowledge on SVT in BD.

Case report

A 48-year-old man with BD was seen at the outpatient clinic of our hospital for painful lesions on his left leg. The patient's medical history included BD with both venous and arterial manifestations, for which he had received cyclophosphamide followed by azathioprine.

The diagnosis of BD had been made 17 years earlier based on recurrent oral and genital ulcers, typical cutaneous lesions, and both arterial thrombosis and deep venous thrombosis (DVT) of the lower extremities. Physical examination showed two tender erythematous linear indurations on the medial side of the left thigh and calf. The lesions were about 3 cm and 10 cm in length, respectively, and multiple varicosities were seen in both legs (Figures 1 and 2). Oral, genital, dermatological, and neurological

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examination showed no active signs of the disease. Duplex ultrasonography revealed echogenic material in the lumen of collateral superficial veins, with no extension to the deep venous system. The patient was started on once-daily subcutaneous low-molecular-weight heparin in combination with elastic compression stockings, with spectacular improvement.

Discussion

Vascular manifestations of BD are reported to occur in up to 40% of patients.² In our experience, vascular features are



Figure 1 Superficial venous thrombosis on the left thigh.



Figure 2 Superficial venous thrombosis on the left calf.

seen in 30% of cases.³ The most common scenario is lower extremity DVT. It usually occurs in the early clinical course of the disease, often within the first few years.²

SVT is the second most common vascular manifestation of BD.⁴ The prevalence of SVT in BD has been reported at rates ranging from 2.1% to 53.3% (Table 1),^{5–16} with a definite male predominance. Usually, it occurs early in the second decade of life.¹⁷ The classical presentation is an erythematous, painful, string-like lesion extending along a superficial vein, which usually occurs on the lower extremities. Physical examination reveals a palpable cord, which is erythematous, tender and warm, with a surrounding edema. Thus, the diagnosis is usually obvious, based on clinical features. SVT typically occurs in varicose veins (chronic venous insufficiency), but can occur in non-varicose veins. Duplex ultrasound is performed to assess the length of the thrombosis and to rule out a concomitant DVT.¹⁸

The greater saphenous vein and the small saphenous vein are by far the most affected segments.¹⁹ Sometimes, SVT occurs after a venipuncture or at the site of heparin injection, and thus it is considered as a vascular pathergy test.⁴ Some reports have shown that SVT may harbor an acquired or inherited thrombophilia such as positive anticardiolipin antibodies, but in the majority of cases, no triggering factor is found.²⁰ Usually, SVT resolves spontaneously over 2–4 weeks, but in some cases, a post-inflammatory skin hyperpigmentation may last for several months.²¹

As shown in Table 1, it seems that DVT is more common than SVT in BD. However, other studies have shown a more frequent occurrence of SVT over DVT.^{5,11} These conflicting data may be partly explained by the fact that some authors included SVT in dermatological manifestations of BD and, thus, patients with ST were excluded from studies reporting the prevalence of vascular features in BD. Moreover, as SVT resolves spontaneously within a few days, it may be under-

Table 1 Prevalence of SVT related to BD in the literature

Authors	Country	Sample size	Diagnosis criterion	Vascular involvement, n (%)	SVT, n (%)	DVT, n (%)
Sarica-Kucukoglu et al ⁵	Turkey	2,319	ISG	332 (14.3)	177 (53.3)	99 (29.8)
Duzgun et al ⁶	Turkey	180	ISG	71 (39.4)	48 (26.7)	104 (57.8)
Wu et al ⁷	China	766	ISG	93 (12.1)	4 (4.3)	78 (83.9)
Sagdiç et al ⁸	Turkey	129	ISG	52 (40.3)	4 (10)	29 (76)
Davatchi et al ⁹	Iran	7,187	Expert opinion	553 (9.1)	140 (2.3)	399 (6.6)
Boura et al ¹⁰	Greece	36	ISG	NA	5 (13.9)	NA
Mousa et al ¹¹	Kuwait	29	O'Duffy	10 (34.4)	7 (24.1)	3 (10.3)
Yücel et al ¹²	Turkey	231	ISG	37 (16)	37 (16)	NA
Pipitone et al ¹³	Italy	137	ISG	42 (30.7)	15 (10.9)	28 (21)
Balta et al ¹⁴	Turkey	521	ISG	30 (5.7)	11 (2.1)	16 (3.6)
Calamia et al ¹⁵	USA	13	ISG	3 (23)	1 (8)	2 (15)
Desbois et al ¹⁶	France	807	ISG	NA	26 (4.4)	560 (95.6)

Abbreviations: BD, Behçet's disease; DVT, deep venous thrombosis; ISG, International Study Group for Behçet's Disease; NA, not available; SVT, superficial venous thrombosis.

estimated in studies. Finally, SVT could be misdiagnosed as other dermatological features such as cutaneous polyarteritis nodosa and erythema nodosum.^{19,22}

In a retrospective study, Coskun et al found that erythema nodosum-like lesions and SVT could be predictive factors for visceral manifestations. They concluded that patients with those lesions should be closely monitored in terms of major organ involvement.¹⁷ Nevertheless, these findings should be validated in large prospective studies. Moreover, in a retrospective histopathological study, the presence of phlebitis in erythema nodosum-like lesions was associated with severe visceral involvement.²³ In another study, erythema nodosum was found to be a prognostic factor for the development of SVT.²⁴ In some cases, the clinical distinction between the two lesions would be challenging.¹⁹ SVT may occur in isolation or coexist concomitantly with DVT. Although there are no guidelines for the management of SVT associated with BD, anticoagulation therapy using enoxaparin has been used successfully in some reported cases.²⁵

Conclusion

SVT is a common manifestation of vascular BD. The diagnosis is primarily clinical, based on three main signs: erythema and tenderness in the distribution of a superficial vein, and the identification of a palpable cord. Duplex ultrasound should be performed on both the affected and contralateral limb to rule out a concomitant DVT.

SVT has a dual importance in prognosis. First, it should raise the suspicion of concomitant deep venous and/or arterial involvement. Second, it may predict the development of future serious visceral manifestations. Therefore, both short-term and long-term clinical monitoring should be performed in patients with SVT.

Ethics approval

Institutional approval was not required to publish this case report.

Disclosure

The authors report no conflicts of interest in this work.

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