Abdominal aortic aneurysm associated with antiphospholipid syndrome: Imaging findings

Clinical History:

A 36-year-old female patient had been diagnosed with antiphospholipid syndrome (APS) at the age of 27 with detection of antiphospholipid antibodies in the serum. At that time, antinuclear antibodies were repeatedly negative. She came to our Emergency Department with fever and pain in the lower dorsal region and epigastrium.

Imaging Findings:

Multidetector computed tomography (MDCT) revealed a large polylobulated saccular aneurysm of the abdominal aorta cranial to the renal arteries, with a maximum diameter of 7 cm (Fig. 1). It was surrounded by a thick and irregular wall of inflammatory appearance that showed moderate enhancement after intravenous contrast material (IVC) administration (Fig. 2). Infiltration of neighbouring tissues and vascular structures was observed, with total occlusion of the coeliac trunk and superior mesenteric artery (arrow and arrowhead in figure 3). Both arteries were recanalised distally via collaterals that mainly arose from the inferior mesenteric artery (arrow in Fig. 4). The right renal artery was also infiltrated, showing almost total obstruction, with a major reduction in right kidney size and poor enhancement of the right versus left renal parenchyma. There was no sign of atherosclerotic involvement of the aorta or of other abdominal arteries.

Discussion:

APS is a clinical condition characterised by recurrent fetal loss, venous and/or arterial thrombosis, and thrombocytopenia associated with elevated antiphospholipid antibody titers [1]. APS was originally reported in patients with systemic lupus erythematosus (SLE) but has more recently been considered an independent syndrome (primary APS) [2]. There are few examples in the literature of an association between APS and onset of abdominal aortic aneurysm [3], and none showed imaging studies of the aneurysm with the marked inflammatory characteristics observed in the present case, which simulated an infectious or mycotic aneurysm [4]. However, it was not clearly demonstrated to be secondary to vasculitis. The coexistence of arterial aneurysms and APS creates a therapeutic dilemma, because the latter must be treated by anticoagulation, which may favour aneurysm bleeding. The present patient was initially diagnosed with primary APS, developing SLE with renal involvement eight years
later. Abdominal aortic aneurysm was clinically manifest at around six months after the SLE diagnosis. Hence, we consider the aneurysm to have been associated with the APS and not with the SLE. Aneurysms related to a primary APS have been reported in splenic, hepatic, and renal arteries [5], pancreatic, renal, and superior mesenteric arteries [6], pulmonary arteries [7], coeliac trunk and superior mesenteric, splenic, renal, and iliac arteries [8], and hepatic artery [9]. In 2008, Szyper-Kravitz et al. [3] described four patients with abdominal aortic aneurysm associated with primary APS, although images were only shown of two of the cases. Our radiological study revealed a large aneurysm with saccular polylobulated morphology and no evidence of mural thrombus or calcification. Only one of the aortic abdominal aneurysms reported by Szyper-Kravitz et al. was saccular, while the remaining cases were fusiform. In contrast, the visceral aneurysms reported in patients with APS were all saccular [5-9]. A unique feature of the aneurysm in the present patient was its “inflammatory” appearance, with replacement of the aortic wall by a thick and irregular wall and the infiltration and destruction of the walls of neighbouring arterial vessels. This behaviour has not been reported in any of the cases consulted. It is highly unlikely that atherosclerosis was responsible for the aneurysm in the present case, given that no signs of this disease were detected in the abdominal aorta not affected by the aneurysm or in the abdominal arteries. The pathology report revealed absence of vasculitis. Mycotic aneurysm was ruled out by running the appropriate tests.

Written informed patient consent for publication has been obtained.

**Differential Diagnosis List:** Abdominal aortic aneurysm associated with APS, Mycotic aneurysm, Infectious process, Arteriosclerotic aneurysm, Connective tissue disease, Immunoglobulin G4 (IgG4)-related disease

**Final Diagnosis:** Abdominal aortic aneurysm associated with APS

**References:**


Description: 36-year-old female patient with abdominal aortic aneurysm associated with antiphospholipid syndrome. CT scan with intravenous contrast in arterial phase. Maximum intensity projection view. Polylobulated saccular aneurysm of abdominal aorta (arrow), with multiple abdominal arterial collaterals (arrowheads). No calcification of the aneurysm wall can be seen. Origin: Department of Radiology, University Hospital “Virgen de las Nieves”, Granada, Spain, 2012
Description: 36-year-old female patient with abdominal aortic aneurysm associated with antiphospholipid syndrome. CT without intravenous contrast (A) and with intravenous contrast in arterial, portal, and excretory phases (B, C, D). Axial images at the height of the spleno-portal axis visualised with the same window. Origin: Department of Radiology, University Hospital “Virgen de las Nieves”, Granada, Spain, 2012
Description: 36-year-old female patient with abdominal aortic aneurysm associated with antiphospholipid syndrome. CT without intravenous contrast (A) and with intravenous contrast in arterial, portal, and excretory phases (B, C, D). Axial images at the height of the spleno-portal axis visualised with the same window. Origin: Department of Radiology, University Hospital “Virgen de las Nieves”, Granada, Spain, 2012
Description: 36-year-old female patient with abdominal aortic aneurysm associated with antiphospholipid syndrome. CT without intravenous contrast (A) and with intravenous contrast in arterial, portal, and excretory phases (B, C, D). Axial images at the height of the spleno-portal axis visualised with the same window. Origin: Department of Radiology, University Hospital "Virgen de las Nieves", Granada, Spain, 2012
Description: 36-year-old female patient with abdominal aortic aneurysm associated with antiphospholipid syndrome. CT without intravenous contrast (A) and with intravenous contrast in arterial, portal, and excretory phases (B, C, D). Axial images at the height of the spleno-portal axis visualised with the same window. Origin: Department of Radiology, University Hospital “Virgen de las Nieves”, Granada, Spain, 2012
Description: 36-year-old female patient with abdominal aortic aneurysm associated with antiphospholipid syndrome. CT with intravenous contrast in arterial phase: volume rendering view. Total occlusion of the coeliac trunk (arrow) and superior mesenteric artery (arrowhead) can be observed, with the distal recanalisation of both arteries. Origin: Department of Radiology, University Hospital “Virgen de las Nieves”, Granada, Spain, 2012.
Description: 36-year-old female patient with abdominal aortic aneurysm associated with antiphospholipid syndrome. CT with intravenous contrast in arterial phase. Volume rendering view showing strong collateral circulation that mainly arises from the inferior mesenteric artery (arrow), secondary to total occlusion of the coeliac trunk and superior mesenteric artery. Origin: Department of Radiology, University Hospital “Virgen de las Nieves”, Granada, Spain, 2012