Unusual association of a positive pANCA pauci-immune extracapillary glomerulonephritis in a lupus patient

Sir,

We report a case of pauci-immune proliferative and crescentic glomerulonephritis in a 22-year-old female with systemic lupus erythematous (SLE) who was admitted because of a rapidly progressive glomerulonephritis syndrome. On admission, the patient had generalized edema, a blood pressure of 130/80 mmHg and anuria. There were no extra-renal signs.

Investigations showed a serum creatinine of 15.2 mg/ dl, hemoglobin of 6.9 g/dl, a lymphopenia (900/cumm), low serum complement C_3 concentration of 20 (50- 120 mg/ dl), positive antinuclear antibodies at 1/640 with homogenous fluorescence, and positive anti-DNA antibodies >300 UI/ml.

Renal histology included 23 glomeruli, there was extracapillary proliferation with circumferential crescents in 13 glomeruli and one obsolescent glomerulus [Figure 1].

Immunofluorescence (IF) examination of the kidney biopsy did not reveal any significant deposit of immunoglobulins and complement. Anti-myeloperoxidase (MPO) antibodies were positive at 212 UI/ml by enzyme-linked immunosorbent assay (ELISA).

Immunosuppressive treatment included corticosteroids and intravenous cyclophosphamid. The renal evolution was characterized by a persistent anuria for 45 days. A progressive amelioration of renal function led to a

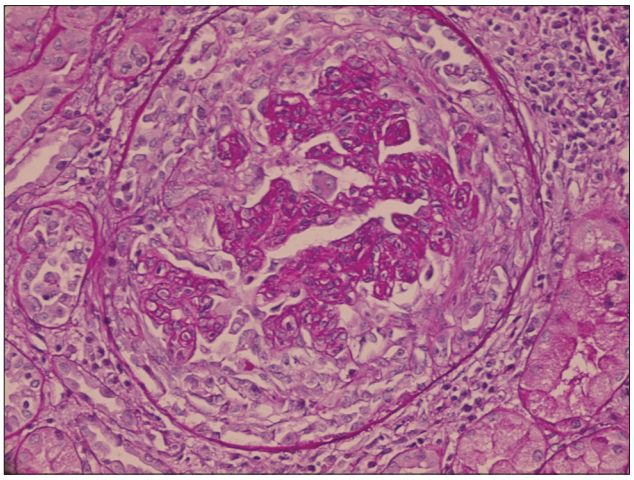

Figure 1: Circumferential extracapillary crescent with flocculus crushing (H and E, ×400)

serum creatinine of 1.7 mg/dl one year after initial treatment with a persistent proteinuria of 2. 4 g/24 hours.

Approximately 20% of patients with SLE have antineutrophil cytoplasmic autoantibodies (ANCA) positivity by indirect IF (IIF), mainly with a perinuclear pattern (p-ANCA).[1] ANCA seropositivity by ELISA is less frequent and the target antigens are most commonly lactoferrin, cathepsin G, and MPO.[1]

There are conflicting reports on the significance of ANCA positivity in patients with SLE.[1] SLE and lupus nephritis (LN) may promote neutrophil degranulation and facilitate ANCA autoantibody formation.[2] Diffuse proliferative LN that contains little or no sub-endothelial deposits is rare.[3] Underlying vasculitic lesions induced by ANCA antibodies were recently incriminated in the pathogenesis of these forms.[4]

Nasr *et al.* reported 10 cases of LN with ANCA-associated glomerulonephritis,[2] the evolution of these patients after the treatment by corticosteroids and cyclophosphamid was marked by lethal infectious complications in three patients, complete remission in six patients with a relapse in one case and resistance to treatment in one patient.

Nasr *et al.* evoked the probability of an overlap between lupus nephropathy and ANCA extracapillary glomerulonephritis and suggested to search systematically the positivity of ANCA by ELISA test in a lupus patient whenever renal histology shows extensive necrotizing lesions with a non-significant endocapillary proliferation and rare sub-endothelial deposits.[2]

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