Case report

Arthritis associated with leucocytoclastic angiitis

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SUMMARY The case described is of a Negro male with recurrent ulcerating skin lesions and episodic arthritis. The association of arthritis with inflammatory dermatoses and a possible pathogenic mechanism for the arthritis are discussed.

Episodic arthritis is associated with a variety of ulcerating, nodular, acniform, or pustular skin lesions. The nosology of these inflammatory dermatoses is confusing, but they can be conveniently classified into 4 groups by clinical and histological criteria: Behçet’s disease, pyoderma gangrenosum, Sweet’s syndrome, and the family of leucocytoclastic angiitides. Behçet’s disease and pyoderma gangrenosum, while giving identical skin lesions, differ histologically from the other groups by the presence of a predominantly lymphocytic infiltration of the dermis and the scarcity of vasculitis or perivascular collections of neutrophils. The other two dermatoses, Sweet’s syndrome and the family of leucocytoclastic angiitides, show perivascular neutrophil infiltrate, but only in the latter is there fibrinoid necrosis of small vessels; it also differs clinically by the presence of ulcerating skin lesions which may scar. Both may be accompanied by fever during the acute episode, and they characteristically respond to corticosteroid therapy. Trentham et al. described a case with features of both these groups and argued that they were part of the disease spectrum. The case history given below lends weight to this argument.

Case report

A Negro male presented in 1972 at the age of 11 with recurrent painful swelling of the small joints of the hands and was found to have multiple circular scars on the legs and several subcutaneous nodules which ulcerated after a few days. A diagnosis of capillaritis was made on biopsy. Antibiotics and nonsteroidal anti-inflammatory drugs were ineffective. Two years later, because of recurrent synovitis of the elbows and ankles accompanied by fever, prednisolone 20 mg on alternate days was added.

Skin lesions, though less frequent, continued chiefly on the extensor surfaces of the limbs but also on the trunk and face (Fig. 1). In 1975 azathioprine to 100 mg daily was added without benefit. In 1978 after an influenza illness he had fluctuating synovitis, including both knees and more frequent skin lesions. Biopsy of one showed a normal epidermis, but vasculitis affecting small vessels, particularly at the dermo-epidermal junction, with fibrin thrombi and a largely polymorphonuclear leucocyte infiltration with leucocytoclastics in the surrounding dermis. Synovial biopsy showed proliferation of capillaries with a polymorphonuclear leucocyte and round cell infiltration. Synovial fluid contained 13·2 × 10⁷/l cells, 98% polymorphonuclear leucocytes. Roentgenograms of affected joints showed periarticular osteoporosis only. There was a mild microcytic hypochromic anaemia, a neutrophil leucocytosis during acute episodes, and an ESR persistently elevated above 50 mm in the first hour. IgG was 20·2 g/l, but other immunoglobulins were not altered. Renal function was normal, as were haemoglobin electrophoresis, culture and serological tests for pathogenic bacteria, viruses, and fungi, liver enzymes, pulmonary function tests, and complement, and C1q binding for circulating immune complexes negative. No new skin lesions appeared for 1 week following 1 g of intravenous methylprednisolone.
Discussion

Despite the clinical and histological heterogeneity of these dermatoses, the arthritis associated with them is similar. It is characteristically an episodic pauciarticular asymmetrical arthritis, particularly of the lower limb, and resolving with neither sequelae nor radiological evidence of joint damage. The pathogenesis of these dermatoses is unknown, as is the causation of the associated arthritis. However, the similarity of this arthritis with that complicating ulcerative colitis and Crohn’s disease is striking and suggests a common pathogenic mechanism. Absorption of antigenic material has been demonstrated in inflammatory bowel disease, as has the presence of circulating immune complexes, so it is of note that circulating immune complexes may also be detected in Behçet’s disease, pyoderma gangrenosum, and leucocytoclastic angitis.

References