Imaging the swollen joint in the young patient

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PATIENT 1
Clinical history
A four year old boy presented to an orthopaedic surgeon with a painful swollen left ankle, having been treated for tonsillitis one week previously. Blood count, plasma viscosity and C reactive protein were normal.

Radiological findings
A lateral radiograph (fig 1) showed distension of the anterior recess of the tibiotalar joint capsule, indicating joint disease. The joint was not aspirated. Intravenous gadolinium-diethylene triaminopentaacetic acid (IV Gd-DTPA) enhanced magnetic resonance imaging (MRI) showed that the distension of the anterior recess consisted predominantly of synovial thickening around a minor effusion (fig 2). Effusions were present in all the tendon sheaths posteriorly and in tibialis anterior (fig 3).

Diagnosis
The initial provisional diagnosis was septic arthritis, but the presence of synovial thickening in a joint and in numerous adjacent tendon sheaths is not typical of septic arthritis and indicated that this provisional diagnosis of infection was wrong. A provisional diagnosis of pauciarticular juvenile chronic arthritis (JCA) was made. Rheumatoid factor was negative, antinuclear antibody (ANA) was positive, and initial slit lamp examination showed no iridocyclitis. The ankle swelling did not resolve on treatment with naproxen, sulphasalazine and ibuprofen, and the ankle began to develop hind-foot valgus deformity. Two months later, the right knee developed a synovitis, whereupon injection of both ankle and knee with triamcinalone under general anaesthetic resulted in complete clinical resolution of synovitis.

PATIENT 2
Clinical history
A 14 year old girl presented to the orthopaedic outpatient clinic with four months of painless swelling of the left knee. Examination revealed knee effusion with a full range of movement. Full blood count and blood biochemistry were normal.

Radiological findings
A radiograph showed a thickened suprapatellar bursa with no bone changes (fig 4). Ultrasound of the knee (fig 5) showed thickening of the synovium, and effusion. Compression of the fluid gave a total synovial thickness of 12 mm (normal range in adults, 1-9-2-9 mm) (fig 6). MRI confirmed distension of the suprapatellar bursa. On T2 weighting, the synovium had low signal intensity and appeared irregular, particularly in the posterior recess of the joint (fig 7). However, there was strong enhancement after IV Gd-DTPA, whereupon the synovium was shown to be smooth and had no fronds typical of pigmented villonodular synovitis (fig 8). Rheumatoid factor was negative and ANA strongly positive (titre = 1 in 800).

Diagnosis
Although it is a rare disease, a diagnosis of pigmented villonodular synovitis (PVNS) was initially considered to be a potential cause of the large effusion in a single joint, but this was excluded by the failure to find typical synovial fronds. Uveitis was excluded by ophthalmological examination.
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Figure 2  Sagittal T1 weighted spin-echo images of left ankle (patient 1). Left: Intermediate-signal intensity synovial hypertrophy of the anterior recess before intravenous gadolinium (arrow). Right: Afterwards, strongly enhancing synovium (large arrow) around a small effusion (small arrow). Enhancing synovium around the flexor hallucis longus tendon is also visible (arrowheads).

The right knee then developed an effusion. A probable diagnosis of pauciarticular JCA was made (a definite diagnosis requires three months of disease with absence of another cause) and the left knee was aspirated and injected with corticosteroids, with good results.

PATIENTS 1 AND 2
Discussion
Two cases of probable pauciarticular juvenile chronic arthritis are described. The first was initially misdiagnosed as septic arthritis and the second as PVNS.

Figure 3  Axial T2 weighted spin-echo image of left ankle (patient 1) showing high signal (white) within the flexor tendon sheaths around the low signal (black) tendons (arrowheads).

Figure 4  Lateral radiograph of the left knee (patient 2) showing a thickened suprapatellar bursa which produces a stripe of soft tissue density (arrowheads) between the quadriceps tendon and the prefemoral fat.

Juvenile chronic arthritis (known as juvenile rheumatoid arthritis in the USA), is a heterogeneous group of systemic inflammatory disorders affecting children younger than 16 years.\(^2\,^3\) There are major subsets, separated on the basis of clinical features at onset which differ in their genetic associations and prognosis. Pauciarticular JCA comprises a group in which four or fewer joints are involved during the first six months of disease, and is the commonest form (55–75% of cases). It can be further subdivided into (at least) three separate types.
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Early onset pauciarticular JCA, which is by far the largest subgroup, includes those with large joint arthritis in the lower limb before the age of six years. Most cases do not show joint erosions; 40–75% are ANA positive, and 20–40% of ANA positive females get chronic iridocyclitis, progressing to ocular damage in 15%.

The second subgroup are older at presentation (over 8 years). Lower limb and large joint involvement, sacroililitis and enthesitis are typical, though it is not usual to detect the sacroiliitis until after the teenage years. Subjects tend to be ANA negative, but HLA-B27 positive.

The third subgroup may present at any time in childhood and involves both large and small joints asymmetrically. Often, psoriasis and psoriatic arthropathy develop in later life, and there may be a family history of this disease.

The development of chronic proliferative synovitis in a child may lead to local disturbance of bone growth, joint laxity with deformity and, occasionally, pannus mediated erosive disease.\(^3\)

**RADIOPHraphs**

Radiographs are the usual first test in the imaging of a patient with a swollen joint, performed to exclude non-inflammatory processes such as tumour, and it is important to interpret signs of an effusion correctly: the lateral radiograph of the ankle in the first patient showed the ‘tear drop sign’—a thickened anterior recess of the tibiotalar joint (fig 1). In the second patient, a lateral radiograph of the knee (fig 4) showed widening of the opaque ‘stripe’ of the suprapatellar bursa, which normally measures less than 5 mm.

**ULTRASOUND**

High resolution real time ultrasound is able to show synovial thickening or effusion in the ankle or knee and can be used to distinguish between them in the latter.\(^4\) This is achieved by compressing the suprapatellar bursa with the transducer, which causes the fluid (which is anechoic and conventionally displayed as black) to be displaced (figs 5, 6).\(^4\) It can give a measure of total synovial thickness, and may be used in patient follow up to assess response to intra-articular corticosteroids or other therapy.\(^1\)\(^,\)\(^4\)

In transverse section, the thickened and relatively echogenic synovium could be seen extending into the superior synovial plica within the anechoic suprapatellar bursa effusion (fig 5). In the ankle, fluid distension of the anterior recess produces an ultrasound equivalent to the tear drop, and tenosynovitis produces widening of the tendon sheath by hypoechoic tissue (fig 9).
MAGNETIC RESONANCE IMAGING

MRI was helpful in the diagnosis of the two patients presented and may have more general application in JCA. Synovial disease and cartilage erosions can be identified. However, both thickening of synovium and effusion have high signal (displayed white) on T2 weighted images and low signal (displayed dark grey) on T1 weighted images because of a high 'free' water content.

Distinguishing the synovial hypertrophy from fluid can be achieved by injecting IV Gd-DTPA: inflamed synovium has high signal on T1 weighted images after injection, whereas fluid remains relatively low in signal. In patient 1, IV Gd-DTPA enhanced images were the only ones to show that most of the anterior recess enlargement consisted of inflamed synovium. Similarly, there was enhancement of the tenosynovium around flexor hallucis longus. In the second patient, although it is possible to differentiate the synovial fluid from the surrounding inflamed thickened membrane on both T2 weighted and unenhanced T1 weighted images, the shape and extent of the synovial thickening was more obvious after IV Gd-DTPA (fig 8).

There are a number of additional advantages to the use of MRI contrast medium. First, in occasional cases of destructive disease, the synovial proliferation progresses from the osteochondral margins centrally, eroding the underlying cartilage and producing marginal defects in the bone which are filled with pannus. The extent of this disease is best assessed by intravenous gadolinium enhanced images. Second, it may help in showing loculation of effusion, which is important if intra-articular therapy is planned. Third, it helps in the diagnosis of hypoplastic menisci, implicated as a cause of joint instability in JCA, because they become covered by pannus.

MRI in PVNS is characterised by extensive frondlike areas of low signal on T1 and T2 weighted images, caused by the presence of haemosiderin. However, advanced cases of rheumatoid type arthritis may have haemorrhagic synovium that cannot be differentiated from PVNS. Large bone erosions may be a feature of late PVNS, and there is usually a Baker’s cyst filled with mass disease.

Conclusion

These cases illustrate the potential of imaging, notably MRI and ultrasound, to show synovial disease in children presenting with undiagnosed joint pain and to lead towards a diagnosis of inflammatory arthritis. In addition, it may be of value in assessing disease progression or the effectiveness of treatment.

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