Concomitant systemic lupus erythematosus and ankylosing spondylitis

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Abstract

The case is reported of a 42 year old white woman meeting currently used diagnostic criteria for both ankylosing spondylitis and systemic lupus erythematosus (SLE). As found in a previously described similar case of a black man, HLA typing showed antigens associated with both SLE and seronegative spondyloarthritis. This case thus supports the hypothesis that the two diseases occur together only when this rare combination of HLA antigens is present.

The coexistence of systemic lupus erythematosus (SLE) and seronegative spondyloarthritis is very rare. Only a few cases have been reported.

We describe here another case of this coexistence. As in the case reported by Nashel et al., our patient showed an unusual combination of HLA antigens associated with both seronegative spondyloarthritis and SLE.

Case report

A 42 year old white woman was admitted to our hospital in January 1986. About one year before, SLE had been diagnosed and treatment started with corticosteroids and chloroquine. Her medical history showed episodes of sciatic pain during the postpartum period in 1979, episodes of inflammatory low back pain and stiffness in the past five years, recurrent episodes of arthritis of the knees, ankles, and wrists and of Achilles tendinitis between 1980 and 1983, malar rash, oral ulcers, alopecia, and Raynaud’s phenomenon since 1983.

Her family history was negative for both SLE and seronegative spondyloarthritis. Physical examination showed the classic butterfly rash over the cheeks and the bridge of the nose, asymptomatic mouth ulcers on the hard and soft palate, alopecia, digital vasculitis and tenderness on both sacroiliac joints and both Achilles tendons. Lumbar and cervical spine movement was normal. Chest expansion was 2.5 cm.

Investigations showed erythrocyte sedimentation rate (Westergren) 25 mm/1st h, C reactive protein 36 mg/l (normal <5 mg/l), positive antinuclear antibody test (1/640, diffuse pattern), positive anti-double-stranded DNA antibody, reduced serum complement concentrations with a total haemolytic complement (CH50) 150–210 U/ml, C4 10 mg/l (normal 200–400 mg/l), and C3 430 mg/l (normal 550–1200 mg/l). Tests for the rheumatoid factor, the anti-extractable nuclear antigen antibody, and the Venereal Disease Research Laboratory test were negative. HLA typing showed A2, A24, B18, B27, CW2, DR3, DR7, DQw2, and DQw3 antigens.

Immunofluorescent studies (lupus band test) disclosed deposits of gammaglobulin and complement components at the dermal-epidermal junction.

An anteroposterior view (fig 1) of the pelvis showed grade 3 bilateral sacroiliitis. Lumbar and dorsal spine radiographs showed subchondral osteitis of the upper corner of the last dorsal vertebrae and of the first lumbar ones. Radiographs of the cervical spine were normal. The lateral view of the calcaneus showed spurs at the insertion of both Achilles tendons.

Prednisone was increased from 6 to 12 mg/day. Subsequently this dose was tapered to 5 mg/day.

In July 1987 the patient had proteinuria and was admitted to our institute again for a renal biopsy.

Laboratory evaluation showed erythrocyte sedimentation rate 36 mm/1st h, antinuclear antibody strongly positive with a diffuse pattern, anti-dsDNA antibody positive, CH50 98 U/ml, C4 60 mg/l, C3 380 mg/l, 24 hour urine protein concentration 4.5 g, and normal serum creatinine concentrations.

A light microscopy study of the renal biopsy specimen (fig 2) disclosed findings of diffuse proliferative lupus nephritis, including mesangial and endothelial cell proliferation, ‘wire loops’, epithelial crescents, nuclear pyknosis and karyorrhexis, and infiltration by polymorphonuclear leucocytes.

Under immunofluorescence diffuse granular deposits of IgG, IgM, C3, and C1q were seen along the basement membrane and the capillary wall.

The patient was given 40 mg/day prednisone. This dose was gradually reduced to the current dose of 5 mg/day.

Discussion

Our patient meets both the New York criteria for ankylosing spondylitis and the American Rheumatism Association criteria for SLE.

The coexistence in the same patient of SLE and seronegative spondyloarthritis is much less than the expected concurrence calculated on the basis of the prevalence of each of them in the general population. Only a few cases, to our knowledge, have been reported. Some authors have reported a high prevalence of sacroiliitis in male patients with SLE, but none of these...
In 1982 Nashel et al reported the case of a 43 year old black man affected by both ankylosing spondylitis and SLE. HLA typing showed A1, A2, B8, B27, C3, DR2, and DR3 antigens. The authors suggested that the rarity of the coexistence of SLE and ankylosing spondylitis in the same patient might depend on the fact that the B27 antigen rarely exists in combination with the DR2 or DR3 antigens, which are associated with SLE, as confirmed by studies of haplotype frequencies in American black subjects.

Our patient, like that of Nashel et al, showed both the B27 antigen, which is associated with ankylosing spondylitis and the other seronegative spondarthritides, and the DR3 antigen, which is associated with SLE. Studies of haplotype frequencies have shown that in white subjects also the B27 antigen exists only exceptionally with DR3.

Our report confirms the hypothesis of Nashel et al that SLE and seronegative spondylarthropathy coexist only when a rare combination of genetically determined markers occurs.1


13 Källman M, Nylen O, Hansén M. Evaluation of quantitative scintigraphic sacroiliitis in patients with active SLE was reported by De Smet et al. The value of quantitative scintigraphy of the sacroiliac joints in detecting early sacroiliitis before the appearance of radiological changes is controversial, however.13 14
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