Systemic Lupus Erythematosus with Stage IV Nephritis in an African Girl

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A 16 year old HIV-negative African girl presented to Princess Marina Hospital in Gaborone, Botswana with a 2 week history of generalized body swelling. It had started as facial puffiness with associated butterfly-like hyperpigmented macular rash around the eyes and cheeks, arthralgia and generalized weakness. One day before presenting she experienced an episode of epistaxis. Examination revealed oral ulcers, scleral reddening, generalized edema, bilateral pleural effusion, and moderate ascites. Blood pressure was 135/85 mmHg, pulse rate 131/min, respiratory rate 21/min and temperature 35.8ºC. Laboratory investigations showed leukocytosis (white blood cells 23,080/µl, neutrophils 65%, lymphocytes 26.9%), anemia (Hb 6.0 g/dl), marked thrombocytopenia (6,000/ µl), raised serum creatinine (186 µmol/L) and urea (17.5 mmol/L), hypoalbuminemia (20.1 g/L), hematuria and proteinuria. Antinuclear (1:320, homogeneous pattern), anti-Smith, anti-ribonucleoprotein antibodies were all positive; anti-dsDNA antibodies and antiphospholipid antibodies were negative; immunoglobulin (Ig) G (5.6 g/L) and IgM (0.4 g/L) levels were low.

A diagnosis of systemic lupus erythematosus (SLE) was reached and treatment was started with blood transfusions, intravenous methylprednisolone 6 mg/kg body weight daily for 3 days followed by 4 mg/kg body weight every other day, antibiotics, antihypertensives, and furosemide. After 10 days the patient had an episode of tonic clonic seizures. A brain computed tomography (CT) scan was normal and phenobarbital was added to the treatment regimen. When the platelet count allowed (24 days after admission), a kidney biopsy was done. Histology showed mild to moderate glomerular and periglomerular chronic inflammatory cell infiltrate comprising mostly lymphoplasmacytic cells, expansion of the glomerular mesangial matrix with obliteration of the Bowman's space in over 50% of the glomeruli, and focal early mesangial matrix sclerosis [Figure 1]. Periodic acid-Schiff stain showed thickened and corrugated glomerular capillary walls with a prominent "wire looping" pattern [Figure 2]. Intracapillary deposits of hematoxyphil bodies and mesangial matrix granularity were also observed. Diffuse proliferative glomerulopathy consistent with stage IV lupus nephritis [1] was diagnosed, and i.v. cyclophosphamide

Figure 1. Histology of renal biopsy showing expansion of the mesangial matrix and focal mesangial sclerosis in a few glomeruli (x 200)

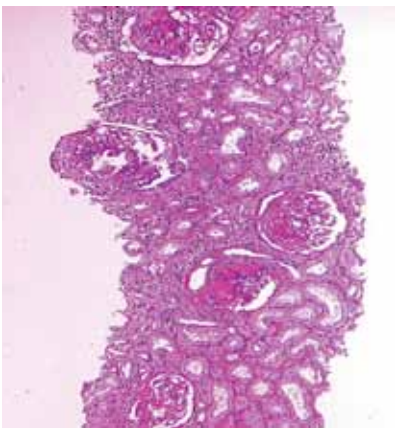
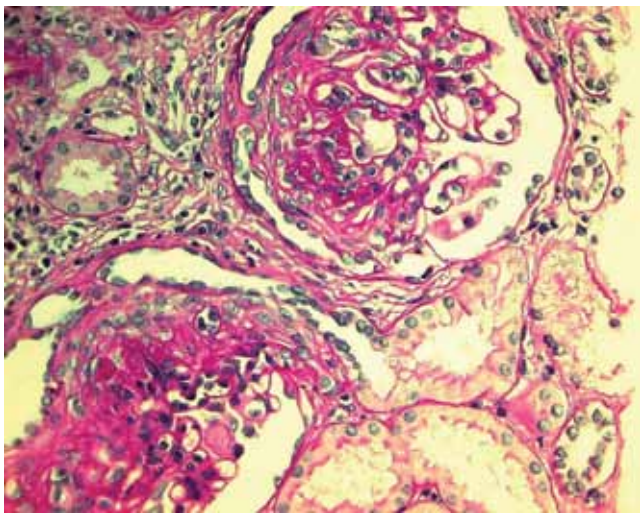


Figure 2. Periodic acid-Schiff (PAS) stain demonstrating thickened glomerular capillary walls with prominent "wire-loop" pattern (x 400)



10 mg/kg body weight every other week was added to the therapy.

Two weeks after discharge the patient's edema resolved; her hemoglobin is 9.8 g/dl, platelets 50,000/μl, white blood cells 11,980/μl, serum urea 10.5 mmol/L, serum creatinine 70 μmol/L, and albumin 18 g/L. She continues treatment with prednisolone (60 mg daily) and cyclophosphamide.

Our patient was anti-Sm and anti-RNP positive, a finding highly prevalent in black Africans [2,3]. However, contrary to the suggestion that patients with Sm/RNP antibodies may have a "benign" form of SLE in which renal and hematologic manifestations are less common and the major manifestations are dermatologic [3], our patient had marked thrombocytopenia and advanced renal disease even though antiphospholipid antibodies (associated with early damage in patients with SLE) [4] were absent. SLE is more aggressive and carries a higher mortality in blacks, in whom nephritis is more common and adversely influences outcome. In a study conducted in Johannesburg, South Africa, 43.8% of patients had nephritis, and the 5 year survival rate in patients with nephritis was significantly lower than in patients without nephritis, 60% vs. 84% respectively [5]. Indeed, nephritis was the only independent predictor of poor outcome and renal failure was the second commonest cause of death [5]. Our patient presented late in the course of nephritis which may adversely influence her outcome.

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