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Erosive arthritis in relapsing polychondritis

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Summary: We report the case history of a 57 year old man who has suffered from typical deforming, relapsing polychondritis for 13 years. He has also developed erosive destructive seronegative polyarthritis involving some of his distal interphalangeal, proximal interphalangeal, metacarpophalangeal, intercarpal, wrist, intertarsal and metatarsophalangeal joints. The distribution of joint involvement in the small joints of the hands and feet is asymmetrical. Both hips and knee joints have also been involved necessitating bilateral total hip and right total knee replacement. The articular associations with relapsing polychondritis are discussed.

Introduction

Relapsing polychondritis (RP) is a relatively rare disease of unknown aetiology characterized by recurrent inflammation and progressive destruction of the cartilages of the external ear, nose, laryngotracheobronchial tree, and the hyaline cartilage of large and small joints. It is a multisystem disorder with involvement of the uveal tract, internal ear, cardiovascular system, skin and occasionally the kidney.

RP has also been associated with other systemic diseases (often involving the joints) in 25–35% of patients. These include rheumatoid arthritis, systemic lupus erythematosus, Sjögren's syndrome, systemic sclerosis, thyroid disease, ulcerative colitis, malignancy, sinusitis, mastoiditis, diabetes mellitus and psoriatic arthritis.[1-3]

It has been suggested that the 5- and 10-year probabilities of survival after diagnosis are 74% and 55% respectively. The most frequent causes of death are infection, systemic vasculitis and malignancy.[4]

The joints are involved in at least 80% of cases of RP and the usual pattern of involvement is migratory, asymmetric, non-nodular, non-erosive, seronegative, and affecting large and small joints as well as parasternal articulations.[5]

We report the case history of a 57 year old man who has suffered from typical deforming RP for 13 years and has since developed distinctive, erosive, destructive, persistently seronegative polyarthritis.

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Case report

RG was first seen in November 1976, then aged 44 years, when he presented with a 6-month history of recurrent swelling of both ears, arthralgia of the large joints of the lower limbs, loose bowel motions and painful red eyes with blurring of vision. Investigations at the time revealed haemoglobin 10 g/dl, white cell count 9.4×10^9/l, erythrocyte sedimentation rate (ESR) 130 mm in the first hour, and rheumatoid factor was negative. A diagnosis of RP was made and the patient was commenced on 30 mg of prednisolone per day with a dramatic response. By December 1977, he was completely asymptomatic, his haemoglobin was 14.4 g/dl and his ESR was 20 mm in the first hour and consequently the prednisolone was stopped.

In May 1978, it was noted that he had persistent proteinuria of 1.0 to 2.0 g/24 hours and his ESR rose to 71 mm/hour but serum urea and creatinine and intravenous pyelography were normal. A renal biopsy showed mild, focal proliferative glomerulonephritis. His antinuclear and dsDNA binding antibodies were negative. During the period of investigations, the patient had an episode of vertigo which was thought to be part of the clinical picture of RP. Oral prednisolone 30 mg a day was recommenced with rapid improvement of the proteinuria and the patient had no further attacks of vertigo. By the end of 1980, the prednisolone was stopped and the proteinuria did not recur.

Eighteen months later, the chondritis of both ears and uveitis recurred and he also developed arthritis affecting both knee and hip joints. The distal (DIP), proximal interphalangeal (PIP), metacarpophalangeal (MCP), wrist and metatarsophalangeal (MTP) joints were swollen and tender. He also had painful costochondritis and nasochond-

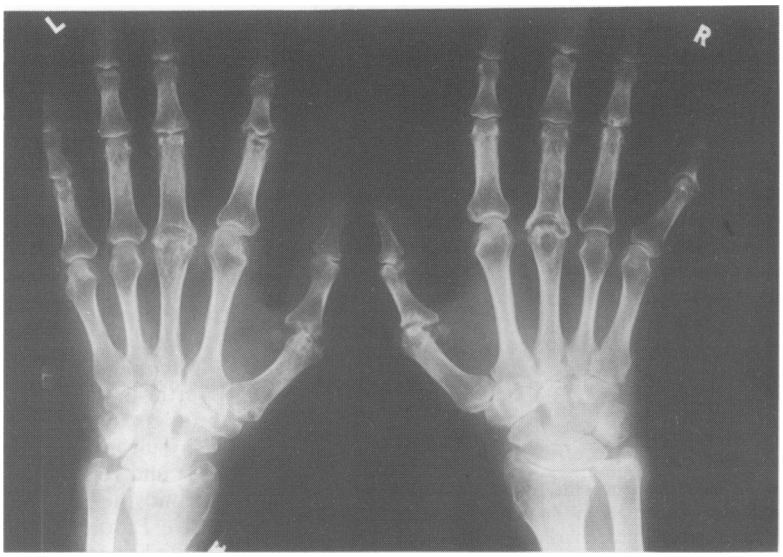

Figure 1 Advanced erosive changes are seen in several DIP, PIP, MCP, intercarpal and wrist joints. Resorptive changes are also clearly shown in the PIP of the right little finger, the MCP of the middle finger on both sides.

ritis. The latter healed with some collapse of the bridge of the nose. Radiological examination of both hands and feet showed several erosive changes with narrowing of the joint space affecting several DIP, PIP, MCP, intercarpal, MTP and intertarsal joints. The rheumatoid factor remained negative.

The arthritis in hands, feet, hips and knees continued to progress and radiological examination performed in August 1986 showed advanced erosive changes in several DIP, PIP, MCP intercarpal and wrist joints (Figure 1). There were also advanced arthritic changes with loss of articular cartilage affecting both knee joints but more marked on the right side (Figure 2), and similar changes were seen in both hips and, again, the appearances were more marked on the right side (Figure 3). In April 1988, he had a right total knee replacement. Total hip replacements were performed on the right in March 1989 and on the left in August 1989.

Since March 1989, he has been on azathioprine 125 mg and prednisolone 7.5 mg daily and remains clinically well.

Discussion

The arthropathy of RP is a major clinical feature of the disease and usually shows two distinct patterns. In patients with 'lone' RP, the arthritis is described as migratory, asymmetric, non-nodular, seronegative and affecting large and small joints, and most probably caused by an underlying inflammatory process affecting the articular cartilage. On the other hand, a minority of patients develop the cardinal features of RP against a background of a well-established chronic arthritic condition, such as rheumatoid arthritis[6,7] and psoriatic arthritis.[3]

Our patient has suffered typical deforming RP associated with a seronegative, erosive, distinctive polyarthritis involving some of his small joints in the hands (incuding the DIP joints) and feet as well as hip and knee joints which necessitated several major joint replacements. His arthritis is quite

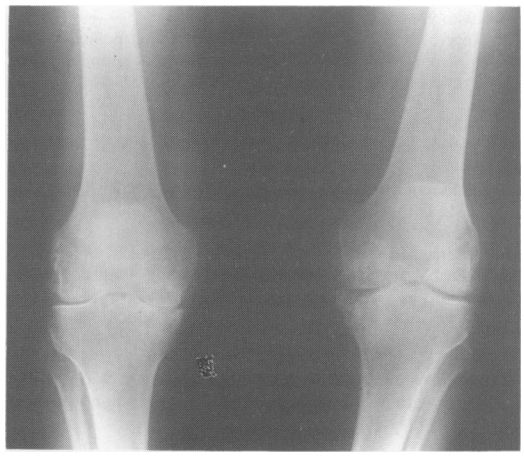

Figure 2 Advanced arthritic changes affecting the right knee, especially the medial compartment are seen. Similar but less severe changes are seen on the left side with some destructive changes affecting the medial tibial plateau.

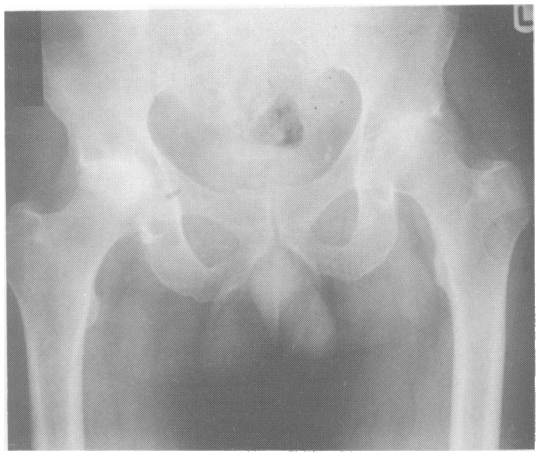

Figure 3 Severe arthritic changes with loss of articular cartilage and some erosive changes in the femoral heads are seen. The appearances are more marked on the right.

different from the arthritis reported in RP. O'Hanlan *et al.*[5] described 16 patients with 'pure' RP polyarthritis and none had developed progressive or deforming arthritis but the average duration of follow-up was only 4.3 years. More recently, Booth *et al.*[8] described the arthritis in their 3 patients with RP as non-erosive and commented that the arthritis was characterized by pure joint space narrowing without erosion but, again the mean period of follow-up was 4.4 years. We have followed-up our patient for 13 years, far longer than the patients of O'Hanlan *et al.* and Booth *et al.*, and it took nearly 5 years for the arthritis to become erosive and rapidly destructive. It may well be that the erosiveness of the arthritis is related to the longer duration of the disease. McAdam[1] suggested that the forefoot is not affected, but this is not our experience, nor the experience of Booth.[6] Previously reported spinal involvement[5] and sacroiliac changes[9] were not seen in our patient.

Our patient's arthritis is also not typical of rheumatoid arthritis because of the asymmetrical distribution of the joint involvement in the small joints of the hands and feet, the prominent DIP joint involvement and the fact that he was persistently seronegative. Psoriatic arthropathy is also highly unlikely in that he has never had psoriasis in his skin or nails and he has been followed-up for a considerable length of time.

It is interesting to note that our patient has had an episode of mild, focal, proliferative glomerulonephritis which has responded promptly to a course of corticosteroids and has not since recurred. Glomerulonephritis and renal vasculitis have been rarely but well-documented in RP.[10-14]

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