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On the Cover

What's the diagnosis? A 42-year-old male with a history of bilateral deep vein thrombosis due to antiphospholipid antibody syndrome (APLS) on warfarin presented to the hospital with a lower leg ulcer and cellulitis and was treated with antibiotics. He presented a few days later with right-sided flank pain and was found to have hematuria and nephrotic range proteinuria. The initial CT scan was positive for perirenal stranding on the right. While in the hospital, he developed severe abdominal pain, nausea and vomiting. Two days after admission he had a repeat CT scan, which demonstrated periaortic stranding consistent with aortitis. The cover image demonstrates this CT scan with contrast (coronal cut through the abdomen). The two midline structures depicted are the inferior vena cava and the aorta. Of note is the segmental attenuation and thickening of the infrarenal aortic wall (green arrows in image below) and periaortic fat infiltration. On kidney biopsy, he was found to have acute thrombotic angiopathy with active intravascular thrombosis, which confirmed the diagnosis of catastrophic antiphospholipid antibody syndrome. After 5 sessions of plasma exchange as well as high dose prednisone, the radiologic findings completely resolved.

“Aortitis” is a pathologic term that refers to an abnormal inflammation of the aortic wall. Clinical features of aortitis are nonspecific and may include fever, abdominal or chest pain, and vascular insufficiency. A wide spectrum of infectious, inflammatory, and idiopathic conditions may result in the development of aortitis. Although Takayasu arteritis and giant cell arteritis are the most common rheumatologic causes of aortitis, other systemic diseases, such as rheumatoid arthritis, systemic lupus erythematosus, Behçet disease, and Cogan syndrome, may also be associated with aortitis. Aortitis has only rarely been described in association with APLS. (Image and text provided by Dr. Adina Voiculescu, Brigham and Women's Hospital, Boston, MA)