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On the Cover

What's the diagnosis? A 31-year-old woman with history of systemic lupus erythematosis (SLE), ITP, membranous nephropathy, stage 3 CKD, cerebrovascular accident, and deep venous thrombosis presented with acute kidney injury (serum creatinine increased from 1.8 mg/dl to 4.7 mg/dl over 3 weeks). Urinalysis revealed 3+ protein and large blood while urine sediment demonstrated 7-10 dysmorphic red blood cells/HPF and 3-5 white blood cells/HPF. There were no casts present. ANA was 1:160 while complement levels were normal and anti-double-stranded DNA was negative. A kidney biopsy was performed. The left panel of the cover image shows near obliteration of the intrarenal vessel with fragmented red blood cells in the wall with initial thickening and thrombi occluding the lumen. The right panel shows luminal thrombus with intimal hyperplasia/fibrosis. Antibodies to β2-glycoprotein-1 were elevated. The patient was diagnosed with antiphospholipid antibody syndrome (APLS) secondary to SLE. The patient received corticosteroids and underwent plasmapheresis. Rituximab was subsequently initiated. Kidney function improved with serum creatinine plateauing at 2.1 mg/dl. APLS is an autoimmune disorder characterized by recurrent arterial or venous thrombosis and/or pregnancy loss, in the presence of persistently elevated levels of anticardiolipin antibodies (and/or anti-β2-glycoprotein-1 antibodies) and/or evidence of circulating lupus anticoagulant. Kidney involvement is quite common in both primary and secondary APLS. Thrombosis may occur at any location within the renal vascu- lature, and the size, type and site of the vessel involved drives the renal manifestation in APLS. Included are renal artery stenosis and/or renovascular hypertension, renal infarction, APLS nephropathy, renal vein thrombosis, and increased allograft vascular thrombosis in transplant patients. In addition, renal manifestations of APLS may co-exist with other pathologies, such as proliferative SLE nephritis. The patient described suffered from APLS nephropathy, which is characterized by kidney injury due to vascular lesions in the arterioles and glomeruli. Thrombotic microangiopathy, as seen in this patient, is the most characteristic lesion of APLS nephropathy and is manifested by fibrin thrombi containing fragmented blood cells along with endothelial cell edema, both which narrow or occlude the vascular lumen. Treatment includes anticoagulation, while corticosteroids and immunosuppressive therapy may sometimes be added. This is particularly true in the setting of catastrophic APLS. Plasmapheresis/plasma exchange is frequently performed in this setting. Rituximab have also been utilized in some cases. (*Images and text provided by Mark A. Perazella, MD, and Gilbert Moeckel, MD, Yale University School of Medicine, New Haven, Connecticut*)