

## CASE STUDIES IN DIAGNOSTIC IMAGING

## Bloody arthritis

D J Annesley-Williams, A Mark Davies, N Evans

**Clinical history**

A 25 year old carpenter presented with a two day history of intense pain in the left ankle. The pain started suddenly when he injured the ankle while working in the joinery, and had been getting steadily worse ever since. The patient was known to suffer from a chronic haematological disorder, and had previously experienced multiple similar episodes affecting in particular the left ankle and right knee joints, often following minor trauma.

On examination he held the ankle dorsiflexed and was unable to weight bear. Movement was limited in all directions. The ankle appeared swollen and was exquisitely tender to palpation. Examination was otherwise normal. The patient was afebrile. Serological tests (rheumatoid factor, erythrocyte sedimentation rate, C reactive protein) were negative. Plain radiographs and magnetic resonance images of the knee and ankle joints were obtained.

**Radiological findings**

## PLAIN RADIOGRAPHIC FINDINGS

Radiographic examination of the left ankle (fig 1) shows a joint effusion with prominence of the soft tissue both anterior and posterior to the joint. The anterior joint space is narrowed with erosions and a large subarticular cyst in the distal tibia. The bones are osteopenic.

The right knee joint (fig 2) shows signs of chronic hyperaemia; loss of secondary trabeculae in the period of osteopenia has led to coarsening of the epiphyseal trabecular pattern. The intercondylar notch is widened and there is joint space narrowing affecting both compartments of the knee joint.

## MAGNETIC RESONANCE IMAGING

Magnetic resonance (MR) images of the left ankle joint were obtained using a one Tesla superconducting magnet (Siemens Impact) (figs 3 and 4). The joint space is narrowed with destruction of articular cartilage, subchondral bone, and subarticular cyst formation on both sides of the joint. Synovial proliferation (dark on all sequences) is seen in both the anterior and posterior recesses of the joint. Marrow oedema, best visualised on the STIR sequence, is also present (fig 4).

MR imaging of the right knee was also performed, which shows similar synovial proliferation and osteoarticular changes (fig 5).



Figure 1 Lateral radiograph of the ankle joint showing osteopenia, joint space narrowing, and a subarticular cyst in the distal tibia.

**Differential diagnosis**

The plain radiographic appearances are those of a degenerative arthropathy—an unusual occurrence at this site, particularly in a young man. The serology does not indicate a generalised inflammatory aetiology. The MR appearances of the synovial hypertrophy (decreased signal intensity on all sequences) imply that there are few mobile protons present to contribute to the image. The most common causes of such appearances are air, foreign body, haemosiderin deposition due to recurrent intra-articular haemorrhage, or fibrous tissue. Based on the MR images the differential diagnosis will include haemophilic arthropathy, pigmented villonodular synovitis, or amyloid arthropathy. Taking the clinical history into account haemophilic arthropathy is the final diagnosis.

**Discussion**

Haemophilia A is the commonest of the hereditary bleeding disorders, with an incidence of between 30 and 120 per million of the population<sup>1</sup> and is caused by deficiency of factor VIII. This gene is located on chromosome Xq 2.8 and is therefore transmitted as a

Department of  
Radiology, North  
Staffordshire Hospital  
Trust, Stoke-on-Trent,  
United Kingdom  
D J Annesley-Williams

The MRI Centre, The  
Royal Orthopaedic  
NHS Trust,  
The Woodlands,  
Birmingham, United  
Kingdom  
A M Davies  
N Evans

Correspondence to:  
Dr D J Annesley-Williams,  
43 Ormes Lane, Tettenhall,  
Wolverhampton WV6 8LL.

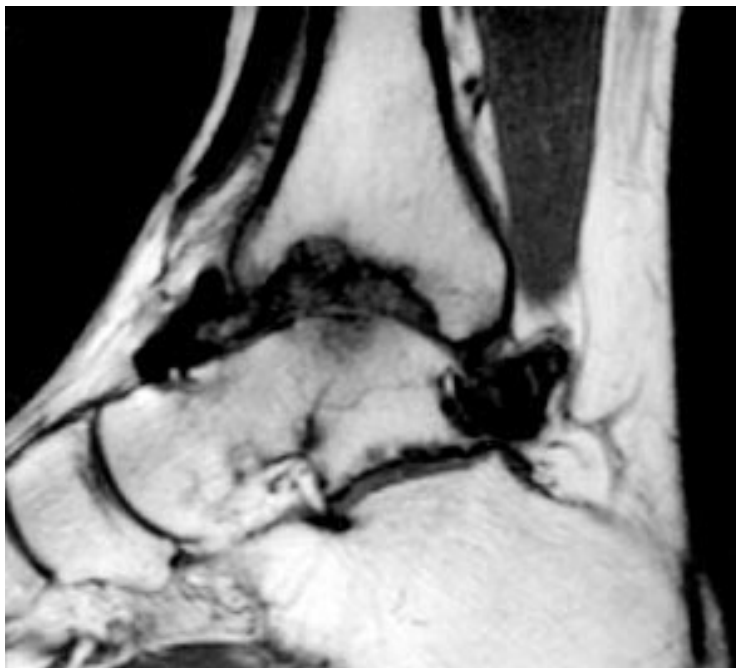
Accepted for publication  
4 October 1996



*Figure 2 Anteroposterior radiograph of the knee joint showing coarsening of the trabecular pattern of the epiphyses and widening of the femoral intercondylar notch. There is loss of joint space involving both medial and lateral compartments.*

sex linked recessive defect. No family history can be established in about one third of cases.

Factor VIII deficiency leads to slowing of thrombin generation through the intrinsic



*Figure 3 Sagittal T1 weighted magnetic resonance image of the ankle joint showing joint space narrowing, subarticular cyst formation in the distal tibia, and low signal synovial proliferation anterior and posterior to the joint.*

pathway. Following trauma or surgery, clot formation is delayed and the clots, once formed, are liable to fragment easily, leading to spontaneous bleeding into major joints. Seventy per cent of haemophilia sufferers have experienced bleeding into a joint by the age of 2 years.<sup>2</sup> In the acute situation distension of the joint capsule causes exquisite pain.

In the young, local hyperaemia leads to enlargement of the epiphysis with altered modelling of the femoral intercondylar notch (fig 2) and premature closure of the growth plate. Periarticular osteoporosis occurs as a result of recurrent haemarthrosis. The appearances on plain film radiography can be indistinguishable from tuberculosis or juvenile chronic arthritis.

The hallmark of haemophilic arthropathy is accelerated destruction of cartilage. Radiologically, this resembles severe osteoarthritis (fig 1), with marked bony deformity and ultimate ankylosis. A florid proliferative synovial overgrowth forms a pannus, similar to that found in rheumatoid arthritis. This causes marginal erosions of the joint surfaces with occasional larger defects, and enhances destruction of articular cartilage (figs 3 and 4). Changes are most commonly found in the knees, ankles, and elbows but also occur, less frequently, in wrists, hips, and shoulders. Long term complications include joint deformity, avascular necrosis, chondrocalcinosis, haemophilic pseudotumours, and premature osteoarthritis.

In some countries positive HIV status is prevalent in haemophilic patients transfused with HIV positive blood products and predisposes to the appearance of septic arthritis in haemophiliacs.<sup>3</sup>

In the past, imaging assessment relied solely on plain film radiography. Currently, ultrasound, computed tomography (CT), and MR imaging are being increasingly used in an attempt to detect haemophilic arthropathy at an early stage. The results of ultrasound examination of the knees in 50 patients with haemophilic arthropathy suggest that sonography allows differentiation between suprapatellar effusion and synovial thickening in the knee in the early stages of the disease. It also reveals early cartilaginous changes and is valuable in following the progression and regression of extra-articular haematomas.<sup>4</sup>

Pseudotumour is a rare complication of haemophilia, occurring in 1-2% of sufferers. It develops most commonly in the large muscles of the pelvis and lower extremity where there is a rich blood supply. If bleeding into a muscle fails to resolve, the haematoma becomes encapsulated and forms a pseudotumour. Intra-articular pseudotumour can also occur following intraosseous bleeding. CT has an important role in detecting pseudotumours and in assessing both the extent of the soft tissue mass and the involvement of bone.<sup>5</sup>

MR imaging can be used to detect early synovial hypertrophy and to examine the status of articular cartilage.<sup>5</sup> Synovial hypertrophy shows decreased to intermediate signal on both T1 and T2 weighted images, with foci of increased signal intensity on T2 weighted



Figure 4 Sagittal STIR (short TAU inversion recovery sequence) magnetic resonance image of the ankle joint showing arthropathy and subarticular cyst formation on both sides of the joint. Marrow oedema, typified by increased signal intensity in the medulla of the distal tibia and talus.



Figure 5 Sagittal T1 weighted magnetic resonance image of the knee joint showing low signal synovial proliferation in the suprapatellar pouch and posterior recess of the knee joint. Degenerative osteoarticular changes are also seen anteriorly.

images due to areas of fluid or inflammation.<sup>6</sup> Following intravenous enhancement with

paramagnetic contrast medium (gadolinium - DTPA), synovial proliferations show an increase in signal intensity allowing improved delineation and quantification of the inflammatory process.<sup>7</sup> Recurrent intra-articular bleeding leads to haemosiderin deposition which yields low signal intensity on both T1 and T2 weighted images. Magnetic resonance imaging may differentiate between acute and chronic bleeding in soft tissue.

The differential diagnosis in this case includes pigmented villonodular synovitis, which is a synovial disease of young adults typically affecting either the joints or the tendon sheaths.<sup>8</sup> It is usually monoarticular, involving the knee in 80% of cases. Effusion and synovial proliferation lead to joint swelling. Plain radiographs show a non-calcified synovial soft tissue mass with marginal erosions on one or both sides of the joint. Synovial proliferation eventually leads to large cystic defects with sclerotic margins. Bone density and joint space are preserved and there is no evidence of hypertrophic new bone formation. On magnetic resonance imaging, pigmented villonodular synovitis also yields low signal intensity on both T1 and T2 weighted images due to haemosiderin deposition within the synovium.<sup>9</sup>

Appearances can be identical in haemophilic arthropathy and thus it may be impossible to distinguish these two conditions on MR imaging appearances alone. The clinical history is therefore invaluable in arriving at the final diagnosis.

Rare causes of low signal intensity within a joint include amyloid arthropathy. This intra-articular deposition of amyloid is well known to be derived from circulating B2 microglobulin in patients on long term haemodialysis.<sup>10</sup>

- 1 Weatherall DJ, Ledingham JGG, Warrell DA. Oxford textbook of medicine, 2nd ed. Oxford: Oxford University Press, 1987:19.213.
- 2 Grainger RG, Allison DJ. Diagnostic radiology: an Anglo-American textbook of imaging, 2nd ed. Edinburgh: Churchill Livingstone, 1992:1574-6.
- 3 Merchan EC, Magallon M, Manso F, Martin-Villar J. Septic arthritis in HIV positive haemophiliacs. Int Orthop 1992;16:302-6.
- 4 Merchan EC, De Orbe A, Gago I. Ultrasound in the diagnosis of the early stages of haemophilic arthropathy of the knee. Acta Orthop Belg 1992;58:122-5.
- 5 Hermann G, Gilbert MS, Abdelwahab IF. Haemophilia: evaluation of musculoskeletal involvement with CT sonography and MR imaging. Am J Radiol 1992;158:119-23.
- 6 Yulish BS, Lieberman JM, Stransjord SE, Bryan PJ, Mulopulos GP, Modic MT. Haemophilic arthropathy: assessment with MR imaging. Radiology 1987;164:759-62.
- 7 Naegele M, Bruening R, Kunze V, Eickhoff H, Koch W, Reiser M. Haemophilic arthropathy of the knee joint: static and dynamic gadolinium-DTPA-enhanced MRI. Eur Radiol 1995;5:547-52.
- 8 Dorwart RM, Genant HK, Johnston WH, Morris JM. Pigmented villonodular synovitis of synovial joints—clinical, pathological and rheumatological features. Am J Radiol 1984;143:877-85.
- 9 Jelinek JS, Kransdorf MJ, Utz JA. Imaging of pigmented villonodular synovitis with emphasis on MR imaging. Am J Radiol 1989;152:337-42.
- 10 Cobby MJ, Adler RS, Swartz R, Martel W. Dialysis related amyloid arthropathy: MR findings in four patients. Am J Radiol 1991;157:1023-7.