

## Case report

# Synovial cysts in juvenile rheumatoid arthritis

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**SUMMARY** In a case of juvenile rheumatoid arthritis with large synovial cysts, cyst fluid aspiration was performed to relieve pain, but recurrence was prevented with salicylate therapy alone. The mechanism of formation of synovial cysts is discussed.

Involvement of the tendon sheaths and bursae is common in adults with rheumatoid arthritis (Palmer, 1969). However, in juvenile rheumatoid arthritis (JRA) involvement of the juxta-articular structures is less common (Brewer *et al.*, 1973). We present a case of juvenile rheumatoid arthritis and prominent multiple cystic swellings of tenosynovial origin.

### Case report

An 8-year-old black male had acute onset poly-articular juvenile rheumatoid arthritis at the age of 4 years. A satisfactory initial response was obtained with salicylates, but subsequent control was inadequate due to poor compliance with therapy. At age 7 he had acute exacerbation of JRA characterised by fever, rash, arthritis, pleurisy, pulmonary infiltrates, and generalised muscle weakness. The bicipital areas were swollen, and the overlying skin was taut and shiny. A gallium scan and serum enzymes (creatinine phosphokinase (CPK), aldolase, aspartate and alanine transaminases (SGOT, SGPT)) were normal. Skin and muscle biopsies from the left upper arm were normal. During this procedure, about 100 ml serous fluid was drained from a compartment overlying the muscle fascia. Cultures were negative. Salicylates failed to control this acute episode and indomethacin was added to the treatment, but had to be discontinued after a hypersensitivity reaction. Steroid therapy was recommended but refused by the mother. On salicylates alone (up to 150 mg/kg

per day), his condition remained essentially unchanged, with painful reaccumulation of fluid in the bicipital areas requiring frequent drainage.

Six months later he was referred to this hospital. Examination showed normal height and weight, temperature 38.9°C, and blood pressure 90/60 mmHg. An erythematous, evanescent, macular rash was present over the trunk, and several keloid scars corresponding to puncture and incision sites were noted over the bicipital areas. Tense and fluctuant swellings measuring 5 × 10 cm were detected in the same areas; slightly less prominent swellings were also noticed at the right wrist and ankle. There were no palpable nodules. Acute and chronic arthritic changes were evident in the cervical spine, temporomandibular, sternoclavicular, shoulder, elbow, wrist, knee, ankle, and interphalangeal joints. Ophthalmological slit-lamp examination was normal. X-rays of the affected joints showed narrowed joint spaces and bone demineralization but no bone destruction or abnormal soft tissue calcification.

Laboratory studies showed haemoglobin 9.3 g/dl, reticulocyte count 2.9%; haemoglobin electrophoresis normal, Coombs's test negative, and haptoglobin 2.93 g/l; leucocyte count 10 400/mm<sup>3</sup> (10.4 × 10<sup>9</sup>/l) with a normal differential count. Wintrobe ESR was 80 mm/h. PPD (5 TU) skin test was negative. Aldolase, SGOT, SGPT, and CPK were normal. Tests for rheumatoid factor (latex fixation), anti-nuclear antibodies, and cryoproteins were repeatedly negative. Serum showed diffuse hypergammaglobulinaemia (IgG 51 g/l IgA 6.2 g/l, IgM 1.97 g/l). Protein electrophoresis showed raised alpha-2 (1.1 g/l) and gammaglobulin fractions.

On the second day of admission, swelling of the left bicipital area had increased and was tense and

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Fig. 1 Synovial cyst of the shoulder joint in the bicipital area.

painful (Fig. 1). Needle aspiration yielded 55 ml blood-stained fluid. Through the same needle, 55 ml Urographin was injected for radiological studies (Fig. 2). Analysis of the cyst fluid showed low viscosity, good mucin clot, specific gravity of 1.035, protein 65 g/l, glucose 20 mg/100 ml (1.11 mmol/l) (blood glucose 80 mg/100 ml; 4.44 mmol/l), C3 0.29 g/l (blood C3 2.85 g/l), trace cryoglobulin, and no rheumatoid factor. Cytological analysis gave 31 000/mm<sup>3</sup> ( $31 \times 10^9/l$ ) leucocytes with 96% polymorphonuclear and 4% mononuclear cells. No neoplastic cells were seen in a cytocentrifuged sample. Bacterial (aerobic and anaerobic), mycobacterial, fungal, and viral cultures were negative.

The patient was given salicylate, 120 mg/kg per day, which produced levels of 25 to 31 mg/100 ml, with prompt clinical improvement. Brief non-compliance with therapy resulted in reaccumulation of fluid over the left bicipital area, requiring drainage of 35 ml turbid fluid which was again sterile. Subsequently he remained asymptomatic, with gradual normalisation of the sedimentation rate and serum proteins and no reaccumulation of fluid over a 10-month period of follow-up.

### Discussion

Tenosynovitis, bursitis, and abnormal fluid accumulation in the form of subcutaneous synovial cysts are frequently seen in adults with rheumatoid arthritis (Palmer, 1969). The best known among these



Fig. 2 X-ray showing contrast media (Urographin) flowing back into the shoulder joint along the long head of the biceps.

are the cysts of the popliteal area, or Baker's cysts (Baker, 1885). Adams first described a bursal cyst communicating with the knee joint in a case of rheumatoid arthritis (Adams, 1840). Since then, synovial cysts have been described in communication with virtually every joint (Gerber and Dixon, 1974). The joint cavity and the cyst usually communicate through a valvular mechanism that allows passage but not return of joint fluid into the cyst (Jayson and Dixon, 1970). Depending on their size, location, and the joint affected, bursal and synovial cysts have been found to simulate inguino-femoral hernias and thrombophlebitis of the calf (Samuelson *et al.*, 1971). They can exert pressure on the neurovascular structures (Palmer, 1969) and rupture into the adjacent tissues, causing painful irritation and tendon destruction (Hall and Scott, 1966).

Tenosynovitis, bursitis, and synovial cysts are presumably infrequent in JRA. Some major reviews make no specific reference to their occurrence (Calabro *et al.*, 1971; Schaller and Wedgwood, 1972;

Grossman and Mukhopadhyay, 1975), while others refer to tenosynovitis without mentioning its incidence (Ansell and Bywaters, 1963; Brewer, 1970; Brewer *et al.*, 1973). Stillman *et al.* (1976) reported a 13% incidence of tenosynovitis in JRA, with a predominance in the polyarticular group. When tenosynovitis is present in JRA it involves chiefly the tendon sheaths of wrists, hands, ankles, and feet. Synovial cysts of the size seen in our patient, and those communicating with the shoulder joint, have not been described in children with JRA.

Several mechanisms have been proposed to explain the formation of these large tenosynovial cysts (Coventry *et al.*, 1959). One theory claims that accumulation of synovial fluid increases intra-articular pressure, forcing the fluid out through valvular structures or through areas of least resistance created by the inflammatory process on the joint wall (Gerber and Dixon, 1974). The fluid is distributed along tendon sheaths and bursae that are in close proximity with the joint capsule. Because of the unique intra-articular course of the long head of the biceps, it seems that the shoulder joint fits perfectly into this mechanism. In fact, the shoulder joint is a frequent site of juxta-articular involvement in adults with rheumatoid arthritis (Gerber and Dixon, 1974). On the other hand, shoulder involvement is relatively uncommon in JRA (Grossman and Mukhopadhyay, 1975), hence the rarity of tenosynovitis of the shoulder in children. In our case, no valvular structure was evident between the tenosynovial cyst and the shoulder joint space, since the contrast media injected into the cyst flowed readily into the joint space.

Another theory proposes the independent formation of inflammatory fluid in the joint space and in the tendon sheaths or bursae (Gristina and Wilson, 1964). Communication between the inflamed joint and the juxta-articular structures may or may not occur. Finally, on rare occasions, the tendon sheaths or the bursae may swell in the absence of any noticeable joint pathology, and the inflammation may eventually find its way into the joint space (Bywaters, 1965).

In our patient, there were other less prominent swellings at the more usual sites, such as the ankles and wrists, in addition to the tenosynovial cysts of the biceps. These swellings did not require drainage, and they resolved with salicylate treatment. Control of the underlying rheumatoid activity frequently leads to resolution of these juxta-articular manifestations of the disease (Samuelson *et al.*, 1971). Occasionally, synovectomy of a recurrently affected

joint has been successful (Jayson *et al.*, 1972). Cyst fluid aspiration is indicated to relieve pain and local tissue destruction, but surgical removal or local steroid injection of the recurrent cysts have not been successful (Samuelson *et al.*, 1971).

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