Destructive arthropathy in chondrocalcinosis articularis

A. J. RICHARDS AND E. B. D. HAMILTON

King's College Hospital, London

It is now well recognized that idiopathic chondrocalcinosis articularis, as well as being associated with acute episodes of pyrophosphate arthropathy, may lead to premature degenerative changes in the joints involved. Some workers have also made occasional reference to the presence of juxta-articular osteolytic lesions (McCarty, Kohn, and Faires, 1962; Currey, 1970.) More recently there have been reports of destructive joint changes which can be the cause of considerable disability (Robinson, 1971; Menkes, Simon, Chouraki, Ecoffet, Amor, and Delbarre, 1973).

Five patients (4 women and 1 man), from a series of eighty patients with idiopathic chondrocalcinosis seen at King's College Hospital, have developed a severe destructive arthropathy. In none of them was there evidence of haemochromatosis or hyperparathyroidism and serological tests for syphilis and for rheumatoid factor were negative. One of the five patients has recently developed diabetes mellitus and another has Paget's disease, although neither of these conditions is causing symptoms.

Case reports

Patient 1, a 75-year-old man, was first seen in 1969 complaining of pain and swelling of both hands and pain in the right knee and neck. Investigations showed a normal erythrocyte sedimentation rate (ESR), a negative Waaler-Rose test, and widespread chondrocalcinosis in radiographs of his hips, knees, and lumbar spine. The serum calcium was in the normal range but the alkaline phosphatase was raised at 99 i.u./litre (normal range 30–85), and Paget's disease of the pelvis was seen radiologically. Apart from the chondrocalcinosis there were degenerative changes in the hips, knees, and spine. The radiographs of the hands are interesting in that they show unusual destructive changes, especially in the right hand (Fig. 1), and faint calcification of hyaline cartilage is present in the metacarpophalangeal joint of the middle finger. The destructive change at this joint is obvious with cyst formation and crumbling of the joint surface, but marginal erosions as seen in rheumatoid arthritis are not present.

Patient 2, a 69-year-old woman, developed pain and swelling of the left shoulder. An effusion of the left shoulder joint was aspirated and calcium pyrophosphate crystals

FIG. 1 *Case 1. Male, aged 75 yrs. Chondrocalcinosis and destructive change in head of third right metacarpophalangeal joint*

FIG. 2 Case 2. Female, aged 69 yrs. Irregularity and destruction of the joint surface of the left shoulder with flattening of the head of the humerus

were seen under the polarizing microscope. Radiologically chondrocalcinosis was seen in the knees, wrists, and symphysis pubis, together with some osteoarthritic change in these joints. The left shoulder shows early destructive change (Fig. 2). There is flattening of the humeral head, irregularity of the articular surface, loss of the bounding line, and the juxta-articular bone is increased in density.

Patient 3, a 75-year-old woman, developed pain and stiffness in many joints in 1948. Over the years she has been developing a widespread destructive arthropathy affecting shoulders, hips, and knees, and chondrocalcinosis was seen in the knees and wrists. The right shoulder showed early destructive change in 1962 (Fig. 3a) with sclerosis of subchondral bone and irregularity of the joint surface. In 1964 the destructive change was more marked (Fig. 3b) and in 1968 gross destruction had occurred (Fig. 3c). There was a very large effusion of the right shoulder and this was found to contain crystals of calcium pyrophosphate on examination. Similar destructive changes are present in the left shoulder, though not so marked (Fig. 4). The severe destructive change was also seen in the right hip joint in 1968 (Fig. 5), in the left knee joint in 1967 (Fig. 6a) with progressing destruction during the following year (Fig. 6b).

In this patient the deep pain sensation was normal and there were no abnormal neurological signs. Her serum calcium, phosphate, and alkaline phosphatase were within the normal range, and the 24-hr urinary calciums were 140 and 156 mg. The serological tests for syphilis were negative and no increase in rheumatoid factor was found. Joint cultures were sterile. Osteomalacia was considered but a bone and jejunal biopsy were normal. Only recently have

raised blood sugars and glycosuria been noticed, but she has no symptoms of diabetes and we do not feel that this has played any part in the causation of her destructive arthropathy. She is able to get about with some difficulty with the aid of full-length calipers.

Patient 4, a 78-year-old woman, presented with pain and swelling of the left knee. Radiographs showed osteoarthritic changes and chondrocalcinosis in the knees, wrists, and hands. The left knee shows destructive change affecting the lateral femoral condyle and tibial plateau (Fig. 7). The lateral radiograph of the right knee shows destruction of the articular surface of the patella which is wrapped round the femur, and there is also calcification in the suprapatellar pouch (Fig. 8). Estimations of serum calcium, phosphate, and alkaline phosphatase have been repeatedly in the normal range. The radiograph of the left hand (Fig. 9) shows erosive change at the radio-ulnar joint, and sclerosis, erosions, and loss of joint space at the radiocarpal joint. Cystic changes are present in the metacarpal heads and calcification of hyaline cartilage of the second and third metacarpal heads can be faintly seen. The radiograph of the right wrist shows a curious erosive lesion affecting the inferior radio-ulnar joint (Fig. 10).

Patient 5, a 74-year-old woman, when first seen in 1968, was complaining of acute pain and swelling of the left knee. Fluid taken from the knee joint showed crystals of calcium pyrophosphate under the polarizing microscope. A radiograph shows destructive change in the lateral compartment of the left knee with loss of joint space (Fig. 11). Chondrocalcinosis is visible in the medial meniscus. The destruction of the left knee joint has pro-

FIG. 3a, b, c Case 3. Female, aged 75 yrs. Progressive changes in the right shoulder leading to complete loss of the humeral head

FIG. 4 Case 3. Female, aged 75 yrs. Severe destruction of the head of the left humerus

gressed and in September, 1972, a compression arthrodesis was performed. Histological examination of excised tissue confirmed calcification of fibrocartilage and hyaline cartilage. Calcification of part of the synovium was also found, and examination under the polarising microscope confirmed the presence of calcium pyrophosphate crystals.

Discussion

Menkes and others (1973) give details of 23 patients with destructive joint changes, chiefly affecting the knees, shoulders, hips, and wrists. Some of the radiographs of our five patients are identical to theirs. They note that involvement of the hip joint may take

the form of a rapidly destructive osteoarthritis and, like the third patient of our series, this severe destructive change may simulate a neurogenic arthropathy.

Destructive changes, especially of the metacarpophalangeal joints, may be seen in haemochromatosis, and there is a known association of chondrocalcinosis with that condition. There are no clinical or biochemical signs of haemochromatosis in our five elderly patients (average age 74 yrs), and it is therefore interesting to find that the radiographs of the hands of the first patient of this series (Fig. 1) show destructive changes of the metacarpophalangeal joints identical to those that may be seen in haemochromatosis

FIG. 5 *Case 3. Female, aged 75 yrs. Dissolution of the right femoral head*

FIG. 6a, b *Case 3. Female, aged 75 yrs. Progressive destruction of the left knee joint*

FIG. 7 Case 4. Female, aged 78 yrs. Destruction of the surface of the femoral condyle and tibial plateau of the outer side of the left knee joint

FIG. 9 Case 4. Female, aged 78 yrs. Erosive change at left inferior radio-ulnar joint. Cystic changes and calcification of hyaline cartilage in second and third metacarpophalangeal joints

(Hamilton, Williams, Barlow, and Smith, 1968; Dymock, Hamilton, Laws, and Williams, 1970). Shoulder radiographs similar to that seen in the second patient (Fig. 2) have also been seen by us in haemochromatosis.

Another interesting feature is the lateral radiograph of the right knee of our fourth patient (Fig. 8). This shows the patella wrapped round the femur, a feature which has been described in hyperparathyroidism (Bywaters, Dixon, and Scott, 1963). Calcification in the suprapatellar pouch is also visible, and there are erosive changes in the inferior radio-ulnar joints. We thought that in view of these radiographic findings the patient might have hyperparathyroidism, and there is a well recognized association of chondrocalcinosis with hyperparathyroidism (Bywaters, 1959; Hosking and Clennar, 1960; Zvaifler, Reefe, and Black, 1962). However, several estimations of plasma calcium, phosphate, and alkaline phosphatase have always been in the normal range, and for this reason

FIG. 8 Case 4. Female, aged 78 yrs. The patella is wrapped round the lower end of the femur and calcification in the suprapatellar pouch

FIG. 10 Case 4. Female, aged 78 yrs. Erosive lesion of right inferior radio-ulnar joint

we do not feel that hyperparathyroidism is the cause of these radiographic appearances.

At the present time we do not know why these destructive changes occur in some patients with chondrocalcinosis articularis and not in others. The severe destructive arthropathy of our third patient is most striking, and some of her radiographs have been shown in an earlier paper on chondrocalcinosis by Atkins, McIvor, Smith, Hamilton, and Williams (1970). Jacobelli, McCarty, Silcox, and Mall (1973)

have recently described the deposition of calcium pyrophosphate dihydrate in neuropathic joints in four cases with polyarticular involvement, all of whom had positive serological tests for syphilis, and have suggested that the two conditions may act synergistically to produce the neuropathic picture. In Case 3 such a mechanism was suspected by us, but serological tests for syphilis were negative.

At the molecular level *in vitro*, Schumacher and Phelps (1972) showed that calcium pyrophosphate

FIG. 11 Case 5. Female, aged 74 yrs. Destructive change in the lateral compartment of the left knee joint

crystals do not produce lysis of the membranes of phagocytic vacuoles in polymorphonuclear leucocytes, whereas sodium urate crystals do. These workers also found that the dramatic sequence of increasing cell necrosis that occurs with urates was not seen with calcium pyrophosphate. There appears to be no correlation with these sequential electron microscopic observations and the severe destructive change *in vivo* in some patients with calcium pyrophosphate arthropathy. A possible explanation for the bone destruction is that this is the result of ischaemic necrosis. None of the five patients has been on corticosteroid therapy, which is sometimes associated with ischaemic necrosis of bone. However, avascularity could possibly occur as a result of the inflammatory changes caused by calcium pyrophosphate crystals in the joint fluid and tissue. The changes we have described do not look like advanced osteoarthritis, except possibly in the knees, although even here it must be stressed that both femoral and

tibial components of the knee joints were affected, and that it was not simply a collapse of the tibial plateau.

Treatment of patients with destructive arthropathy is initially on a symptomatic basis using drugs such as phenylbutazone or indomethacin to control the pain. Later surgical procedure such as arthrodesis (Patient 5) or total excision arthroplasty may be required.

Summary

A well-marked destructive arthropathy has been seen in five patients with chondrocalcinosis articularis. The joints involved included the metacarpophalangeal, wrists, shoulders, hips, and knees. There was no evidence of rheumatoid arthritis, hyperparathyroidism, haemochromatosis, or neurological abnormality in any of the patients.

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