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What's the diagnosis?

Case description: The patient is a 64-year-old woman with a history of recent coronary artery bypass graft who presented with a one-week history of a purpuric rash on her ankles which spread to thighs, trunk, and arms. Creatinine increased from baseline of 0.9 to 3.1 mg/dL, and urine analysis revealed protein and blood. The clinical suspicion was for vasculitis; serologic studies including anti-neutrophil cytoplasmic antibody (ANCA) were negative, and complement levels were normal. A kidney biopsy was performed, and the rash responded well to prednisone.

Images:

- Left panel: Granular mesangial and peripheral capillary wall deposition of IgA by immunofluorescence microscopy.
- Middle panel: Medullary angiitis, with prominent leukocytoclastic neutrophilic reaction in the medulla, and medullary hemorrhage.
- Right panel: Vascular necrosis in the medulla, with dissolution of the arterial media (arrows) and replacement by red blood cells and fibrin. Jones silver stain highlights the outline of residual vascular myocytes.

Teaching points: This is an unusual case of IgA nephropathy/Henoch-Schönlein purpura, with predominantly extraglomerular vasculitic-like lesions and associated medullary angiitis and hemorrhage. Glomerular changes were modest and limited to minimal mesangial expansion and focal endocapillary hypercellularity.

- Medullary angiitis is rare (0.19% of cases in the largest series from one institution, Hum Pathol. 2013 Apr;44(4):521-5).
- Most cases (63%) are associated with ANCA vasculitis.
- Approximately 20% of medullary angiitis may be attributed to IgA nephropathy, as in this case.

Rare cases of IgA nephropathy with features not well-represented in the Oxford classification validation may have different prognostic implications.

(Images and text were provided by Nicole K. Andeen, MD, Oregon Health & Science University, Portland, Oregon and Kumar Dinesh, Renal Care Consultants, Medford, Oregon)