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On the Cover

What’s the diagnosis?

Case description: The patient is a 57 year old man with a history of hypertension and chronic kidney disease with a serum creatinine of 2.68 mg/dL 4 years ago, which has gradually risen to 9.7 mg/dL. At initial workup, duplex ultrasound revealed an atrophic left kidney with no blood flow in the left renal artery, and possible stenosis of the artery supplying the upper pole of the right kidney. Four years later, during a hospitalization for a gastrointestinal bleed, abdominal imaging revealed an abdominal aortic aneurysm and masses in the right kidney. A nephrectomy was performed.

Images:
- Left panel: Renal mass, radiographically concerning for renal cell carcinoma.
- Middle panel: Granulomatous vasculitis with numerous giant cells, disruption of the elastic lamina and focal necrosis, involving the right upper pole renal artery. Proximal to this, the artery showed marked damage to the muscular and elastic portions of the wall, stenosis, and features of recanalization.
- Right panel: Histology of the renal “mass” reveals granulomatous and giant cell-rich vascular and interstitial inflammation.

Teaching points: This represents a very unusual case of granulomatous arteritis causing renal artery stenosis and renal mass lesions. The differential diagnosis for granulomatous inflammatory processes involving large and medium-sized arteries includes:
- Infectious etiologies: particularly those related to acid fast, spirochetal or fungal organisms. Corresponding special stains were negative for microorganisms.
- Sarcoid vasculitis: a rare complication of sarcoidosis. Patients with sarcoid vasculitis generally have lung involvement and sometimes central nervous system involvement, which were not present in this patient.
- Giant cell arteritis and Takayasu arteritis: these are difficult to distinguish histologically, and are primarily differentiated clinically.
  - Takayasu arteritis is more common in individuals <40 years old, and giant cell arteritis is more common in individuals >50 years old.
  - Giant cell arteritis usually presents with headaches and can be diagnosed by temporal artery biopsy.
  - Takayasu arteritis typically involves the aorta and its major branches, and has been classified into different types based on the involvement of the aorta. Takayasu arteritis is more common in some ethnicities and in women, who frequently have involvement of the thoracic aorta and its branches; men are more likely to have involvement of the abdominal aorta and its branches.

Given the constellation of available clinical and pathologic findings, we favor the interpretation of Takayasu arteritis, and suggest that this multifocal granulomatous arteritis may be a unifying diagnosis to explain the patient’s bilateral renal disease, abdominal aortic aneurysm and intestinal bleeding. (Images and text provided by Nicole K. Andeen and Kelly D. Smith, University of Washington – Pathology, Seattle, Washington.)