

Long term evolution of adult onset Still's disease seen in an infectious diseases department

Sir: Adult onset Still's disease was first reported by Bywaters¹ in 1971, who stated that the long term articular prognosis was good. Several studies have shown, however, that severe joint lesions do occur and often leave patients with severe sequelae.²⁻⁵ Cabane *et al*⁶ found that the incidence of hip disease was 50% and that hip replacement was required in seven out of 18 cases. This letter reports the long term outcome of 10 patients with adult onset Still's disease who presented to an infectious diseases department and were followed up for more than three years.

All 10 patients were more than 16 years old (mean 36 years (range 20-56)) and fulfilled the American Rheumatism Association criteria for adult onset Still's disease⁶; in eight patients all four major criteria (fever, arthritis, rash, leucocytosis) were present and the diagnosis was definite. Diagnosis was only probable in two patients because of the absence of the characteristic rash. Arthritis most commonly affected the knees (seven patients), wrists (four), and ankles (two), and was monoarticular (six) or pauciarticular (four) in distribution. The patients were followed up for a mean of 8.9 years (range 3-17).

Long term outcome was assessed according to the disease activity scale defined by Cush *et al*² and is reported in the table. Systemic disease activity was the dominant aspect of the illness in eight out of 10 patients in whom articular involvement was mild or absent. Five of these eight patients had a single episode of systemic disease followed by clinical and biological remission. Currently, they have discontinued treatment after a mean duration of 20 months and a mean follow up period of six years. Three of these eight patients had a polycyclic systemic disease pattern defined by multiple episodes of systemic disease without severe articular involvement. Chronic articular disease was identified in two other patients with polycyclic systemic disease superimposed on persistent peripheral arthritis. Joints in these two patients, including shoulders, wrists, and knees, were affected with asymmetric polyarticular disease. It should be noted that the hip joints were spared. The patients were negative for rheumatoid factor. Radiographs showed moderate subchondral erosions without narrowing of the articular space. Chronic arthritis in these two patients was controlled by a methotrexate regimen. The first patient was treated with this drug for two years (15 mg weekly) and is now asymptomatic, 2.5 years after discontinuation of all treatment. The methotrexate regimen was recently started by the second patient and is controlling the arthritis. Corticosteroid

treatment alone had been ineffective in both these patients. Articular destruction was moderate in both patients and arthrodesis or articular prostheses were not required. Finally, the outcome for all 10 patients was favourable and they remained in functional class I of the American Rheumatism Association functional status classification.

Our observations confirm the favourable prognosis of the systemic manifestations of adult onset Still's disease. Furthermore, they suggest that the prognosis of the articular manifestations may also be favourable. This is not consistent with several earlier studies.³⁻⁵ This discrepancy might be due to differences in the way in which the patients were recruited. Our study group comprised patients admitted to an infectious diseases department owing to prolonged fever of unknown origin without prominent articular signs. Previous studies used groups of patients admitted to departments of internal medicine or rheumatology. Cush *et al*² indeed showed that patients with mild articular manifestations at presentation tend to have a considerably better functional status after prolonged follow up than those with more severe articular manifestations. The shorter follow up periods in this study, as compared with previous studies, might explain the more favourable articular prognosis suggested by our results. Nevertheless, in the group described by Cabane *et al*⁶ joint destruction appeared between one and eight years after the diagnosis and hip destruction in less than four years. In contrast, no hip disease or marked articular destruction were found in our patients with a mean follow up of 8.9 years.

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Septic arthritis due to *Nocardia caviae*

Sir: A 75 year old man from a nursing home with past medical history of osteoarthritis, mild diet controlled non-insulin dependent diabetes mellitus, and hypertension was admitted in October 1988 with loculated left pleural effusion and infiltrates in the left lower lung field. Despite extensive evaluation no definite cause could be found. The patient was treated intravenously with antibiotics and was discharged after his condition stabilised.

In January 1990, while he was in hospital after peripheral vascular surgery, he developed left knee joint pain. Physical examination at that time showed an elderly, moderately built man with blood pressure 130/70, pulse rate 84/min, temperature 37.5°C. Chest examination showed decreased breath sounds and dry rales at the left base. There was a grade II/VI systolic ejection murmur at the right upper sternal border with regular heart rhythm. Examination of the left knee showed heat, marked tenderness, decreased range of motion, and moderate synovial effusion. The aspirated joint fluid was purulent and had a rosy colour. The white blood cell count showed 415 × 10⁹ cells/l, of which 93% were neutrophils. Empirical treatment was started with nafcillin intravenously 6 g/day. After three days culture of this fluid yielded *Nocardia caviae*. Drug treatment was switched to trimethoprim 160 mg-sulphamethoxazole 800 mg every 12 hours orally with subsequent addition of amoxicillin-clavulanate 500 mg every eight hours orally. Blood and sputum cultures were negative. The chest radiograph was stable and showed persistent minimal left pleural effusion with pleural reaction, which had not changed for 14 months. An x ray examination of the left knee showed degenerative changes, soft tissue swelling with joint effusion, and no sign of osteomyelitis.

Left knee arthrocentesis was performed periodically to monitor the progression of the disease. The leucocyte count in the synovial fluid gradually decreased with change of colour to opaque yellow despite persistent growth of *N. caviae*. After six weeks of antibiotic treatment the synovial fluid white cell count was 4.9 × 10⁹ cells/l and it was sterile. The patient's symptoms and signs of septic arthritis gradually improved. Treatment with amoxicillin-clavulanate was continued for a total of 12 weeks. Trimethoprim-sulphamethoxazole was discontinued after eight weeks because of a minimal rise in blood urea and creatinine. No further synovial fluid accumulation has been noted, and the patient has continued to have a functionally stable left knee.

Nocardia caviae is an uncommon cause of human disease, usually seen in immunocompromised hosts. Although septic arthritis caused by *N. asteroides*¹⁻³ and *N. brasiliensis*⁴ has been described, nocardial arthritis due to *N. caviae*, to our knowledge, has not been previously reported.

The predisposing factors for nocardial infection in the patient described here were mild, diet controlled non-insulin dependent

Treatment and outcome of 10 patients with adult onset Still's disease followed up for more than three years

Case No	Duration of follow up (years)	Treatment*	Duration of initial treatment (months)	Outcome
1	8	CS, MTX	12	Chronic articular
2	14	CS, MTX	6	Chronic articular
3	6	CS	36	Polycyclic systemic
4	17	CS	8	Polycyclic systemic
5	15	CS	180	Polycyclic systemic
6	3	CS	5	Monocyclic systemic
7	7	CS	30	Monocyclic systemic
8	5.5	CS, ASA	30	Monocyclic systemic
9	5.5	CS	30	Monocyclic systemic
10	8	ASA	6	Monocyclic systemic

*ASA=acetylsalicylic acid; CS=corticosteroid; MTX=methotrexate.