Relapsing polychondritis and Behçet’s syndrome in a patient with HIV infection

Relapsing polychondritis is a systemic disorder characterised by recurrent inflammatory lesions involving cartilaginous structures, the cardiovascular system, the eyes, and the ears. Connective tissue diseases, dysmyelo poetic and myeloproliferative syndromes, ulcerative colitis, and thyroid diseases have been reported in association with relapsing polychondritis.1 We present a patient with coexisting relapsing polychondritis, Behçet’s syndrome, and infection with the human immunodeficiency virus (HIV).

A 25 year old man presented with a six month history of recurrent oral and genital ulcerations, followed by redness of his left eye and generalised polyarthritis. There was a past history of intravenous drug abuse and hepatitis B infection. Two months later he developed pain and swelling of his left pinna and nasal cartilage, and necrotising skin lesions on his legs, fingers, and toes. On examination the left pinna was erythematous, swollen, and tender. Several ulcers were noted on the buccal mucosa and penis. There were signs of uveitis in the left eye and necrotising skin lesions suggesting vasculitis (fig 1). The characteristic saddle nose deformity was evident. Radiographs of the hands and knees were normal. Laboratory findings included: erythrocyte sedimentation rate 23 mm/1 h, haemoglobin 100 g/l, leucocyte count 10-4 x 10^9 with a normal differential count, platelet count 77 x 10^9/l. Tests that gave negative or normal results included serum creatinine, urine analysis, total haemolytic complement, bilirubin, transaminases, alkaline phosphatase, hepatitis B surface antigen, hepatitis B surface antibody, Veneral Disease Research laboratory tests, rheumatoid factor, and antinuclear antibodies. Hepatitis B core antibody and serology for HIV performed by enzyme linked immunosorbent assay were positive. The number of CD4 positive lymphocytes was 350/mm^3, and a biopsy specimen from the left pinna showed diffuse loss of basophilic staining of matrix (fig 2). Despite treatment with prednisone 60 mg/day and dapsone 100 mg/day, buccal, genital, and skin lesions persisted and methotrexate 15 mg/week was added; it stopped four months later because of lack of efficacy. During the next two years, while receiving prednisone 5-30 mg/day and azithromycin (AZT) 600 mg/day, the patient developed severe flares of oral and genital ulcers, bilateral auricular chondritis, and oesophageal candidiasis, he eventually died from a pulmonary infection.

There is a broad spectrum of rheumatic manifestations associated with HIV infection;2 vasculitis is one of them.3 Relapsing polychondritis and Behçet’s syndrome are rare multisystem diseases in which vasculitis appears to be an important factor. The coexistence of the two conditions has been reported previously,4 but not in patients infected with HIV. Our patient fulfilled the diagnostic criteria proposed by McAdam for relapsing polychondritis (bilateral auricular chondritis, nasal chondritis, uveitis, and polyarthralgia) and by the International Study Group for Behçet’s disease (recurrent oral and genital ulcers, and uveitis).5 The relationship between HIV and connective tissue diseases is unknown. The development of the latter seems not to be affected by the stage of immunosuppression.4 It is possible that the HIV interacts with endothelial cells, leading to the release of chemotactic mediators, the development of vasculitis and, in this patient, relapsing polychondritis and Behçet’s syndrome. The five year estimated survival rate of patients with relapsing polychondritis and systemic vasculitis is 45%, similar to that reported for polyarteritis nodosa.6 In patients with autoimmune diseases infected with the HIV, each case must be judged carefully, weighing the risks of immunosuppressive therapy in an immunodeficient patient.7 AZT has not been reported to be effective. In our patient, aggressive therapy was warranted to control the severe vasculitis. Antimicrobial prophylaxis (pentamidine, butenosin; broad spectrum antibiotics) may be necessary for preventing infectious diseases.8


Human leucocyte antigen typing in rheumatoid arthritis/polymyositis overlap syndrome

Rheumatoid arthritis/polymyositis (RA/PM) overlap syndrome is an uncommon disorder.9 One study of 76 patients with myositis found no cases of RA.2 Halla et al first reported 'rheumatoid myositis', and noted that muscle necrosis was associated with mild synovitis.1 Non-erosive arthritis has been described in PM associated with the Jo-1 antibody.4 We describe five patients with erosive, seropositive RA, and electromyographically and biopsy proven PM, their autoantibodies and human leucocyte antigen (HLA) alleles.

Figure 1 Necrotising vasculitic lesions on the patient’s fingers.

Figure 2 Biopsy specimen of the left pinna, showing diffuse loss of basophilic staining of the matrix.