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On the Cover

What is the diagnosis? A 73-year-old man was seen for rising serum creatinine (1.1 mg/dl → 3.8 mg/dl) associated with malaise and fatigue. One-week prior, the patient was exposed to antibiotics for a urinary tract infection. Vital signs were stable and physical examination revealed clear lungs, normal heart sounds, benign abdomen, and 1+ lower extremity edema without skin rash. Urinalysis showed 1+ blood and trace leukocyte esterase. Urine sediment examination revealed isomorphic red blood cells, white blood cells, renal tubular epithelial cells, a few finely granular casts, and a white blood cell cast. The patient underwent kidney biopsy to evaluate acute kidney injury with active urine sediment. Hepatitis B surface antigen and hepatitis C antibody were negative, while hepatitis B surface antibody was positive. MPO-ANCA was positive at low titer. The patient received oral prednisone 60 mg/day with improvement of kidney function (serum Cr at 1.8 mg/dl).

The kidney biopsy revealed an interstitial infiltrate consisting of lymphocytes, plasma cells, and eosinophils as well as a medium vessel vasculitis consistent with polyarteritis nodosa. As seen on the cover image, a medium sized vessel stained with PAS, silver and trichrome demonstrates a leukocytic infiltrate within the vessel wall. Narrowing of the vessel lumen is also present.

Polyarteritis nodosa (PAN) is a rare disorder that consists of a necrotizing vasculitis affecting primarily medium-sized arteries. It can affect all age groups, with a peak incidence in the fourth and fifth decades. PAN is often associated with positive hepatitis B surface antigen, but this finding is not universal. PAN is most often a multi-system disease, but can be limited to vasculitis involving one organ system. Patients generally present with constitutional symptoms and a vague systemic illness depending on the organs involved. Muscle, nerve, gastrointestinal tract, skin, joint, kidney, and lung are some of the organs involved. ANCA serology is often negative unless there is a concomitant small vessel vasculitis. Angiography reveals multiple aneurysms (beading) of affected vessels most commonly involving the celiac axis and renal vessels. Biopsy of muscle or sural nerve may demonstrate the necrotizing vasculitis. It is generally recommended to avoid kidney biopsy due to a risk of aneurysmal rupture and bleeding; however, renal biopsy has been performed safely. PAN histology consists of focal necrotizing arteritis consisting of a mixed cellular infiltrate within the vessel wall. (Images and text provided by Eric Chang, MD, Randy Luciano, MD, PhD, Gilbert Moeckel, MD and Mark A. Perazella, MD, Yale University School of Medicine, New Haven, Connecticut)