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CASE REPORT

A case of Behçet's disease associated with necrotizing small vessel vasculitis

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Abstract Cutaneous manifestations are an important feature of Behçet's disease (BD) and are classified as a major diagnostic criterion by the International Study Group (ISG). Necrotizing vasculitis as a skin manifestation in patients with BD has been reported rarely. In this report, we describe a patient who fulfills ISG criteria for the diagnosis of BD with necrotizing small vessel vasculitis developed necrosis on distal part of the fifth finger of right foot and the heel.

Keywords Behçet's disease · Necrotizing vasculitis

Introduction

Behçet's disease (BD) is a systemic vasculitis of unknown etiology, characterized by recurrent oral and genital ulcers and uveitis. Cutaneous, articular, neurologic, intestinal, pulmonary, urogenital, and vascular manifestations have also been observed. It can affect all types and sizes of vessels [1]. Cutaneous manifestations are an important feature of BD and are classified as a major diagnostic criterion by the International Study Group (ISG) [2]. A number of cutaneous manifestations, including Sweet's syndrome-like lesions, erythema multiforme-like lesions, infiltrated erythema, palpable purpura, hemorrhagic bullae, extragenital ulcerations, superficial migratory thrombophlebitis and acral purpuric papulonodular lesions, are described [3, 4]; however, some of these are uncommon. Cutaneous vasculitis in BD is predominantly a venulitis or thrombophlebitis, with relative sparing of the arterial compartment [5]. Necrotizing vasculitis as a skin manifestation in patients with BD has also been reported rarely [6–11]. In this report, we describe a BD patient with necrotizing small vessel vasculitis developed necrosis on distal part of the fifth finger of right foot and the heel.

Case report

A 21-year-old woman was referred with a 2-year history of recurrent oral and genital ulcers, erythema-nodosum like lesions and bilateral uveitis. Based on the clinical findings, a diagnosis of BD was established. After diagnosis of BD, treatment was started with colchicine 1.5 mg/day, azathioprine 150 mg/day, cyclosporine 200 mg/day and low dose aspirin. One year later, she was re-admitted to the hospital with recurrent uveitis of left eye, erythema-nodosum like lesions and ischemic lesions on the fifth finger of right foot and the heel. Laboratory studies revealed white blood cell count of 11,400 per mm^3 [4,400–11,300 per mm^3], platelet count of 559,000 per mm^3 [150,000–450,000 per mm^3], C-reactive protein of 142 mg/l [<5 mg/l], and erythrocyte sedimentation rate of 106 mm/h. Urinalysis, renal function, and liver-function tests were normal. Serological work up was negative, including antinuclear antibody, antineutrophilic-cytoplasmic antibody, lupus anticoagulant, anticardiolipin antibody

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and rheumatoid factor. Genetic study including factor V Leiden and prothrombin 20210A gene mutations were also negative. Protein-C, protein-S and antithrombin III levels were in normal ranges. Thorax computed tomography was normal. Although a histopathological examination was not done, based on testing for the primary causes of hypercoagulopathy yielding negative results, clinical diagnosis of small vessel vasculitis with BD was made and therapy was initiated with intravenous pulse of methylprednisolone 1 g given three consecutive days and monthly pulse cyclophosphamide. Later, prednisolone continued with 1 mg/kg/day orally. Thereafter, intravenous three monthly pulses of cyclophosphamide (1 gram once a month) were given, oral cyclophosphamide continued with 150 mg/day. Prednisolone was dropped below 20 mg/day over a 12-week period. Despite appropriate immuno-suppressive therapy, necrosis developed on distal part of the fifth finger of right foot and the heel after 5 months (Fig. 1). Amputation of distal part of the fifth finger of right foot (Fig. 2) and debridement of necrotic site of the heel were done and split thickness skin grafting was used for repair of soft-tissue defect resulting from necrotic site of the heel. Preoperative laboratory studies revealed white blood cell count of 11,300 per mm^3, platelet count of 199,000 per mm^3, C-reactive protein of 15.3 mg/l, and erythrocyte sedimentation rate of 48 mm/h. She is currently free of all presenting symptoms in the 8 months of this therapy. Soft-tissue defect on the heel has been reduced.

Discussion

BD is distinctive among the vasculitides in that it can affect all types and sizes of vessels. Large vessel involvement is observed in 25–50% of patients with BD [12, 13]. When we review of the literature, some

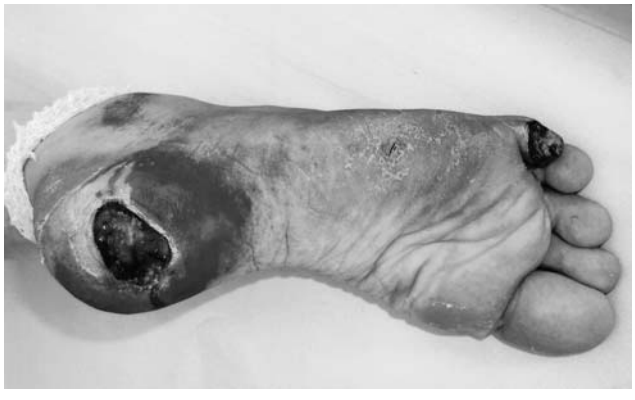

Fig. 1 Necrosis on the distal part of fifth finger of right foot and the heel

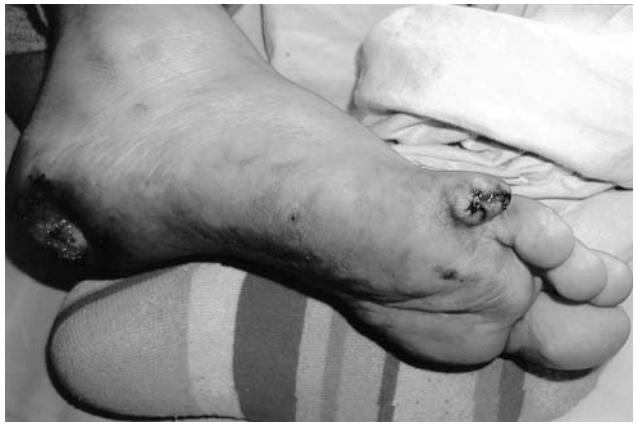

Fig. 2 Amputation of distal part of the fifth finger of right foot

cases of BD with severe necrotizing vasculitis as a skin manifestation have been described [6–11]. Cutaneous vasculitis in BD is predominantly a venulitis or thrombophlebitis, with relative sparing of the arterial compartment. Chen et al. [5] reported that approximately half (48%) of BD patients with cutaneous lesions had either lymphocytic (31%) or leukocytoclastic vasculitis (17%). They have suggested that vascular inflammation is the pathologic basis of the skin lesions in BD and that the histologic spectrum ranges from fully developed necrotizing vasculitis with marked fibrinoid necrosis of vessel walls to perivascular inflammation with or without a marked interstitial infiltrate. Plotkin et al. [7] reported that a patient with chronic recurrent migratory superficial thrombophlebitis and marked cutaneous hyperreactivity (pathergy) who developed leukocytoclastic vasculitis with recalcitrant leg ulcerations 9 years after the onset of his illness. Cutaneous polyarteritis- nodosa-like lesions and necrotizing panarteritis involving small and medium-sized arteries in the dermis–subcutis junction have also been reported rarely with BD [10, 11]. Vikas et al. [10] reported that their patients had both venous and arterial involvement, the former with thrombotic angiopathy and the latter with acute vasculitis.

Although a histopathological examination was not done in our case, based on testing for the primary causes of hypercoagulopathy yielding negative results, clinical diagnosis of vasculitis with BD was made. Vasculitis is well-known characteristic feature of BD. Accordingly, we considered vasculitis with BD as the most likely diagnosis in this case. Our patient had no history of venous thrombosis before the development of necrotizing vasculitis. One interesting point of our patient is that, to the best of our knowledge, there is no previous report of such a case of BD with necrotizing small vessel vasculitis that developed necrosis on distal part of the fifth finger of right foot and the heel. The

association of BD and small vessel vasculitis is represent a rare condition. Our knowledge of the prevalence, pathogenesis and therapeutic options of small vessel vasculitis in BD patients are limited.

References

1. Yazıcı H, Yurdakul S, Hamuryudan V, Fresko I (2003) Behçet's syndrome. In: Hochberg MC, Silman AJ, Smolen JS, Weinblatt ME, Weisman MH (eds) Rheumatology. Mosby, London, pp 1665–1669
2. International Study Group for Behçet's Disease (1990) Criteria for diagnosis of Behçet's disease. Lancet 335:1078–1080
3. Magro CM, Crowson AN (1995) Cutaneous manifestations of Behçet's disease. Int J Dermatol. 34:159–165
4. King R, Crowson AN, Murray E, Magro CM (1995) Acral purpuric papulonodular lesions as a manifestation of Behçet's disease. Int J Dermatol 34:190–192
5. Chen KR, Kawahara Y, Miyakawa S, Nishikawa T (1997) Cutaneous vasculitis in Behçet's disease: a clinical and histopathologic study of 20 patients. J Am Acad Dermatol 36:689–696
6. Cornelis F, Sigal-Nahum M, Gaulier A, Bleichner G, Sigal S (1989) Behçet's disease with severe cutaneous necrotizing vasculitis: response to plasma exchange—report of a case. J Am Acad Dermatol 21:576–579
7. Plotkin GR, Patel BR, Shah VN (1985) Behçet's syndrome complicated by cutaneous leukocytoclastic vasculitis: response to prednisone and chlorambucil. Arch Intern Med 145:1913–1915
8. Lee SH, Chung KY, Lee WS, Lee S (1989) Behçet's syndrome associated with bullous necrotizing vasculitis. J Am Acad Dermatol 21:327–330
9. Zouboulis CC, Kurz K, Bratzke B, Orfanos CE (1991) Adamantiades-Behçet's disease: necrotizing systemic vasculitis with a fatal outcome. Hautarzt 42:451–454
10. Vikas A, Atul S, Singh R, Sarbmeet L, Mohan H (2004) Behçet's disease with relapsing cutaneous polyarteritis-nodosa-like lesions, responsive to oral cyclosporine therapy. Dermatol Online J 9:9
11. Liao YH, Hsiao GH, Hsiao CH (1999) Behçet's disease with cutaneous changes resembling polyarteritis nodosa. Br J Dermatol 140:368–369
12. Müftüoglu A, Yurdakul S, Yazici H, Tüzün Y, Pazarli H, Altuğ F (1986) Vascular involvement in Behçet's disease. A review of 129 cases. In: Lehner T, Barnes CG (eds) (1986) Recent advances in Behçet's disease. Royal Society of Medicine Services International Congress and Symposium Series no. 103, London, pp 255–260
13. Koç Y, Güllü İ, Akpek G, Akpolat T, Kansu E, Kiraz S, Batman F, Kansu T, Balkancı F, Akkaya S, Telatar H, Zileli T (1992) Vascular involvement in Behçet's disease. J Rheumatol 19:402–410