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 Bywaters in 1971, who stated that the long term articular prognosis was good. Several studies have shown, however, that severe joint erosions do occur and often leave patients with severe sequelae. Cabane et al 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 36.2, range 20–56) and fulfilled the American Rheumatism Association criteria for adult onset Still's disease; in eight patients all four major criteria (fever, arthritis, rash, leucocytosis) were present and the diagnosis was considered to be only probable in two patients because of the absence of the characteristic rash. Arthritis most commonly affected the knees (seven patients), wrists (four patients), and ankles (two), and was monarticular (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 presentation defined by Cush et al 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 disease pattern who had persistent peripheral arthritis. Joints in these two patients, including shoulders, wrists, and knees, were affected by asymmetrical 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 arthroplasties 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. 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 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 in our study, compared with previous studies, might explain the more favourable articular prognosis suggested by our results. Nevertheless, in the group described by Cabane et al 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.

BERTRAND GOEDEAU
CATHERINE LEPorT
CHRISTIAN PERRonne
DOMINIQUE MON-CERON
JEAN-LOUIS Vilde
Department of Infections and Tropical Diseases
Bichat/Claude Bernard Hospital
Paris, France

MARCEL FRANS KAHN
Department of Rheumatology
Bichat/Claude Bernard Hospital
Paris, France

Correspondence to: Dr B Godeau, 68 Bd Arago, 75013 Paris, France.


Septic arthritis due to Nocardia caviae

Sir: A 75 year old man from a nursing home with past medical history of osteoarthritis, diabetes mellitus, and hypertension was admitted in October 1988 with localized left pleural effusion and infiltrates in the lower left lung field. Despite extensive evaluation no definitive cause could be uncovered. 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 a dry cough and normal breath sounds with no rales at the 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 not continued due to the high risk of infection. Cultures of amoxicillin/clavulinate 500 mg every 8 hours orally. Blood and sputum cultures were negative. The chest radiograph was stable and showed persistent minimal left pleural effusion with no 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 sepsis. The patient had no history of chronic kidney disease.

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-clavulinate 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 and N brasiliensis has been described, nocardial arthritis is rare. In our knowledge, it has not been previously reported.

The predisposing factors for nocardial infection in the patient described here were mild, diet controlled non-insulin dependent diabetes mellitus, and hypertension which was treated with diet and oral hypoglycaemic agents. The patient had no history of peripheral vascular disease.
diabetes mellitus and hypertension, inactive lifestyle at a nursing home with chronic osteoarthritis and minimal persistent left pleural effusion of over nine years duration and of uncertain cause. The patient had also been treated for herpes zoster infection about one year before the development of nocardial arthritis. Possibly, therefore, he might have had a subnormal immune defence mechanism against opportunistic infections. The patient's chest radiograph showed marked improvement in pleural disease after treatment of the arthritis with trimethoprim-sulphamethoxazole-amoxicillin-clavulanate. This suggests that the portal of entry was the respiratory tract, and the patient might have had chronic pleural disease with culture negative nocardial infection.

Successful treatment of nocardial septic arthritis seems to require prolonged antibiotic treatment for three to six months or longer. From among all the antibiotics tested in vivo and in vitro, the use of trimethoprim-sulphonamide combination has been of the most consistent success. There is some evidence to suggest that amoxicillin-clavulanate may be useful in the treatment of nocardiosis. In summary, septic arthritis due to _Nocardia caviae_ occurred in an elderly patient with multiple underlying diseases and was successfully treated with trimethoprim-sulphonamide-amoxicillin-clavulanate combination antibiotic regimen. To our knowledge, _N caviae_ arthritis has not been previously reported. Nocardial infection must be considered when undiagnosed, longstanding pleuropneumonic disease persists in an immunocompromised patient who later develops septic arthritis. Finally, although it requires the long term use of antibiotics, septic arthritis due to _N caviae_ can be treated successfully.

N P TORRE B K KIM
Sisters of Charity Hospital
2157 Main Street
Buffalo, New York 14214
USA

**Letters to the editor**

Female: male sex ratio among family members with primary Sjögren’s syndrome

<table>
<thead>
<tr>
<th>Women</th>
<th>Men</th>
<th>χ² or Fisher’s test</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Index cases (n)</td>
<td>38</td>
<td>2</td>
<td>20.3</td>
</tr>
<tr>
<td>Total relatives (n)</td>
<td>150</td>
<td>74</td>
<td>13.3</td>
</tr>
<tr>
<td>Siblings in study (n)</td>
<td>49</td>
<td>37</td>
<td>2.5</td>
</tr>
<tr>
<td>Offspring of probands (n)</td>
<td>19</td>
<td>22</td>
<td>0.11</td>
</tr>
</tbody>
</table>

*Refers to the potential pool of relatives.*


**Figure 1** Technetium-99m labelled nanocolloid scintigram. Accumulation in the joints and along the tenosynovial sheath of the fourth left finger.
Septic arthritis due to Nocardia caviae.

N P Torre and B K Kim

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