Renal cortical necrosis: A life-threatening manifestation of primary antiphospholipid antibody syndrome

Sir,

The kidney is a major target organ in antiphospholipid antibody syndrome (APLAS).[1] A 28-year-old female patient with a history of three first trimester abortions presented with decreased urine output for 2 weeks following an episode of fever. She was hypertensive for the past 4 years and was on irregular treatment. On admission, her serum creatinine was 7.4 mg/dl, urine examination showed 3+ albuminuria and active urinary sediments, and ultrasonography revealed normal sized kidneys. Antinuclear antibody and anti-dsDNA were negative, C3, C4 levels were normal. Lupus anticoagulant and anticardiolipin anticoagulants were positive. The patient was dialyzed, and renal biopsy was performed which

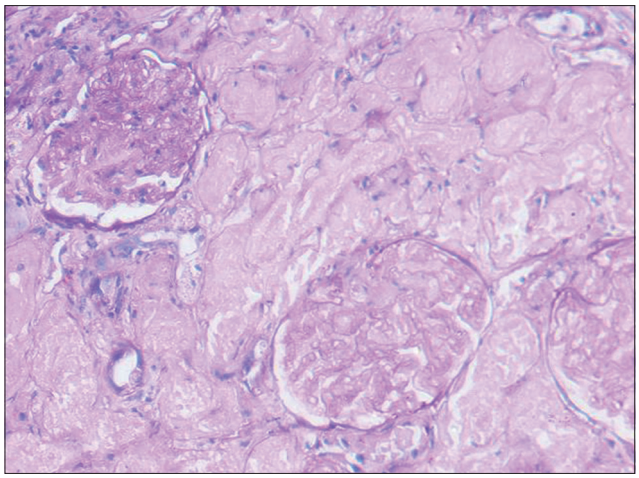

Figure 1: Figure showing patchy cortical necrosis with ghost glomeruli and tubules

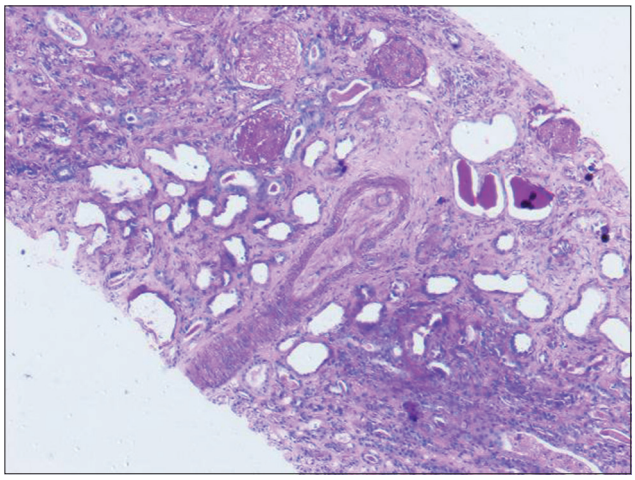

Figure 2: Mucoid intimal hyperplasia of medium sized artery

revealed the evidence of patchy cortical necrosis in the cortex showing ghost glomeruli and tubules [Figure 1]. Small and medium-sized blood vessels showed mucoid intimal hyperplasia with arteriosclerosis causing marked luminal narrowing [Figure 2]. Immunofluorescence was negative for IgG, IgM, IgA, and C3 with overall features suggestive of patchy cortical necrosis with features of malignant hypertension. In view of the possibility of primary APLAS, she was stared on aspirin, prednisolone, and antihypertensives. Her urine output gradually improved to about 1.5–2 L/day. The serum creatinine, however persistent at around 4–5 mg/dl. Her lupus anticoagulant and anticardiolipin antibody levels remained high even after 12 weeks confirming the diagnosis of primary APLAS.

Cortical necrosis continues to be encountered in our part of the world.[2] Renal cortical necrosis accounts for 2% cases of acute kidney injury (AKI) in Western world while it accounts for 7–9% cases of AKI in developing countries.[3] Obstetric causes are responsible for more than 50% cases of acute cortical necrosis while other causes include snake bite, hemolytic uremic syndrome, acute pancreatitis, acute gastroenteritis, and septicemia.[2] APLAS is a constellation of diverse clinical manifestations related principally to hypercoagulable state. Renal manifestations pf primary APLAS include renal artery lesions, glomerular capillary thrombosis, thrombotic microangiopathy, renal cortical necrosis, renal vein thrombosis, and APSN.[1] Cortical necrosis seems to be due to occlusion of small parenchymal blood vessels. These are generally asymptomatic, however, if they are multiple or generalized they may lead to patchy or diffuse cortical necrosis.[4] Another presentation of renal cortical ischemia is slowly progressive ischemia causing renal failure in long-term. Clinically, the patient will have hypertension, proteinuria, and progressive renal failure.[5] Regarding treatment, class of therapeutic drugs, intensity, and duration of treatment remain controversial. Prednisolone, aspirin, heparin, low-molecular-weight heparin, and plasmapheresis have been used in the treatment with variable results.

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Conflicts of interest

There are no conflicts of interest.

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