Case report

Relapsing polychondritis mimicking rheumatoid arthritis

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SUMMARY A woman with relapsing polychondritis presented with progressive and deforming polyarthritis (but always negative for rheumatoid factor) 14 years before the appearance of typical clinical and histological changes of nasal and auricular cartilage destruction.

Key words: nose deformity, ear deformity.

Case report

Relapsing polychondritis is a rather rare but severe systemic disease, characterised by recurrent episodes of inflammation and destruction mainly of cartilaginous structures. The principal destructions occur in the respiratory tract (larynx, tracheobronchial cartilage), sensory organs (ears, nose, and eyes), cardiovascular system (leading to aortic insufficiency, aortic and other arterial aneurysms, vasculitis), and peripheral joints.^{1–4} Recent reports show possible involvement of the kidneys.^{5–7}

We have observed a patient with symmetrical erosive polyarthritis of 14 years' duration mimicking classical rheumatoid arthritis, but with persistent absence of rheumatoid factor or other autoantibodies. The purpose of the following case report is to draw attention to the possibility of joint erosions in relapsing polychondritis.

CASE HISTORY

The patient, a woman born in 1918, had Bell's palsy of the left facial nerve in 1940 with a recurrence in 1956. In 1968 and 1972 she had facial 'erysipelas' with swelling of the nose, frontal region, and both cheeks. The symptoms abated slowly with parenteral penicillin treatment. Simultaneously with the second inflammatory episode the patient became hard of hearing. In the course of 1973 symmetrical synovitis

developed in both knees, ankles, radiocarpal joints, and a few proximal interphalangeal joints of both hands. The patient also complained of morning stiffness of longer than one hour in the afflicted joints. Rheumatoid factor negative rheumatoid arthritis was diagnosed, and treatment with chloroquine and systemic, low dose corticosteroids produced substantial improvement of all symptoms.

A few months later radiotherapy of the left shoulder and right knee was necessary owing to intractable pain. In 1978 pulmonary infarction and pneumonia with bilateral pleural effusions after deep venous thrombosis of the left calf occurred.

Until 1984 the polyarthritis showed an undulating course with asymptomatic periods of several months. On clinical examination the peripheral joints showed persistent bilateral, low grade synovitis of the metacarpophalangeal joints, hyperextension of the interphalangeal joint of the right thumb, swan neck deformity of various fingers (Figs 1a and b), reduced wrist motility, extension deficits of both elbows, and subluxation of all metatarsophalangeal joints (Fig. 2). An x ray examination of the hands (Figs 3a and b) and forefeet (Figs 4a and b) showed erosive destructions of the radiocarpal, intercarpal, metacarpophalangeal, isolated proximal interphalangeal, and metatarsophalangeal joints bilaterally. Rheumatoid factor and antinuclear autoantibodies remained persistently negative. In 1984 chloroquine was replaced by aurothiomalate, without substantial effect. In the same year there was spontaneous appearance of saddle nose deformity (Fig. 5). In

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Fig. 1a

Fig. 1b

Fig. 1 (a) Left and (b) right hand showing synovial swelling of radiocarpal, metacarpophalangeal, and isolated proximal interphalangeal joints and swan neck deformity of several fingers bilaterally.

Fig. 2 The right and left forefoot showing spreading, hallux valgus deformity, and dorsal subluxation of the second to fifth metatarsophalangeal joints bilaterally.

Fig. 3a

Fig. 3b

Fig. 3 x Rays of (a) the left and (b) the right hand showing juxta-articular osteopenia, erosive destruction of the radiocarpal, intercarpal, metacarpophalangeal, and isolated proximal interphalangeal joints bilaterally.

October 1985 the patient developed leucopenia ($3.0 \times 10^9/l$) and thrombocytopenia ($92 \times 10^9/l$), the cause remaining unclear. Both findings persisted after parenteral gold treatment was stopped. In 1986 painful swelling of the tragus of the left ear occurred, preventing further use of the hearing aid and necessitating excision of the tragus. Histology showed focal necrosis of the hyaline cartilage, chronic granulating perichondritis, and surrounding scar tissue (Fig. 6). A few months later the patient was readmitted to hospital because of upper gastrointestinal haemorrhage due to stomach ulcer caused by non-steroidal anti-inflammatory drugs.

FURTHER INVESTIGATIONS

In October 1986 further investigations showed an erythrocyte sedimentation rate of 90 mm/1st h;

haemoglobin 74 g/l; packed cell volume 0.24; normal erythrocyte indices. Leucocytes were $2.5 \times 10^9/l$ with normal differential; thrombocytes $45 \times 10^9/l$. Rheumatoid factor (RF) (IgM RF, IgA RF, IgE RF, and IgD RF) and antinuclear antibodies (including all subgroups) were negative. There were no circulating immune complexes. Normal values were obtained for electrolytes, blood urea nitrogen, creatinine, serum aspartate transaminase, serum alanine transaminase, alkaline phosphatase, glucose, and for serum IgG, IgM, IgA. There was hypoalbuminaemia. No light chains were found. Urinary sediment was normal. Bone marrow aspiration showed focal increase of polymorphic lymphoid elements, without selective monoclonal increase in any subpopulation, and without progression in the follow up examination 12 months later.

Fig. 4a

Fig. 4b

Fig. 4 x Rays of (a) the left and (b) the right forefoot showing diffuse osteopenia, erosive destruction, and subluxation of all metatarsophalangeal joints.

HLA typing showed positive antigens of the loci A2, A29, Bw44, Bw4, Cw5, DR4, and DR7. Antiphospholipid antibody was negative (March 1988).

Discussion

Relapsing polychondritis with erosive and deforming joint involvement has to our knowledge only once been reported.⁸ This is rather unusual for a systemic disorder characterised by episodic inflammation and destruction of cartilage and its surroundings. The present case report is intended to draw attention to the possibility that relapsing polychondritis may mimic rheumatoid arthritis by developing a symmetrical and erosive deforming polyarthritis of small and large joints. Our patient showed recurrent inflammation of ears and nose, leading to destruction of the left tragus and saddle nose deformity. She also developed an erosive deforming arthritis, mainly involving the radiocarpal, metacarpophalangeal, metatarsophalangeal, and isolated proximal interphalangeal joints bilaterally. The arthropathy misled the treating physicians for

many years because of its erosive nature and the late appearance of auricular and nasal destruction. Her serum was persistently negative for rheumatoid factor, she never developed subcutaneous nodules, and she showed no evidence of a metabolic disease despite extensive investigation. Several unusual systemic features (leucopenia, thrombocytopenia, and deep venous thrombosis) could imply coexistent systemic lupus erythematosus. The patient, however, does not fulfil the revised criteria for the classification of systemic lupus erythematosus proposed by Tan *et al.*⁹ Antinuclear autoantibodies (including all subgroups) were persistently undetectable. In addition, the antiphospholipid antibody titre was negative.

Until now the arthropathy of relapsing polychondritis has been considered as migratory, asymmetric, anodular, non-erosive, and rheumatoid factor negative, affecting large and small peripheral joints as well as parasternal and sacroiliac articulations.^{3 10 11} The feet have been reported to be spared.³ Our case, therefore, is the first with definite involvement of the metatarsophalangeal

Fig. 5 Saddle nose deformity appearing many years after development of symmetrical erosive polyarthritis.

Fig. 6 Histology of the tragus showing focal necrosis of the hyaline cartilage, chronic granulating perichondritis, and surrounding scar tissue.

joints. The arthritic attacks were episodic, not temporally related to the extra-articular inflammation of nose and ears, and were at times very painful.

Destructive and erosive joint lesions in patients with relapsing polychondritis are still a matter for debate. Braunstein *et al* and O'Hanlan *et al* suggest that erosive joint changes are not a feature of true relapsing polychondritis, but probably due to coexistent rheumatoid arthritis or another inflammatory arthropathy.^{10 11} Such coexistence (with rheumatoid arthritis or lupus erythematosus) has been frequently recorded.^{1 3 12-15} In contrast, Johnson *et al* showed erosive articular changes indistinguishable from those of classical rheumatoid arthritis in one case of relapsing polychondritis.⁸ To conclude, after exclusion of other conditions we believe that relapsing polychondritis is the cause of the symmetrical and erosive lesions in the radiocarpal, metacarpophalangeal, metatarsophalangeal, and isolated proximal interphalangeal joints of this patient. She

fulfills the diagnostic criteria of relapsing polychondritis, as proposed by McAdam *et al*.³

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