Case report

A case of sarcoid arthritis in a child

A. L. THOMAS, C. S. THOMAS, J. A. DODGE, AND J. D. JESSOP

From the Departments of Rheumatology and Child Health, University Hospital of Wales, Heath Park, Cardiff CF4 4 XW

SUMMARY An 8-year-old girl presented with dyspnoea, weight loss, erythema nodosum, and an arthritis. A diagnosis of sarcoidosis with joint involvement was made. The clinical features of this unusual arthritis in a child and the response to corticosteroid therapy are described.

Sarcoidosis in children is relatively uncommon. The diagnosis is often made at a much later stage in the disease than in adults.¹ Studies in the United States have shown that the disease is more common in black children than Caucasians, reflecting the relative incidence in adults in the same area.² Joint involvement in children is rare, with only 13 cases being previously reported.³⁻⁶ A case of sarcoid arthritis is described in a Caucasian child with many of the distinctive clinical features of the disease.

Case report

A girl aged 8 years 10 months of Welsh descent was seen in February 1981 at the University Hospital of Wales with an 8-month history of weight loss and shortness of breath together with painless knee and ankle effusions for 18 months. She was the fourth of 5 children, and her parents and siblings were alive and well. She had been in good health and had developed normally until the age of 4½ years, when she developed persistent erythema nodosum, predominantly on the legs but also on the arms. She was otherwise well, and repeated blood counts and antistreptolysin titres, erythrocyte sedimentation rates, and chest x-rays remained normal. Serum rheumatoid factor was never found, and Mantoux tests were repeatedly negative.

In June 1979 she developed painless effusions in her knees and later her ankles. There was no therapeutic response to salicylates. One year later, now aged 8 years, she had early finger clubbing, her liver and spleen had become palpable, and her elbow

joints were now transiently involved. During the next 8 months her general condition deteriorated, with a 6 kilogram weight loss; she became short of breath on exercise and developed a dry unproductive cough.

At this stage she was referred to the Department of Child Health at the University Hospital of Wales for further investigations. Physical examination showed that her weight was on the 10th and height on the 50th percentile respectively. She had grade IV finger clubbing, her skin was normal, with no evidence of erythema nodosum, and her eyes showed no abnormality on slit-lamp examination. She was dyspnoeic at rest but not cyanosed. No abnormality was found in her cardiovascular system. Her respiratory rate was 32 per minute; chest expansion was poor, but there were no abnormal findings on auscultation. Her liver was enlarged 4 cm and the spleen 2 cm. No neurological signs were detected. There were warm, nontender effusions of both her knee (Fig. 1) and ankle joints, with boggy synovial proliferation and thickening of adjacent synovial sheaths. Joint movement was full and painless, and her other joints were normal.

Laboratory tests showed haemoglobin 12.5 g/dl, leucocytes $6.9 \times 10^9/l$, ESR 8 mm/h, liver function tests normal, IgM 0.2 g/l, other immunoglobulins normal. Serum rheumatoid factor, antinuclear antibody, smooth muscle antibody, and mitochondrial antibodies were not present. Chest x-ray showed bilateral hilar and right paratracheal lymphadenopathy with a 'ground-glass' appearance throughout both lung fields (Fig. 2). X-rays of knees and ankles revealed effusions with extensive periarticular soft tissue shadowing but no erosions or cartilage loss. A marked restrictive pattern was recorded on respiratory function tests, PFR 115 l/min, FEV₁

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Correspondence to Dr J. D. Jessop, Department of Rheumatology, University Hospital of Wales, Heath Park, Cardiff CF4 4XW.

0.35 l, FVC 0.48 l. Serum angiotensin converting enzyme level was markedly elevated at 165 IU/l (the upper limit of normal at our laboratory for a child of her age is 65 IU/l). 20 ml of inflammatory synovial fluid with a protein content of 35 g/l and negative rheumatoid factor were aspirated from each knee. Liver biopsy showed noncaseating granulomata typical of sarcoidosis (Fig. 3).

Fig. 1 Knees, showing effusions and boggy synovial thickening.

She was treated with oral prednisolone 20 mg daily, with a prompt beneficial response, and the dosage was gradually reduced to a maintenance dose of prednisolone 10 mg on alternate days at the end of 3 months. At this time examination showed her to have some cushingoid features, grade I finger clubbing, liver enlarged 2 cm, and impalpable spleen. There were no abnormal signs in her joints. The chest x-ray appearances had returned to normal, serum angiotensin converting enzyme level was within the normal range at 29 IU/l, and repeat respiratory function tests showed an improvement with PFR 205 l/min, FEV₁ 0.5 l, FVC 0.75 l.

Nine months later the patient was well and maintaining her growth velocity and had no dyspnoea on exercise or at rest. Episodes of erythema nodosum had persisted, lesions appearing every 3/4 weeks. Apart from pain in her left second toe there were no other joint symptoms. She was mildly cushingoid, finger clubbing remained grade I, but there was no lymphadenopathy, and her eyes were normal. There were no chest signs, liver and spleen were impalpable. She had minimal synovial swelling of her right ankle, the terminal interphalangeal joint of the left second toe was swollen but not tender. All other joints were normal. Her haemoglobin was 12 g/dl, ESR 14 mm/h, serum angiotensin converting enzyme 31 IU/l. X-rays of her feet were normal, as was her chest x-ray. Respiratory function tests had continued to improve but still showed a restrictive pattern, PFR 270 l/min, FEV₁ 1.1 l, FVC 1.25 l.

Fig. 2 Chest x-ray with bilateral hilar and right paratracheal lymphadenopathy; pulmonary infiltration.

Fig. 3 Liver biopsy showing a noncaseating granuloma. (Haematoxylin and eosin, $\times 133$).

Discussion

Sarcoidosis is relatively uncommon in children and the true incidence is difficult to assess. In Japan, where routine chest x-ray screening is common, many more cases of children with asymptomatic bilateral lymphadenopathy are discovered.⁷ Erythema nodosum and hilar lymphadenopathy are rare presenting features. To date only 331 cases of childhood sarcoidosis have been reported, but only 13 had joint involvement,⁶ all of whom presented below the age of 5 years. Children with sarcoid arthritis rarely have pulmonary disease but typically have uveitis, localised cutaneous lesions, and a distinctive arthritis with mild constitutional symptoms. The arthritis is persistent, nondestructive, and affects predominantly the large joints. It is characterised by large, painless, boggy synovial proliferation and tendon sheath effusions with little limitation of movement and normal x-ray appearances.³

Our case presented with this typical arthritis and also with features found more commonly in older children with sarcoidosis, namely, weight loss, cough, bilateral hilar lymphadenopathy and parenchymal lung changes. Finger clubbing has not been described in childhood sarcoidosis but is becoming increasingly recognised as a feature of adult disease, particularly where there is lung involvement.⁸

Childhood sarcoid arthritis may be mistaken for juvenile chronic arthritis (JCA), particularly when it is associated with uveitis. There is no way of distinguishing uveitis occurring in the 2 conditions, but antinuclear antibodies are found in 88% of patients with JCA and uveitis⁹ and are absent in sarcoidosis.

Erythema nodosum is a feature of inflammatory bowel disease but our patient had no bowel involvement. The other features of this case, namely, the marked degree of hepatosplenomegaly, minimal loss of joint function, and lack of radiographic change after an unremitting course for several years, involvement of tendon sheaths, lack of response to salicylates, and finally lung involvement are typical of childhood sarcoid arthritis, but none is pathognomonic. Estimation of serum angiotensin converting enzyme can be useful both in the diagnosis of active sarcoidosis and in following the response to treatment.

There have been no long-term studies of children with sarcoid arthritis. Those studies of children with sarcoidosis but without joint involvement reveal a poorer prognosis in those with extrathoracic disease.¹¹ It is encouraging that this patient has made a good clinical, radiological, and biochemical response to steroid therapy and has not at any time showed evidence of uveitis, but there is still clinical evidence of disease activity. The prognosis must therefore remain guarded.

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