

Case report

Idiopathic peripheral enthesopathy without spondylarthritis

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SUMMARY A 40 year old man who had a systemic inflammatory enthesopathy without spondylarthritis and HLA-B27 is described. The presence of hypergammaglobulinaemia and the effectiveness of glucocorticoid therapy suggested the possibility of its aetiology being 'autoimmune' in nature.

A 40 year old Japanese man had suffered from rheumatic fever in 1958 and received an aortic valve replacement for his rheumatic valvular disease in 1980. When he visited our rheumatology clinic in June 1985 he had polyarthralgia, low grade fever (37.8°C), and weight loss and had not been able to work for one month because of marked general body discomfort. There had been no trauma of any sort. A recurrence of rheumatic fever was considered initially because of the patient's past history and discontinuation of penicillin prophylaxis three years before.

Detailed physical examination of the joints, however, showed no objective signs of active synovitis. Instead, marked tenderness and radiating pains were demonstrated at the sites of entheses, where muscles are inserted into and originate from bones. The affected entheses were those of the right sternocleidomastoid muscle, the right deltoid muscle, both extensor digitorum communis muscles, the right biceps femoris muscle, and the right extensor digitorum brevis muscles. The pain at these entheses was aggravated by contraction against resistance of the relevant muscles, thus confirming that these pains originated in the enthesopathies. The sacroiliac joints and spine were normal on physical examination. The chest expansion was not limited. Neither skin rash nor subcutaneous nodules were found.

Laboratory tests showed haemoglobin 10.7 g/dl

(107 g/l), white blood cell count $5.0 \times 10^9/l$ (7% eosinophils), platelets $310 \times 10^9/l$, erythrocyte sedimentation rate (ESR) 124 mm/1st h, fibrinogen 10.2 g/l, IgG 24.1 g/l, IgA 6.24 g/l, IgM 0.94 g/l, CH₅₀ 58 U (normal 30-40 U), and C reactive protein (CRP) 5+. Tests for rheumatoid factors, antinuclear antibodies, LE cells, anti-DNA antibodies (Faust assay), precipitating autoantibodies, and circulating immune complexes (C1q binding) were all negative. The serum creatine kinase level was 33 IU/l (normal 27-85 IU/l). Repeated tests for β haemolytic streptococcal infection by throat cultures, antistreptolysin O, and antistreptokinase were also negative. HLA typing showed A2, 31(w19), B7, 40, Cw3, w7, DR1, w8, w52, DQw1, and w3. The histology of an

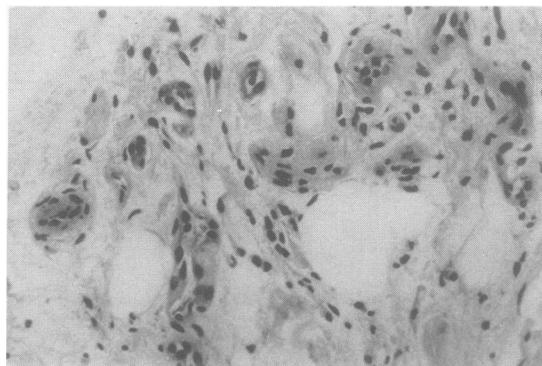


Fig. 1 Histology of the enthesis obtained from the site of origin of the right extensor digitorum communis muscle, showing slight degeneration of the collagen fibres and mild inflammatory infiltrate, especially around the capillaries.

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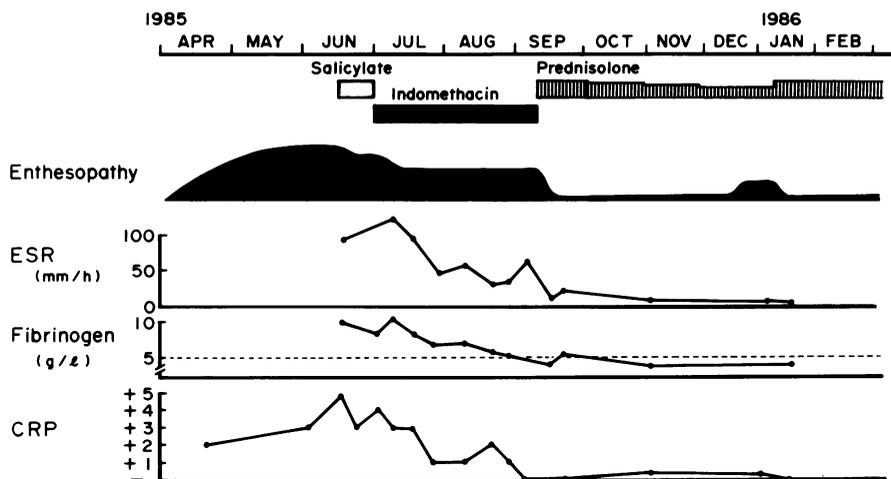


Fig. 2 Course of the clinical and laboratory findings.

enthesis removed by biopsy from the right extensor digitorum communis muscle showed a mild inflammatory infiltrate (Fig. 1). Radiographic examination of the peripheral and axial joints showed no abnormal signs such as erosion, calcific deposit, or bony overgrowth (enthesophyte).

Initially, the patient was treated with 3 g salicylate daily, which did not improve the symptoms significantly. Subsequent treatment with 150 mg of indomethacin a day was also not very effective. Finally, 10 mg of prednisolone a day produced dramatic improvement. Despite gradual reduction of prednisolone (1 mg a month) clinical flare occurred. He is now maintained with the initial dose. The clinical and laboratory course is illustrated in Fig. 2.

Discussion

Entheses are sites of tendon and ligament attachment to bone, and enthesopathy is a disease process which occurs at these sites.^{1,2} It may be inflammatory, degenerative, endocrine, metabolic, or traumatic in nature.³ It has been emphasised that inflammatory enthesopathy occurs frequently in HLA-B27 associated spondyloarthropathies, especially in ankylosing spondylitis.¹⁻⁵

Physical and radiographic examination showed that this patient had no involvement in his axial skeleton. The negative result for HLA-B27 was compatible with this case having a different enthesopathy from those in the various spondyloarthropathies or 'seronegative polyarthritides'.^{6,7} The

association of HLA-B27 with ankylosing spondylitis holds also in the Japanese population,⁸ though HLA-B27 itself is found very rarely in Japanese people.⁹

The marked systemic symptoms, the positive acute phase reactants, the presence of hypergammaglobulinaemia, the presence of inflammatory infiltrates in the fibrous tissue at the site of an enthesis, and the dramatic effectiveness of steroid therapy in this case all suggest the possibility that the condition of the patient is inflammatory in nature and 'autoimmune' in origin.

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