Behçet's disease in a patient with immunodeficiency virus infection

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Abstract

A patient with human immunodeficiency virus (HIV) infection who developed Behçet's disease is described. As various vasculitis syndromes have been encountered recently in association with HIV infection it is suggested that Behçet's disease may be related to the HIV infection in this patient.

Behçet's disease is a multisystem disease that may have mucocutaneous, ocular, intestinal, articular, vascular, urogenital, pulmonary, neurological, and other features.¹⁻³ The cause of Behçet's disease is unknown, but viral agents and autoimmune mechanisms are currently suggested as important factors in its pathogenesis.⁴⁻⁵ We describe a case of Behçet's disease developing in a young black woman with HIV infection. As far as we know, this is the first such case reported.

Case report

A 27 year old black Ugandan woman presented with an eight month history of fatigue, fevers, night sweats, anorexia, photophobia, painful oral and genital ulcers, and painful pretibial nodules. She had also had three episodes of swollen and painful feet, ankles, knees, and hands, recurrent episodes of bilateral conjunctivitis, and pustules at venepuncture sites. Owing to her systemic symptoms and because she had immigrated from Uganda she was tested by an enzyme linked immunosorbent assay (ELISA) and found to have antibodies to HIV, confirmed by Western blot analysis. There was no history of urethritis, inflammatory bowel disease, or psoriatic skin lesions. On examination she appeared chronically ill with mild generalised lymphadenopathy. Oral hairy leucoplakia was noted. There was a marked bilateral pretibial erythema nodosum. Two large genital ulcers were found. The haemoglobin was 89 g/l (normochromic, normocytic), white blood cell count $2.6 \times 10^9/l$, and platelet count $463 \times 10^9/l$. An absolute CD4 count of 20/l (normal >420) and a T4/T8 ratio of 0.08 (normal 0.8-2.5) were found. Serological tests showed no hepatitis B virus. Serological markers for autoimmune disease were negative. Liver and kidney function tests were normal. Cultures from genital ulcers, stool blood, and urine were negative. Bone marrow aspirate was normal. Biopsy specimens of both oral and genital ulcers showed no evidence of malignancy or infectious agents. Human leucocyte antigen (HLA) typing showed A9, 30; B42, 17; Bw4/Bw6; DR3, DR8;

DRw52. There was no radiological evidence of sacroiliitis or spondylitis.

The patient was diagnosed as having Behçet's disease in association with HIV infection stage IV C2. Over the following three weeks the genital ulcers enlarged and deepened, and a new painful ulcer developed on the oral buccal mucosa. Despite a trial of oral acyclovir 1000 mg/day for 10 days she developed right nodular episcleritis, two large sacral ulcers, and the erythema nodosum recurred. She was treated with prednisone 40 mg/day and thalidomide, without improvement. Both drugs were discontinued and colchicine 1.2 mg/day was started, with marked improvement in two weeks and subsequent healing of the ulcers over the next four months. Treatment was subsequently started with zidovudine, and the patient was still asymptomatic at her eight month follow up.

Discussion

The patient described in this report fulfils the criteria for the diagnosis of Behçet's disease according to Mason and Barnes⁵ as well as O'Duffy.⁶ In addition to extremely painful recurrent oral and genital ulcers, she had conjunctivitis and nodular episcleritis, erythema nodosum, polyarthritis, and pathergy. She also had serological evidence of HIV infection, possibly related to two heterosexual contacts in an area with endemic HIV infection (Uganda). Although oral ulcers are associated with HIV infection, the presence of the genital ulcers and the other features confirm the co-occurrence of the two conditions. The differential diagnosis included Reiter's disease, which has recently been described in patients with HIV.^{7,8} Our patient had no clinical evidence of spondylitis or sacroiliitis, however, nor of urethritis or diarrhoea. She did not have features of any other spondyloarthropathy.

The cause of Behçet's disease is unknown, but viral agents or autoimmune mechanisms are currently suggested as possible important factors in its pathogenesis.^{3,4} Recent studies have shown an increase in herpes simplex virus type 1 homologous DNA from the nuclei of peripheral blood lymphocytes in patients with Behçet's disease.⁹ Furthermore, some patients with Behçet's disease have responded to acyclovir, whereas others, like our patient, have not.¹⁰⁻¹² Our patient has remained in remission while taking zidovudine, without any other anti-inflammatory drugs.

Various vasculitis syndromes have been found in association with HIV infection.^{13,14} The relation between vasculitis and HIV infec-

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tion remains unclear, but the HIV virus may have a direct effect on the vessel wall or might be participating through an immune complex mechanism.¹³ In view of the evidence of co-existence of HIV infection and vasculitis we suggest that Behçet's disease may be related to the HIV infection in this patient.

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