

## AKI, Anemia, and Thrombocytopenia in a Patient with Leg and Flank Pain

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### Case Description

A 56-year-old woman presented to the emergency department for sudden-onset pain in her left leg and right flank. She had a history of deep vein thrombosis in her right leg and had discontinued anticoagulation 3 months previously. Physical examination revealed hypotension, tachycardia, and bilateral ankle edema 1+. Blood tests were relevant for elevated serum creatinine (2.2 mg/dl), BUN (57 mg/dl), and hemoglobin 9.4 gdl, and platelet count was low (54,000 cells/ $\mu$ l). Urine dipstick was negative for hematuria, with a spot albumin-creatinine ratio of 30 mg/g.

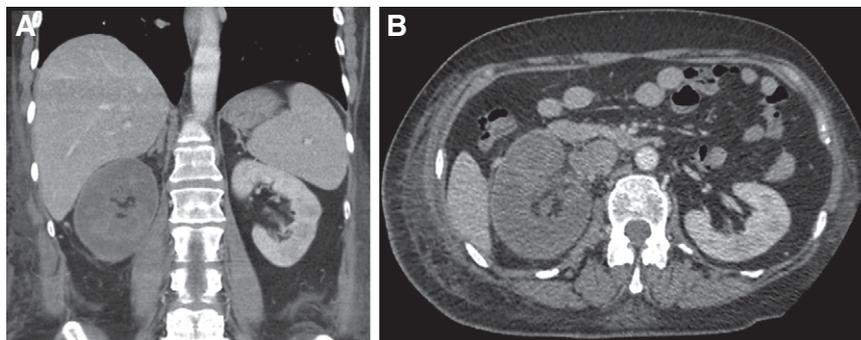
Imaging studies confirmed deep venous thrombosis in the left lower extremity, inferior vena cava, and bilateral pulmonary arteries. Computed tomography (CT) scans showed an enlarged, nonenhancing right kidney (Figure 1A), with an enhancing “cortical rim sign” (Figure 1B), and a filling defect within the renal vein. She was immediately started on anticoagulation with low molecular-weight heparin. Further blood tests confirmed high titers of both anti- $\beta$ 2 glycoprotein (IgG, IgM) and anticardiolipin antibodies (IgG, IgM). There were no features of SLE or any other autoimmune diseases.

The patient was treated with intravenous immune globulin, pulse methylprednisolone, and continued anticoagulation for probable catastrophic antiphospholipid syndrome (CAPS) and discharged uneventfully on

prednisone and vitamin K antagonists. At 3-month follow-up, the patient remains stable, with an eGFR 97 ml/min per 1.73 m<sup>2</sup>, despite the absence of right kidney uptake on scintigraphy due to infarction (Figure 2). Platelet count and hemoglobin were normal, and anticardiolipin antibodies were persistently positive.

We describe a case of CAPS with unilateral large-vessel renal occlusion with permanent kidney damage. Antiphospholipid syndrome affects the kidneys in 25% of patients, mainly *via* venous, arterial, or microvascular thrombosis (1). The classic triad of renal vein thrombosis includes acute flank pain and hematuria, and, although rare, it has been reported to cause renal infarction. A subset of patients present with widespread thrombosis and end-organ damage, with mortality approaching 50% despite aggressive treatment, termed CAPS (2). Histopathology reveals noninflammatory vascular occlusion, and care must be taken to exclude other forms of kidney damage (thrombotic microangiopathy, membranous nephropathy, or proliferative GN).

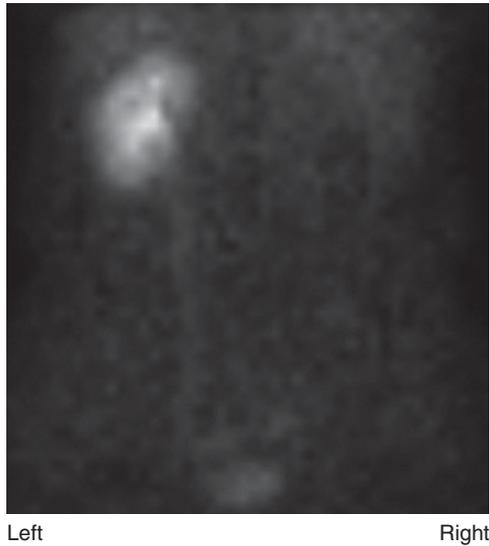
Prompt diagnosis and early treatment are critical to prevent complications. There are no randomized clinical trials to guide treatment in patients with CAPS, and combination therapy includes steroids, intravenous Ig, and anti-CD20, on the basis of case reports and case series (3).



**Figure 1.** | Kidney imaging upon initial presentation. (A) Coronal abdominal computed tomography (CT) shows an enlarged right kidney with nonenhancing parenchyma. (B) Axial abdominal CT reveals a thin cortical enhancing rim.

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**Figure 2.** | Renal scintigraphy (using diethylene triamine penta-acetic acid-mercaptoacetyltriglycine) demonstrates the absence of radioisotope uptake in the right kidney, at 3-month follow-up.

### Teaching Points

- APS affects the kidneys in nearly 25% of patients, damage can occur through major vessel thrombosis and/or thrombotic microangiopathy.
- A high suspicion index and confirmatory testing allow for early treatment aimed at preventing thrombosis and end-organ damage.

### Disclosures

All authors have nothing to disclose.

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The patient's informed consent was obtained before submitting this manuscript.

### Author Contributions

J.I. Lara Prado wrote the original draft; F. Pazos Pérez provided supervision and was responsible for the validation and visualization; and all authors reviewed and edited the manuscript.

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