Behçet's syndrome with gangrenous bilateral lower limbs

Yathish Gattimallanahali, Gurmeet Mangat, Balakrishnan Canchi, Girish Kakade

Department of Rheumatology,
PD Hinduja Hospital and
Medical Research Centre,
Mumbai, Maharashtra, India

Correspondence to
Dr Yathish Gattimallanahali,
yathish_gc@yahoo.co.in

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DESCRIPTION

A 25-year-old Indian woman presented with recurrent oral and genital ulcers that had started in 2008. She had suffered from posterior uveitis that same year and been treated with oral prednisolone 40 mg/day tapered over 6 months, after which she recovered fully with complete restoration of vision without the need for any other immunosuppressive agents. In 2009, she developed acute onset gangrene of the right foot, which needed above-ankle amputation (figure 1). In 2011, she consulted a rheumatologist for a non-healing ulcer of the right thigh. A diagnosis of Behçet's syndrome was made and she was treated with prednisolone 1 mg/kg taper along with azathioprine 100 mg/day. She did not have a family history of Behçet's syndrome nor of any other autoimmune disorder. Over the next 6 months, there was complete resolution of the skin and orogenital ulcers. She was in remission until July 2015, on maintenance steroid and azathioprine, at which time she initially consulted us for acute onset gangrene of the left foot (figures 1 and 2). Physical examination revealed left foot gangrene with a clear line of demarcation and absent left dorsalis pedis and posterior tibial arterial pulsation.

Laboratory findings showed an increase in serum C reactive protein (71.16 mg/L). Rheumatoid factor, antinuclear antibodies, antineutrophil cytoplasmic antibodies (ANCA), antiphospholipid antibodies (APLA) and lupus anticoagulant were all negative. CT angiogram showed no flow in left anterior tibial and dorsalis pedis arteries (figure 3). The patient underwent above-ankle amputation of the left foot. Histopathology of the amputated foot showed arterial wall inflammation with thrombosis and inflammatory infiltrate in the dermis and subcutaneous tissue. An intravenous pulse of methylprednisolone 500 mg/day for 3 days was

Figure 2 Gangrenous left foot.

administered and a course of intravenous pulses of cyclophosphamide 500 mg every fortnightly was advised. At the patient's latest follow-up, she had received 6 pulses of intravenous cyclophosphamide and was on 10 mg prednisolone. The amputated stump had healed well and she was undergoing active physiotherapy and rehabilitation (figure 4).

Behçet's syndrome is a chronic, relapsing and debilitating systemic vasculitis of unknown aetiology, characterised by recurrent oral ulcers, genital ulcers, ocular and skin involvement, and other multisystem features. It can cause significant morbidity leading to functional impairment, disability affecting quality of life and mortality. There are no specific diagnostic tests for Behçet's syndrome, the diagnosis is made on the basis of the clinical picture, with typical features balanced by exclusion of other differential diagnoses. The differential diagnosis of a young patient presenting with gangrene of limbs is wide, and includes ANCA-associated vasculitis, polyarteritis nodosa, APLA syndrome and other procoagulant states. Careful exclusion of all these conditions with appropriate investigations is essential.

Genome-wide association studies (GWAS) have confirmed HLA-B51 association, however, the potential interactions of this allele with other genetic and non-genetic risk factors, and with disease phenotype, is yet to be confirmed.¹ In our

Figure 1 Amputated right foot and left foot gangrene with clear line of demarcation.

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Figure 3 CT angiography of lower limbs showing no flow in left anterior tibial and dorsalis pedis artery.

Figure 4 Patient undergoing active physiotherapy and rehabilitation with right foot prosthesis.

patient, who was already an established case of Behçet's syndrome with a proven vasculitis, HLA-B51 was not carried out due to financial constraints and also as it would not have changed the course of management.

Arterial involvement is rare and has been reported in about 1–12% of patients with Behçet's syndrome.² Any part of the arterial tree can be affected, with aneurysms or occlusions in the abdominal aorta, carotid, pulmonary, femoral and popliteal arteries. Disease seems more aggressive with high mortality in patients who have their disease onset at 14–24 years of age.³ With time, the disease seems to become less aggressive, with a smaller number of exacerbations and decreased mortality. Arterial involvement in Behçet's syndrome is very rare in the Indian population—reported only in a few cases. In spite of its rarity, it becomes very important to keep vasculitis as one of the differentials in any young patient with thrombotic events, as missing the diagnosis can lead to devastating complications.

The treatment of arterial involvement involves a combination of steroids and cyclophosphamide as a first-line therapy, progressing to infliximab in severe refractory and life-threatening cases. Surgical repair of peripheral arterial aneurysms, if indicated, should be performed after controlling the disease with adequate immunosuppression. Anticoagulation is not used unless there is a prothrombotic tendency in the subject and an absence of aneurysms.

Learning points

- ▶ Vasculitis in Behçet's syndrome can occur in spite of the patient being on immunosuppressive drugs.
- ▶ Vasculitis in Behçet's syndrome needs early diagnosis and aggressive management.
- ▶ Frequent monitoring is required, especially in young patients who have higher risk for vascular events.

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