Collapsing focal segmental glomerulosclerosis in a patient with systemic lupus erythematosus

Sir,

A 17 year old girl presented with progressive edema, puffiness of the face, frothy urine, dysuria, and intermittent headache of one week duration. She had no history of photosensitivity, joint pain, skin rash, hair loss, neurological deficits, or constitutional symptoms. On examination, she had anasarca, hypertension (blood pressure - 160/90), pallor, raised jugular venous pressure, oral ulcers, ascites, swelling of both knee joints, tenderness over both wrists, elbow, knee joint, subtalar joint, and metacarpophalangeal joint. She was evaluated for a cause of symmetrical polyarthritis with possible renal involvement.

Investigations revealed hemoglobin 7.1 g/dl, total leukocyte count 5.1 × 10^3/mm^3, platelets 1.53 × 10^3/µl, serum albumin 2.4 g/dl, urine albumin 3+, serum creatinine 2.5 mg/dl, erythrocyte sedimentation rate 90 mm in 1st h, urine protein 5445 mg/24 h, serum anti ds DNA 800 IU/ml, blood C3 and C4 were 0.4 g/dl (low) and 0.05 g/dl (low) respectively.

She underwent a renal biopsy which demonstrated diffuse mesangial and endocapillary hypercellularity with neutrophill infiltration and wire loop thickening. Seven of seventeen viable glomeruli had moderate to marked podocyte hyperplasia. Two glomeruli showed collapsed glomerular tuft with hyperplastic podocytes and periodic acid–Schiff positive droplets. There was no fibrin deposition on special stain. Immunofluorescence showed a "full house" pattern with 3+ deposits in mesangium and capillary loops for IgG, IgM, C3, C1q, kappa, lambda, and fibrinogen. The biopsy was suggestive of diffuse proliferative lupus nephritis (LN) - Class IV A with collapsing glomerulopathy (CG) [Figure 1].

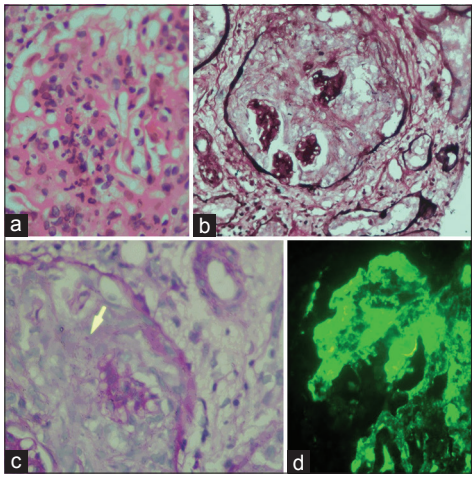

Figure 1: (a) Proliferative glomerulonephritis (H and E, ×400), (b) Collapsed glomerular tuft surrounded by podocyte proliferation (Periodic acid methenamine silver stain, ×400), (c) Periodic acid–Schiff positive granules in the pseudo-crescent (periodic acid–Schiff, ×400), and (d) Strong positivity of C1q along the capillary loops and mesangium (IF for C1q, ×400)

She was given intravenous pulse methylprednisolone followed by oral prednisolone and was initiated on cyclophosphamide monthly cycles. Her proteinuria reduced to 1.5 g/day and creatinine dropped to 1.4 mg/dl after six cycles of cyclophosphamide. She had an episode of cryptosporidium diarrhea recently, resulting in the rise of S.creatinine to 3.9 which settled to 1.5 mg/dl at discharge. Currently, she is on mycophenolate mofetil and oral steroids.

CG is a podocytopathy with segmental or global collapse of the glomerular capillary tuft and podocyte proliferation in Bowman's space,[1] resembling crescentic glomerulonephritis. CG is essentially a disease of podocyte proliferation and can be differentiated from true crescents by their cuboidal morphology and presence of protein reabsorption droplets.[2] First case of CG in LN was reported in 1979.[3] Patients with CG and LN usually present with severe renal failure, massive proteinuria and usually have a poor response to steroids and immunosuppressive therapy.[4] These patients carry a poor prognosis and majority of them end up on dialysis.[5] At present, there is no evidence-based treatment approach for CG with LN owing to the rarity of this disease. Current treatments are based on regimens used to treat focal segmental glomerulosclerosis in HIV patients. Complete remission is <10% and a partial remission rate is reported to be around 15%.[4] Our case demonstrates possible milder form of the same disease which responded to pulse methyl prednisolone and monthly cyclophosphamide therapy.

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Conflicts of interest

There are no conflicts of interest.

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