Case report

Unusual radiographic features in a female patient with ankylosing spondylitis

TIMOTHY J CUNNINGHAM, PETER M CUMBER,
GORDON EVISON, AND PETER J MADDISON

From the Royal National Hospital for Rheumatic Diseases, Upper Borough Walls, Bath

SUMMARY A 71 year old woman suffering from ankylosing spondylitis with aggressive peripheral joint disease for 46 years developed unusual radiographic features in the hips and knees. The pathogenic processes responsible are uncertain, and no histology is yet available. Although she had been treated with radiotherapy to the knees, there was no similar history to explain the hitherto unreported appearance of the hips.

Key words: hip disease, radiology.

Peripheral joint involvement in ankylosing spondylitis (AS) is more common in women than men,¹ the most frequently affected site being the hip.² Radiological knee involvement occurs in approximately 30% of patients with disease of long duration.³ Involvement of the peripheral joints has been reported as an indicator of poorer prognosis.⁴ The case reported here illustrates a protracted history of destructive peripheral joint involvement in a woman with AS with atypical radiographic features.

Case report

A 71 year old woman had had ankylosing spondylitis for 46 years. One year after the onset of pain and stiffness in the lumbar region ankylosing spondylitis was diagnosed and she was treated with radiotherapy to the lumbar spine only. Four years later she developed involvement of several peripheral joints, and gold therapy was given for six months. Further courses of radiotherapy were given 12 to 14 years after the onset of the disease to the cervical and dorsal spine, the right elbow, and left knee and after

a further 20 years to both knees, left elbow, and right wrist, with limited benefit. Peripheral joint disease remained a major problem, with numerous intra-articular corticosteroid injections being given to both knees. At the most recent examination there was persistent synovitis of the right shoulder, left wrist, and both knees. The patient had never had psoriasis or other cutaneous disease, symptoms of genitourinary or gastrointestinal disease, or symptoms suggestive of iritis. The patient had suffered from insulin dependent diabetes since the age of 39, complicated by diabetic retinopathy, and mild hypertension, though renal function was normal. There was no family history of ankylosing spondylitis, psoriasis, or rheumatic disease apart from her father having had mild osteoarthritis of his hands.

Clinical examination showed a well looking woman, with bilateral cataracts, optic atrophy in the left eye, and bilateral diabetic retinopathy with microaneurysms. Cardiovascular examination was normal apart from a non-radiating ejection systolic murmur in the aortic area. The cervical spine had only a few degrees of lateral rotation, and thoracic and lumbar spines were fused. Chest expansion was less than a centimetre. Peripherally there was synovitis of the right shoulder, left wrist, and both knees, with associated effusions and lateral instability of the knees.

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Correspondence to Dr Timothy J Cunningham, Royal National Hospital for Rheumatic Diseases, Upper Borough Wall, Bath BA1 1RL.

Investigations confirmed that the patient was HLA-B27 positive and in addition was DR4 negative. In 1983 the rheumatoid factor first became positive by nephelometry at 101 IU/l (less than 40 units considered negative).

Radiological examination showed fusion of both sacroiliac joints, the pubic symphysis, and the cervical, thoracic, and lumbar spine. The appear-

ance of the hips was unusual in showing marked 'waisting' of both femoral necks, which contained large, well demarcated cystic lesions. Fig. 1 shows the progression of these changes since 1967. Throughout there was relative preservation of the joint space, though erosive changes developed in both greater and lesser trochanters. Progression of the changes in the knee is shown in Fig. 2. The

Fig. 1a

Fig. 1b

Fig. 1 Progression of radiological changes in the pelvis from 1967 (a) to 1985 (b), showing fusion of sacroiliac joints, pubic symphysis, and atypical hip destruction.

Fig. 2a

Fig. 2b

Fig. 2 *Progression of changes in the knees from 1967 (a) to 1985 (b), showing destruction of lateral femoral condyle and lateral tibial plateau with loss of joint space of the right knee.*

right knee developed gross destructive changes, particularly in the lateral femoral condyle and lateral tibial plateau, with loss of articular cartilage. Loss of joint space was less marked on the left but there was development of marginal erosions on the tibia. x Rays of the hands (Fig. 3) showed marked carpal destruction with lysis and tapering of the distal ulnar shafts. Small erosions were present at

the right fifth metacarpophalangeal joint and third proximal interphalangeal joint.

Discussion

There is no doubt concerning the diagnosis of ankylosing spondylitis in this patient, who presents virtually complete axial fusion and severe continuing

Fig. 3 Hand x rays (1985) showing marked carpal destruction with lysis and tapering of the distal ulnar shafts.

peripheral joint involvement. She has never developed psoriasis, and the rheumatoid factor only became positive 40 years after the onset of progressive destructive peripheral joint disease. Furthermore, there have never been other features to suggest coexistent rheumatoid disease.

Characteristic radiological appearances of hip involvement in ankylosing spondylitis include concentric joint space narrowing, which may produce axial migration of the femoral head, and osteophytosis, typically appearing early on the lateral aspect of the femoral head.⁵ Bony ankylosis of the hip is reported to be more a feature of AS in women.⁶ Two radiographic patterns of spondylitic hip disease were reported by Forestier *et al*⁷; firstly, a non-destructive ankylosing form in young patients and secondly, a slower unilaterally destructive process in older individuals. The radiological appearances of the hips in this case are quite atypical for AS and to our knowledge not yet described in the literature. In a review of the hip radiographs of 150 patients with AS seen at the Royal National Hospital for Rheumatic Diseases (40 of whom were female and 24 of whom had undergone hip replacement surgery) similar cystic change and 'waisting' of the femoral necks was not observed.

Radiation induced tissue destruction or the effects of previous long acting corticosteroid injections may have played a part in the severe damage seen in the knees of the patient but cannot explain the changes in the hips. Several mechanisms are

probably operating to produce this unusual radiological appearance. The cystic changes in the femoral necks for example, are similar to those shown in pigmented villonodular synovitis due to proliferation of the synovial membrane, and marked synovial proliferation in a tightly encapsulated joint may be responsible for the cysts developing in this case. 'Waisting' of the femoral neck may be analogous to the spindling frequently seen in the metacarpal and metatarsal knees of patients with seronegative spondylarthritides and seen in the distal ulnae of this patient.

The radiological picture described here is only rarely seen, but we describe this case to add to the spectrum of peripheral joint disease seen in AS.

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