

CJASN

Clinical Journal of the American Society of Nephrology

June 2018 • Vol. 13 • No. 6

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On the Cover

What's the diagnosis?

A 25 year-old woman with SLE and antiphospholipid antibodies presented with altered mental status and AKI with serum creatinine (sCr) 1.8 mg/dL (baseline 0.5 mg/dL). Her BP was 119/79 mmHg. Urinalysis revealed 1847 RBC/hpf with RBC casts and 2.9 g of urine protein in 24 hours. C3 and C4 levels were both decreased. ANA, dsDNA, Smith, and RNP antibodies were positive. CT head demonstrated acute bilateral cerebellar infarcts. She was treated with iv heparin which was stopped 8 hours before kidney biopsy and restarted 24 hours afterward. Biopsy revealed RPS/ISN Class IV lupus nephritis. She was treated with methylprednisolone iv 1 g daily x3 days followed by oral prednisone 60 mg daily, mycophenolate mofetil, lisinopril, and hydroxychloroquine. Her mental status significantly improved, and the sCr returned to 0.5 mg/dL within 8 days. By 1 month, sCr was 0.7 mg/dL with uPCR 0.77 g/g.

Light microscopy showed mesangial matrix expansion, mesangial proliferation without endocapillary proliferation or crescents, and large immune deposits within the capillary walls ("wire loops") (Figures 1 and 2). On immunofluorescence, diffuse, intense mesangial, capillary wall, and peritubular staining was present for C1q, IgG, IgA, IgM, C3, kappa, and lambda. Electron microscopy demonstrated subendothelial, intramembranous, subepithelial, mesangial, and peritubular capillary deposits with tubuloreticular inclusions (Figure 3).

A diagnosis of Class IV lupus nephritis can be made on the basis of different criteria, including the presence of extensive subendothelial immune deposits without endocapillary proliferation. Induction therapy for Class IV lupus includes a combination of glucocorticoids plus either IV cyclophosphamide or oral mycophenolate mofetil.

(Text and images provided by Sadichhya Lohani, MD, Mohamad Hanouneh, MD, and C. John Sperati, MD, MHS, Johns Hopkins University, Department of Medicine, Division of Nephrology, Baltimore, Maryland)