Introduction

Jaccoud was a French clinician from the 19th century who reported a peculiar joint deformity in patients with rheumatic fever [1]. He observed the development of an ulnar deviation that was reducible, which indicated that the joint spaces were not destroyed. We have no evidence that Jaccoud was interested in the kidney or that he ever saw a patient with nephritis. Perhaps the patient reported here would have raised his interest in our specialty.

Case

A 69-year-old woman was admitted because of generalized weakness and decreasing renal function. Two months earlier, her creatinine had been normal. Three months before admission, she developed an upper respiratory tract infection. Rheumatoid arthritis was diagnosed 20 years earlier and had been treated with corticosteroids and methotrexate. This treatment was terminated 10 years prior to admission. Pertinent physical findings included her hands (Figure 1). At rest, she had bilateral ulnar deviation with swan neck deformities. However, synovial thickness, redness or arthropathy were not evident. As a matter of fact, the ulnar deviation could be reduced, which she could also do voluntarily (Figure 2). She had telangiectasias on her chest and livedo reticularis on the upper legs and back.

The haemoglobin was 8.9 g/dl, the haematocrit was 28 vol%. The leucocyte count was 7500/μL; the platelets numbered 399 000/μL. The C-reactive protein was 144 mg/l. The creatinine concentration was 481 μmol/l, the urea concentration was 20 mmol/l, total proteins were 73 g/l and the albumin concentration was 30 g/l. On protein electrophoresis, no M-gradient was present and no monoclonal paraproteins were detected in serum and urine. Nevertheless, she had 1.8 g/day proteinuria. The urine showed acanthocytes and erythrocyte casts. The roentgenograms of her hands showed no metacarpal destruction and reduction of the deformities was possible. The terminal thumb digital joints showed osteoarthritis, but not rheumatoid arthritis.

Question

Dr Jaccoud would have diagnosed rheumatic fever. Can you help him make a better diagnosis?

We thought the patient had systemic lupus erythematosus. The anti-nuclear autoantibody (ANA) titres were 1:2560 (HEp2 indirect immunofluorescence assay) and double-stranded DNA antibody titres were 1:40 (normal range <1:10), verified by a *Chritidia luciliae* assay. The ELISA for double-stranded DNA antibodies was 72 U/ml (normal range <101), for antibodies against nucleosomes 197 U/ml (normal range <20), for antibodies against centromeres 0.1 (normal range: ratio <1.1) and for antibodies against Scl-70 0.1 (normal range: ratio <1.1). Perinuclear antineutrophil cytoplasmatic antibody (p-ANCA) titre was 1:64 (ELISA). The antibodies had specificity for myeloperoxidase (MPO).

Fig. 1. Ulnar deviation of four digits from each hand. The thumbs show a deviation in the opposite direction. The fingers 3–5 also show the presence of swan neck deformity. Nevertheless, the metacarpophalangeal joints show no evidence of destructive arthropathy as would be expected in rheumatoid arthritis.
ANCA with typical cytoplasmatic stains (c-ANCA) were negative in ELISA. Complement factor C1q was 93 mg/l (normal range 122–208 mg/l), complement factor C3 0.7 g/l (normal range 0.79–1.52 g/l) and C4 0.175 g/l (normal range 0.180–0.490 g/l). Anticardioloopin-IgG was 99.7 kU/l (normal range < 48), Anticardioloopin IgM was 14.8 kU/l (normal range < 44), and beta2-glycoprotein-I antibody was 2.2 kU/l (normal range < 5.1). Lupus anticoagulant was negative. A renal biopsy showed a proliferative glomerulonephritis, crescent formation, small vessel vasculitis, round cell infiltrates and fibrinoid necrosis of the blood vessels (Figure 3). The immunofluorescence disclosed a pauci-immune staining pattern (data not shown).

What is your diagnosis?
Answer to the question on the preceding page

Pauci-immune rapidly progressive anti-MPO glomerulonephritis with incidental Jaccoud’s arthritis.

Comment

Our patient had abnormal antinuclear antibody titres, antibodies to double-stranded DNA, arthritis of both hands, and acute glomerulonephritis. We were confident she had systemic lupus erythematosus; however, the significant p-ANCA titre gave us cause for reflection. Fifty years ago, Bywaters [2] pointed out the similarities between the arthritis reported by Jaccoud and the joint deformations that he observed in patients with systemic lupus erythematosus. Bywaters was of the opinion that Jaccoud’s arthritis is not uncommon in systemic lupus erythematosus patients. In contrast to rheumatoid arthritis in which a destructive arthropathy occurs, Jaccoud’s arthritis features synovitis with capsular fibrosis without articular destruction. Our patient had four finger bilateral ulnar deviation of >20°, swan-neck deformities of three fingers, and a Z deformity of both thumbs. With these criteria, our patient scores eight points on a scale requiring five points to make the diagnosis of Jaccoud’s arthritis [3].

Myeloperoxidase antibodies, p-ANCA, occur in ~0–20% of patients with systemic lupus erythematosus [4]. We are aware of only one report in which a convincing case for systemic lupus erythematosus was made in a patient with pauci-immune nephritis [5]. Conceivably, ANCA may initiate segmental necrotizing and crescentic glomerulonephritis without subendothelial deposits in a few patients with systemic lupus erythematosis [5,6]. Possibly, our patient had systemic lupus erythematosis and associated p-ANCA. The latter then might have initiated crescentic glomerulonephritis on the basis of an immune complex-independent cell-mediated action. Important in this case is the appreciation of Jaccoud’s arthritis, which in nephrological circles should generally suggest systemic lupus erythematosus.

Conflict of interest statement. None.

References


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