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Severe Vascular Behçet Disease Presenting with Multisite Thrombosis and Pyoderma Gangrenosum in a Young Man: A Case Report

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Case Report

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Abstract

Background: Pyoderma gangrenosum is a rare neutrophilic dermatosis that can complicate Behçet disease. The simultaneous presentation of catastrophic multisite thrombosis and pyoderma gangrenosum as initial manifestations of Behçet disease is exceptionally rare. This case describes a young male with previously undiagnosed Behçet disease presenting with this unusual combination.

Case presentation: A 19-year-old man presented with painful necrotic leg ulcers evolving over 40 days. He later developed fever, cough, hemoptysis, and progressive thrombosis despite therapeutic anticoagulation. Thrombosis involved deep veins, mesenteric vein, and hepatic artery. Oral aphthae appeared on day 16 of hospitalization. Autoimmune and thrombophilia workups were negative including antiphospholipid antibodies. A clinical diagnosis of Behçet disease was established. The patient received pulse methylprednisolone, nine plasma exchange sessions, and cyclophosphamide. His ulcers healed completely after six cyclophosphamide cycles with no recurrence at follow-up.

Conclusions: This case presents a rare instance of pyoderma gangrenosum as the initial manifestation of Behçet disease with catastrophic multisite thrombosis. Negative antiphospholipid antibodies distinguished this presentation from catastrophic antiphospholipid syndrome. This case raises the possibility that pyoderma gangrenosum in Behçet disease may associate with a more severe thrombotic phenotype. Further studies are needed to determine this association, especially in the absence of classic mucocutaneous manifestations at presentation.

Introduction

Behçet's disease (BD) represents a chronic, relapsing vasculitis involving multiple organ systems and affecting blood vessels of all dimensions(1). The condition is most common along the ancient "Silk Road," with typical onset between 20 and 30 years of age(2, 3). Oral ulceration represents the most frequent initial manifestation, occurring in over 98% of patients(2, 4). Vascular involvement can occur in up to 50% of patients with Behçet syndrome(5). Venous manifestations are reported in 7% to 29% of patients, with deep vein thrombosis being most frequent. Arterial lesions occur less commonly(1.6%), manifesting predominantly as in situ thrombosis and aneurysms(6). A unique feature of BD is the coexistence of venous thrombosis and arterial aneurysms(1).

Pyoderma gangrenosum (PG) is a rare neutrophilic dermatosis that can complicate Behçet disease. The diagnosis of PG remains primarily clinical, supported by typical ulcer morphology and exclusion of other causes. Reported cases describe PG lesions in BD patients as painful, destructive ulcers with irregular, ragged overhanging edges.(7, 8).

The simultaneous presentation of catastrophic multisite thrombosis and pyoderma gangrenosum as initial manifestations of Behçet disease is exceptionally rare. This case describes a young male with previously undiagnosed BD presenting with this unusual combination.

Case presentation

A 19-year-old man with no known past medical history presented in late September 2024 with painful, progressively enlarging ulcerative lesions on both lower legs. The lesions had evolved over approximately 40 days and had become necrotic. One week before admission, he developed fever, chills, and productive cough. He denied dyspnea, pleuritic chest pain, hemoptysis, arthralgia, ocular or oral dryness, malar rash, photosensitivity, neurologic symptoms, or gastrointestinal or urinary complaints. His past history was notable for occasional oral aphthae. Family history revealed five miscarriages in one of his sisters.

On initial examination, the patient appeared ill and febrile. Vital signs included blood pressure 148/91 mmHg, heart rate 100 bpm, respiratory rate 20/min, temperature 38.2°C, and oxygen saturation 97% on room air. Dermatologic examination

revealed multiple necrotic ulcers on both legs, the largest measuring approximately 4×3 cm on the anterior left shin (Fig. 1). No active discharge was noted. Other systemic examinations were unremarkable.

Initial laboratory evaluation showed leukocytosis, thrombocytopenia, and markedly elevated inflammatory markers (Table 1). Liver function tests revealed significant elevations in transaminases and alkaline phosphatase, while total and direct bilirubin levels remained within normal limits. Abdominal ultrasonography demonstrated splenomegaly (135 mm).

Table 1

Key laboratory findings during hospitalization and 6-month follow-up. Laboratory parameters were measured at presentation (Day 0), during hemoptysis-associated intensive care unit (ICU) admission, after initiation of immunosuppressive therapy, at discharge, and at the 6-month follow-up. The table demonstrates the temporal evolution of inflammatory markers, hematologic indices, coagulation parameters, and biochemical markers of tissue injury throughout the disease course and in response to treatment.

Time point	Laboratory test	Result	Reference range	Time point	Laboratory test	Result	Reference range
Day 0 (Presentation)	WBC (/μL)	13800	4–10 ×10³	Day 14 (After immunosuppressive therapy initiation)	ESR (mm/hr)	1	< 20
	Hb (g/dL)	14.3	12–16		Ferritin (ng/mL)	573	30–400
	PLT (/μL)	90000	150–450 ×10³		Triglyceride (mg/dL)	117	< 150
	ESR (mm/hr)	85	< 20	Day 25 (Cyclophosphamide initiation / steroid adjustment)	PLT (/μL)	60000	150–450 ×10³
	AST (U/L)	357	< 40	Day 38 (Discharge)	WBC (/μL)	10000	4–10 ×10³
	ALT (U/L)	557	< 40		Hb (g/dL)	11.1	12–16
	ALP (U/L)	473	44–147		PLT (/μL)	350000	150–450 ×10³
	Viral markers (HBsAg, anti-HCV, HIV Ab)	Negative	—		AST (U/L)	34	< 40
	Blood culture (BacT/ALERT)	Negative	—		ALT (U/L)	178	< 40
Day 4 (ICU-Hemoptysis)	WBC (/μL)	14200	4–10 ×10³		ALP (U/L)	481	44–147
	Hb (g/dL)	11.2	12–16		LDH (U/L)	753	140–280
	PLT (/μL)	40000	150–450 ×10³	6-month follow-up	WBC (×10³/μL)	5.2	4–10
	Procalcitonin (ng/mL)	6.01	< 0.1		Hb (g/dL)	12.8	12–16
	Interleukin-6 (pg/mL)	719	< 7		PLT (×10³/μL)	274	150–450
	PT (sec)	19.4	11–14		ESR (mm/hr)	13	< 20
	PTT (sec)	85	25–35		AST (U/L)	17	< 40
	INR	1.5	≈ 1		ALT (U/L)	25	< 40
	LDH (U/L)	1316	140–280		ALP (U/L)	286	44–147
					LDH (U/L)	281	140–280

Further diagnostic evaluations, including infectious, hematologic, and rheumatologic workups, were performed. A dermatology consultation was obtained; the cutaneous lesions were considered clinically consistent with pyoderma gangrenosum, though a biopsy was not performed due to the patient's lack of consent. Peripheral smear and bone marrow aspiration showed no evidence of hematologic malignancy.

Comprehensive autoimmune evaluation, including ANA, anti-dsDNA, anti-liver kidney microsomal antibody (LKM), anti-mitochondrial antibody (AMA M2), p-ANCA, and c-ANCA, was negative. C3 and C4 levels were within normal limits. Antiphospholipid antibody panel (lupus anticoagulant, anti-cardiolipin IgG/IgM, anti-β2 glycoprotein I IgG/IgM) was negative. Infectious workup, including blood cultures, HIV, HBV, and HCV, was negative.

During hospitalization, the patient developed non-massive hemoptysis. Chest computed tomography (CT) revealed ground-glass opacities, raising initial concern for diffuse alveolar hemorrhage (Fig. 2). Concurrently, fever rose to 40°C, and peripheral smear demonstrated toxic granulation.

With worsening clinical status, marked elevations in inflammatory and tissue injury markers including lactate dehydrogenase (LDH), erythrocyte sedimentation rate (ESR), procalcitonin (PCT), and interleukin-6 (IL-6) were observed (Table 1). The patient was transferred to the intensive care unit (ICU), and empiric broad-spectrum antibiotics (vancomycin 1 g every 12 hours and meropenem 1 g every 8 hours) were initiated. Blood culture was obtained and remained negative. A single session of plasma exchange was performed.

Despite persistent fever, echocardiography was performed to evaluate for endocarditis; left ventricular ejection fraction was normal, and no pathological findings were reported. Due to ongoing hemoptysis, CT pulmonary angiography was performed and demonstrated pulmonary embolism with evidence of infarction in the lungs, spleen, and liver. Intravenous heparin infusion was initiated for anticoagulation.

Despite this therapy, the patient subsequently developed deep vein thrombosis in both upper and lower extremities, as well as thrombosis of the superior mesenteric vein (SMV) and the hepatic artery. Repeat autoimmune evaluation, including the ENA panel, remained negative. Thrombophilia workup, including antithrombin III, protein C, protein S, and factor II (prothrombin), was within normal limits. Antiphospholipid antibody panel was sent and subsequently returned negative.

Subsequently, with procalcitonin declining to 2.19 ng/mL and no identifiable infectious source, the infectious disease service discontinued antibiotics. Given the history of oral aphthae, multiple thromboses, and normal thrombophilia workup, catastrophic antiphospholipid syndrome (CAPS) and Behçet disease were considered, and the patient was transferred to the rheumatology service. Repeat ENA panel remained negative. Pulse methylprednisolone (1 g daily for 5 days) was initiated. Due to concern for CAPS, the patient underwent nine daily sessions of plasma exchange, followed by high-dose dexamethasone (8 mg every 8 hours).

On follow-up laboratory testing, the erythrocyte sedimentation rate (ESR) decreased to 1 mm/hr. Despite normalization of ESR, ferritin was 573 ng/mL; however, no evidence of hemophagocytic lymphohistiocytosis (HLH) was found (Table 1). On day 16 of hospitalization, overt oral aphthae were observed on repeat oral examination (Fig. 1). Ophthalmologic evaluation was normal, and HLA-B51 testing was negative. A pathergy test was performed and turned out negative.

Given the multiple thromboses, cutaneous lesions clinically consistent with pyoderma gangrenosum, and oral aphthae, a diagnosis of Behçet disease was established. On day 25 of hospitalization, cyclophosphamide (500 mg) was initiated. Due to thrombocytopenia, dexamethasone was transitioned to prednisolone 50 mg daily (Table 1). After starting prednisolone, the patient developed knee pain and effusion; MRI demonstrated evidence of infarction, and the prednisolone dose was reduced to 30 mg daily.

A second dose of cyclophosphamide (500 mg) was administered 12 days after the first dose. Subsequently, respiratory symptoms, hemoptysis, oral aphthae, and cutaneous lesions improved significantly. Anticoagulation was transitioned to warfarin, and the international normalized ratio (INR) was maintained within the therapeutic range of 2.5 to 3.5.

With improvement in clinical status, resolution of thrombocytopenia, normalization of liver function tests, and reduction in lactate dehydrogenase (LDH) (Table 1), the patient was discharged on the following medications: azathioprine 50 mg daily, prednisolone 30 mg daily, colchicine 1 mg daily, alendronate, calcium–vitamin D supplementation, and monthly cyclophosphamide (1 g).

Follow up

After six cycles of cyclophosphamide, the patient reported no new cutaneous lesions, arthralgia, morning stiffness, oral aphthae, genital ulcers, or hemoptysis. The necrotic ulcers of the lower limbs had healed, leaving only residual scarring (Fig. 1). Cardiac evaluation for pulmonary hypertension showed a normal left ventricular ejection fraction and a pulmonary artery pressure of 25 mmHg (borderline elevation, but no clinical evidence of significant pulmonary hypertension). Abdominal vascular Doppler ultrasonography was unremarkable, with no evidence of residual thrombosis. The patient remained clinically stable and was maintained on prednisolone 5 mg every 12 hours, azathioprine 50 mg every 12 hours, colchicine 1 mg daily, folic acid 5 mg daily, and warfarin adjusted according to PT/INR.

Timeline

Time	Clinical Course
Day – 40	Onset of ulcerative skin lesions on the lower extremities
Day – 7	Development of systemic symptoms, including fever, Respiratory distress
Day 0 (Admission)	Hospitalized because of worsening systemic symptoms and progressive necrotic skin lesions. Laboratory tests revealed leukocytosis, thrombocytopenia, splenomegaly and abnormal liver function tests. Clinical diagnosis of pyoderma gangrenosum was made. Hematologic malignancy and autoimmune diseases were excluded.
Day 4	Clinical deterioration with high-grade fever, non-massive hemoptysis, and markedly elevated inflammatory markers. Peripheral blood smear showed toxic granulation. The patient was admitted to the ICU.
Days 4–13	Broad-spectrum antibiotics and one session of plasma exchange were administered. CT pulmonary angiography demonstrated pulmonary embolism with venous and arterial thrombosis, despite anticoagulation, multiple arterial and venous thromboses developed. Coagulation disorders and Rheumatologic disorders were excluded.
Day 13	Clinical stabilization following intensive care management.
Day 16	Transferred to the rheumatology service. Pulse intravenous methylprednisolone and serial plasma exchange were initiated for suspected catastrophic antiphospholipid syndrome and Behçet disease. Oral aphthous ulcers became clinically evident.
Day 25	Behçet disease was diagnosed. Cyclophosphamide was initiated, and corticosteroid therapy was continued.
Day 38	Marked improvement in respiratory symptoms and cutaneous lesions. The patient was discharged in stable condition.

Discussion

This report describes a 19-year-old man who had Behçet's disease (BD) presenting with pyoderma gangrenosum leg ulcers and catastrophic multisite thrombosis. Despite receiving therapeutic anticoagulation, the patient experienced progressive thrombosis affecting both arteries and veins, a presentation that resembled catastrophic antiphospholipid syndrome.

Catastrophic antiphospholipid syndrome (CAPS) represents an uncommon, life-threatening disorder defined by recurrent vascular occlusions together with sustained antiphospholipid antibodies. The rapid emergence, elevated death rate, and reliance on classification criteria in clinical settings all complicate the diagnosis of CAPS.(9). Definite CAPS classification requires the presence of antiphospholipid antibodies on two or more occasions, at least 12 weeks apart. Our patient had negative antiphospholipid antibody panels on repeated testing, effectively excluding CAPS. According to the 2023 ACR/EULAR classification criteria for APS, diagnosis incorporates six clinical domains alongside two laboratory domains. (10). CAPS predominantly affects small vessels, leading to microangiopathic multiorgan failure within a short period(9). By comparison, Behçet's disease (BD) is a chronic form of vasculitis capable of involving both veins and arteries across all

vessel sizes(11). Venous thrombosis in BD is driven primarily by inflammation rather than traditional hypercoagulable mechanisms(12). Anticoagulation using heparin followed by warfarin represents the primary treatment approach for thrombotic events in APS.(13). For BD-related thrombosis, immunosuppressants are the cornerstone of therapy, and adding anticoagulation remains controversial(12). Our patient's negative serology, disease course exceeding one week, and response to immunosuppression favored Behçet over CAPS.

The International Criteria for Behçet's Disease (ICBD) provides a standardized scoring system for diagnosis. A total score of 4 or more points classifies a patient as having Behçet disease. The ICBD criteria demonstrate significantly higher sensitivity (94.8%) compared to the older International Study Group criteria (85.0%)(14). This improved sensitivity is particularly notable in patients with severe vascular manifestations(15). Our patient achieved a score of 4 points based on oral aphthosis (2 points), vascular thrombosis (1 point), and pyoderma gangrenosum skin lesions (1 point). The pathergy test was negative in our patient, but ICBD criteria do not require a positive result for diagnosis(14). Thus, our patient meets the validated international diagnostic threshold for Behçet disease.

Pyoderma gangrenosum (PG) is diagnosed primarily on clinical grounds, as biopsy findings are non-specific and pathergy risk limits its utility. A retrospective study of 54 PG patients found that biopsy had limited diagnostic promise due to low sensitivity and lack of specific findings(16). PG appears to be overrepresented among patients with Behçet disease (BD) compared with the general population. Among 1462 BD patients in a French cohort, only 6 cases (0.4%) of PG were identified, giving a prevalence of approximately 400 per 100,000 BD patients. All reported PG lesions in BD patients occurred on the limbs(17). The association between PG and BD is considered exceptional, particularly in pediatric populations(18). The most common location for pyoderma gangrenosum lesions is the lower extremities, particularly the pretibial region. In patients with Behçet disease specifically, all reported pyoderma gangrenosum cases occurred on the limbs(17, 19). In our case, the patient declined biopsy consent, but clinical features supported PG diagnosis. The rarity of PG in BD (0.4%) makes our patient's presentation with PG as the initial manifestation noteworthy.

Lower extremity vein thrombosis is the predominant form of vascular involvement, which affects forty percent of Behçet's syndrome patients and greatly raises mortality risk. Recent retrospective clinical studies have documented male-to-female ratios between 3.86:1 and 4.4:1, indicating a strong male predominance of vascular involvement in Behçet's disease.(20, 21). In a retrospective study of 796 BD patients, vascular involvement was observed in 12.8% of cases. Of these patients, 70.6% presented with venous lesions and 54.9% with arterial lesions, while simultaneous venous and arterial involvement occurred in 25.5%.(21). A Japanese cohort study found arterial and venous lesions in 31% and 81% of affected patients, respectively, including 12% with both types(22). Our patient developed extensive thrombosis including deep vein thromboses, mesenteric vein thrombosis, and hepatic artery thrombosis, representing simultaneous arterial and venous involvement.

In BD, excessive neutrophil activation and related pathways directly drive endothelial dysfunction, platelet activation, and thrombus formation. The pathogenesis of thrombosis in this condition is mediated by an altered immune-inflammatory response, not by conventional cardiovascular risk factors. Consequently, managing thrombosis in BD depends primarily on immunosuppressive agents rather than on anticoagulant therapy.(5). A phase 2 randomized controlled trial compared infliximab versus cyclophosphamide for severe BD. Among 52 patients (71% with vascular involvement), complete response at week 22 was 81% for infliximab versus 56% for cyclophosphamide. Adverse events occurred in 30% of infliximab patients compared to 64% receiving cyclophosphamide. The posterior probability of achieving ≥ 70% complete response was 97.4% for infliximab and 6.0% for cyclophosphamide. Our patient received cyclophosphamide with good clinical response, though infliximab may offer superior outcomes(23).

A case of juvenile BD with bilateral optic neuritis showed no improvement with methylprednisolone alone. Serum plasmapheresis was performed, resulting in improved clinical outcomes, and the patient was discharged on azathioprine maintenance therapy. Our patient received nine plasma exchange sessions due to initial CAPS concern, which may have contributed to disease control(24).

A systematic review and meta-analysis examined thrombosis relapse rates in Behçet's syndrome. Combined immunosuppressives and anticoagulants had a pooled relapse rate of 17.7% (95% CI, 9.7%-30.1%). Immunosuppressives alone had a relapse rate of 43.7%, while anticoagulants alone had 54.9%. Combination therapy was associated with significantly lower relapse risk compared to either treatment alone. Our patient was discharged on azathioprine, colchicine, prednisolone, and warfarin(25). After six cyclophosphamide cycles, our patient's necrotic leg ulcers healed with only residual scarring. He reported no new cutaneous lesions, oral aphthae, or hemoptysis at follow-up. Pulmonary artery pressure was borderline at 25 mmHg without clinical evidence of significant pulmonary hypertension. In young patients with pyoderma gangrenosum, unexplained thrombosis, and oral ulcers, clinicians should suspect Behçet's disease early(5, 26). Anticoagulation alone fails because inflammation drives thrombosis in Behçet's syndrome(5, 25). Immunosuppression with glucocorticoids and cyclophosphamide or infliximab is the cornerstone of therapy(23).

Conclusion

This case presents one of the rare instances of pyoderma gangrenosum as the initial manifestation of Behçet's disease in a young male patient with catastrophic multisite thrombosis. Our patient had negative antiphospholipid antibodies on repeated testing, distinguishing this presentation from catastrophic antiphospholipid syndrome. Treatment with glucocorticoids and cyclophosphamide effectively controlled both cutaneous lesions and thrombotic events, while anticoagulation alone was insufficient. The extensive arterial and venous thrombosis observed in this patient, despite therapeutic anticoagulation, highlights the importance of considering Behçet's disease as a potential underlying diagnosis in young patients with unexplained widespread thrombosis.

Abbreviations

ACR
American College of Rheumatology
AMA
Anti-Mitochondrial Antibody
ANA
Anti-Nuclear Antibody
anti-dsDNA
Anti-Double Stranded DNA Antibody
anti-β2GPI
Anti-Beta-2 Glycoprotein I Antibody
APS
Antiphospholipid Syndrome
BD
Behçet's Disease
c-ANCA
Cytoplasmic Anti-Neutrophil Cytoplasmic Antibody
CAPS
Catastrophic Antiphospholipid Syndrome
C3
Complement Component 3
C4
Complement Component 4
CT
Computed Tomography
ENA

Extractable Nuclear Antigen
ESR
Erythrocyte Sedimentation Rate
EULAR
European Alliance of Associations for Rheumatology
HBV
Hepatitis B Virus
HCV
Hepatitis C Virus
HIV
Human Immunodeficiency Virus
HLA-B51
Human Leukocyte Antigen B51
HLH
Hemophagocytic Lymphohistiocytosis
ICU
Intensive Care Unit
IL-6
Interleukin 6
INR
International Normalized Ratio
LDH
Lactate Dehydrogenase
LKM
Liver-Kidney Microsomal Antibody
MRI
Magnetic Resonance Imaging
p-ANCA
Perinuclear Anti-Neutrophil Cytoplasmic Antibody
PCT
Procalcitonin
PG
Pyoderma Gangrenosum
PT
Prothrombin Time
SMV
Superior Mesenteric Vein

Declarations

Ethics approval and consent to participate:

Written informed consent was obtained from the patient for publication of this case report. Ethical approval was obtained from the Shahid Sadoughi University Ethics Committee, approval code: IR.SSU.MEDICINE.REC.1404.330.

Consent for publication:

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal upon request. Availability of data and

materials:

Competing interests:

The authors declare that they have no competing interests.

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Author Contribution

Hamidreza Bashiri supervised the case and managed the patient. Nastaran Tavangar wrote and edited the manuscript and prepared the charts and figures. Elham Sadat Tabatabaei performed data collection and verification. All authors read and approved the final manuscript.

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Figures



Figure 1

Clinical manifestations during hospitalization and follow-up. (a) Necrotic ulcerative lesions on the lower extremities with violaceous undermined borders, clinically diagnosed as pyoderma gangrenosum, during hospitalization. (b) Residual atrophic scarring of the previous skin lesions at follow-up. (c) Oral aphthous ulcer observed during hospitalization.

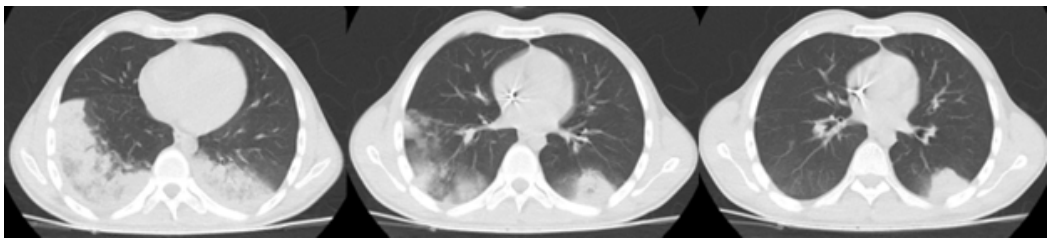


Figure 2

Chest computed tomography findings during hemoptysis. Chest computed tomography images obtained at the time of hemoptysis, demonstrating representative axial sections with ground-glass opacities, consistent with pulmonary involvement.