Lesson of the Week

Felty's syndrome presenting without arthritis

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Felty's syndrome classically consists of the triad of rheumatoid arthritis, splenomegaly, and neutropenia, often with additional features such as weight loss, lymphadenopathy, and leg ulceration. We describe three patients in whom neutropenia preceded clinical evidence of arthritis thus delaying the correct diagnosis. In one patient the condition has remained asymptomatic, and in another articular disease became apparent 10 years after presentation with neutropenia.

Case reports

Case 1—A 73 year old woman was found to have neutropenia when admitted to hospital with postherpetic neuralgia. Four years earlier, an unexplained erythrocyte sedimentation rate of 120 mm in first hour and a high titre of rheumatoid factor (1/2560) had been detected. Investigations showed: haemoglobin concentration of 11·7 g/dl, white cell count $3{\cdot}0 \times 10^9$/l, platelets 224×10^9/l, erythrocyte sedimentation rate 38 mm in first hour, Rose Waaler test positive, and antinuclear antibody test negative. The bone marrow showed a low leucoerythrogenic ratio, and a radioisotope scan showed splenomegaly. There was no evidence of arthritis, but on reassessment seven months later asymptomatic swelling of several metacarpophalangeal joints was noted. Investigations confirmed the presence of leucopenia (white cell count $2{\cdot}1 \times 10^9$/l, 32% neutrophils) and rheumatoid factor; *x* ray examination showed erosions in wrists, fingers, and toes.

Case 2—A 57 year old woman presented with carpal tunnel syndrome and painless swelling of the ipsilateral extensor tendon sheath. The spleen was not palpable and the joints appeared normal. Investigations showed haemoglobin concentration of 14·9 g/dl, white cell count $3{\cdot}0 \times 10^9$/l (18% neutrophils). Results of the Rose Waaler test were positive (titre of 1/256) and of antinuclear antibody test were weakly positive. Joint radiographs showed no abnormality. The bone marrow was mildly hypocellular with increased granulocyte precursors. Synovium removed at the time of carpal tunnel decompression one year later showed typical histological appearances of a rheumatoid nodule. Two years after her initial attendance she developed a mild polyarthritis with splenomegaly, the white cell count remaining low at $2{\cdot}7 \times 10^9$/l. *X* ray films now showed small erosions of the metatarsophalangeal joints. Six months later the spleen was no longer palpable and the white cell count had risen to $4{\cdot}7 \times 10^9$/l (49% neutrophils). It has remained normal for three years.

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Case 3—A 72 year old woman developed a febrile illness and on investigation was found to have haemoglobin concentration of 12·8 g/dl, white cell count $2{\cdot}0 \times 10^9$/l (11% neutrophils), and erythrocyte sedimentation rate 62 mm in first hour. Although the neutropenia persisted no cause was found, but 10 years later she developed widespread joint pain. Examination now showed a symmetrical polyarthritis with splenomegaly, and a latex test for rheumatoid factor was positive. The bone marrow was normal. Five years later she had a persistent neutropenia and mild arthritis accompanied by a mild peripheral neuropathy. Radiographs showed small erosions of the hands.

Comment

These patients were striking because of their mild, delayed, and, in one case, absent joint symptoms despite subsequent radiological evidence of joint erosions. They responded to simple anti-inflammatory agents, none requiring treatment with so called "second line" drugs. In no case was an alternative cause for the neutropenia discovered, and, in particular, drugs could not be implicated.

Usually, articular manifestations of Felty's syndrome are considered to be more severe than in uncomplicated rheumatoid arthritis.[1] Nevertheless, although some reports describe advanced joint destruction radiologically, the arthritis is often clinically quiescent.[2 3] Such observations suggest that the neutropenia may be related to a reduction of synovitis. Thus when Felty's syndrome occurs early in the course of rheumatoid disease it may coexist with mild arthritis. This may explain why arthritis was for so long absent or undetected in our patients.

Although neutropenia and splenomegaly only rarely precede all evidence of articular disease,[1] our report emphasises the need to consider Felty's syndrome as a cause of neutropenia even in the absence of joint symptoms.

References

[1] Sienknecht CW, Urowitz MB, Pruzanski W, Stein HB. Felty's syndrome. Clinical and serological analysis of 34 cases. *Ann Rheum Dis* 1977;**36**: 500-7.
[2] Ruderman M, Miller LM, Pinals RS. Clinical and serological observations on 27 patients with Felty's syndrome. *Arthritis Rheum* 1968;**11**:377-84.
[3] Barnes CG, Turnbull AL, Vernon-Roberts B. Felty's syndrome: a clinical and pathological survey of 21 patients and their response to treatment. *Ann Rheum Dis* 1971;**30**:359-74.
[4] Heyn J. Non-articular Felty's syndrome. *Scand J Rheumatol* 1982;**11**: 47-8.