

Antiphospholipid Syndrome and the Aorta: A Rare Presentation

ANJALI SHROFF, HAN-OH CHUNG, NADER A. KHALIDI and ALEX SPYROPOULOS

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Antiphospholipid Syndrome and the Aorta: A Rare Presentation

To the Editor:

Antiphospholipid syndrome (APS) is a disorder of coagulation that is usually manifested by arterial or venous thrombosis, or pregnancy-related complications such as miscarriage, stillbirth, preterm delivery, or severe preeclampsia¹. The syndrome occurs due to the autoimmune production of antibodies against the cell membrane phospholipid.

A 39-year-old woman presented to the emergency department with severe abdominal pain. An abdominal computed tomography (CT) angiogram showed complete occlusion of the distal abdominal aorta from the origin of the left renal artery, common and external iliac artery occlusion, extensive infarcts of kidneys and spleen, and a superior mesenteric artery thrombus (Figures 1 and 2).

She had a history of intermittent left upper-quadrant abdominal pain with radiation to the back with nausea and diaphoresis. Her history was significant for hypertension, a remote history of peptic ulcer disease, and chronic back pain. Her obstetric history was G5P4A1 with one spontaneous abortion at 10 weeks' gestation. Her medications were hydrochlorothiazide, and oxycodone/paracetamol and ibuprofen as needed for back pain. She was not taking oral contraceptives. She was a tobacco smoker with a 12.5 pack-year history. She drank alcohol occasionally and denied any recreational or intravenous drug use. There was no family history of thrombosis or malignancy.

On examination, she was hypertensive (blood pressure 159/115) and tachycardic (120 bpm) but was otherwise stable and afebrile. Her examination was unremarkable except for some abdominal tenderness on her left side with some costovertebral angle tenderness.

A complete blood count showed hemoglobin 124 g/l (normal 115–165 g/l), leukocyte count $20.2 \times 10^9/l$ (normal $4.0\text{--}11.0 \times 10^9/l$), and platelet count 635 (normal $150\text{--}400 \times 10^9/l$). Electrolytes were normal and creatinine was elevated at 109 $\mu\text{mol/l}$ (previously 60 $\mu\text{mol/l}$). Her urinalysis revealed 3+ proteinuria with 2+ blood.

An extensive hypercoagulable investigation was performed. Protein C

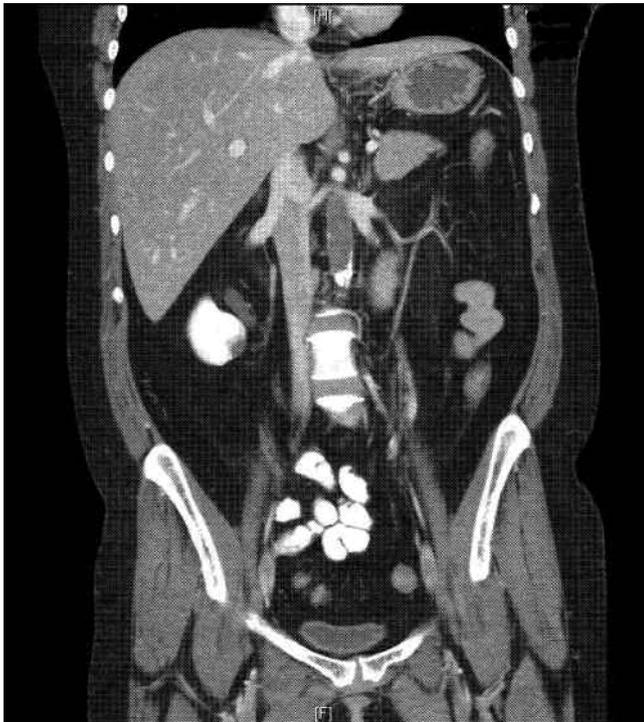


Figure 1. Abdominal computed tomography coronal slice shows large aortic thrombus extending infrarenally.

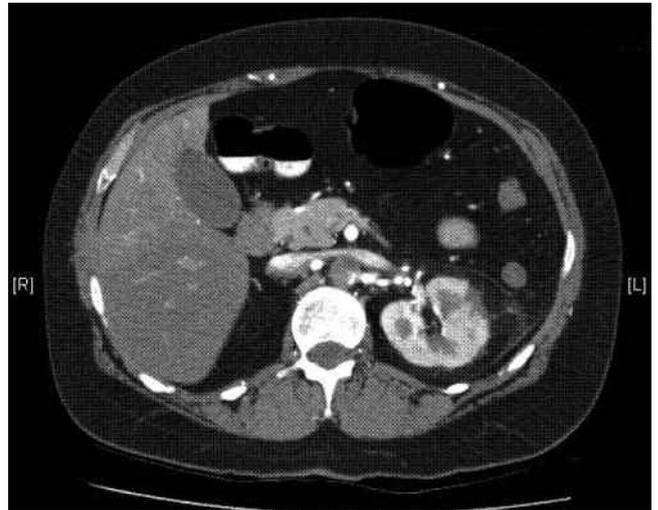


Figure 2. Axial computed tomography reveals large aortic thrombus and left renal infarcts.

and S levels and activated protein C resistance (APCR) ratio were normal. Prothrombin gene mutation screen and Janus Kinase 2 (JAK2) were negative. Homocysteine level was insignificantly elevated at 14.8 (normal 4–12). Paroxysmal nocturnal hemoglobinuria was ruled out with negative cell markers for CD55 and CD59. A 24-hour urine collection was negative for nephrotic syndrome.

Screening for APS revealed a negative VDRL screen, but a positive dilute activated partial thromboplastin time with a positive 1:1 mix, suggesting the presence of a lupus-like inhibitor. As a result, she was also investigated for a possible connective tissue disease (CTD). Her history was unrevealing for any symptoms of CTD. She had an elevated erythrocyte sedimentation rate (46) and C-reactive protein (13.8). Antinuclear antibody, extractable nuclear antigens, rheumatoid factor, anticitrullinated protein antibody (anti-CCP), antineutrophil cytoplasmic antibodies, complement levels, immunoglobulins, and protein electrophoresis were all normal. Screening for hepatitis B and C and HIV revealed a positive hepatitis C serology. However, she was negative for cryoglobulins.

Polyarteritis nodosa and other large-vessel vasculitides were ruled out as she did not fit diagnostic criteria and the CT angiogram showed no evidence of aneurysms or vessel wall thickening consistent with inflammation. Buerger's disease was ruled out as she had no evidence of acral involvement. Blood cultures and transesophageal echocardiography were negative for evidence of endocarditis.

With her positive nonspecific inhibitor, a presumptive diagnosis of APS was made. She was anticoagulated with dalteparin, transitioned to warfarin and was discharged home. Her followup visit 5 months later confirmed the presence of a lupus-like inhibitor.

APS is considered secondary if it occurs with other related autoimmune diseases such as systemic lupus erythematosus. Primary APS is diagnosed when the presence of other related diseases is excluded.

The Sapporo criteria² require one clinical criterion (vascular thrombosis or pregnancy mortality before 10 weeks) and one laboratory criterion (one of lupus anticoagulant, anticardiolipin antibody, or anti- β_2 -glycoprotein antibody are positive twice at least 12 weeks apart) for diagnosis of APS. The 3 laboratory criteria are used because of the variability of APS cases, as not all patients present homogeneously^{3,4}.

Our patient had an extremely rare form of thrombosis in the great vessels. To our knowledge, there are only 4 other case reports in the literature^{3,4,5,6} — none with the feature of complete aortic occlusion observed in this patient.

Thus, our patient fit the clinical criteria with extensive splenic, kidney, superior mesenteric artery, and aortic thrombi, and pregnancy loss at 10

weeks' gestation. She has also had 2 confirmed episodes of a lupus-like inhibitor. She was negative for the anticardiolipin antibody, and the anti- β_2 -glycoprotein test is not done at our institution. This suggests a rare form of primary APS with involvement of the great vessels.

ANJALI SHROFF, MD; HAN-OH CHUNG, MD, Department of Internal Medicine; NADER A. KHALIDI, MD, Department of Rheumatology; ALEX SPYROPOULOS, MD, Thrombosis, McMaster University, Hamilton, Ontario, Canada.

Address correspondence to Dr. Shroff; E-mail: anjali.shroff@gmail.com

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