Symptomatic thrombus of Infrarenal Abdominal Aorta in a patient with Antiphospholipid Syndrome. A case report and review of the literature

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Abstract:

A previously healthy 58-year-old man presented to the emergency department with nine days history of right lower extremity rest pain (class Fontaine III, Rutherford IV). The ankle-brachial index in the right and left leg was 0.67 and 0.75, respectively. Computed tomography angiography (CTA) revealed the presence of thrombus in the infrarenal aorta, starting one centimeter below the renal arteries with occlusion of the right common iliac artery. A rheumatological workup showed antiphospholipid antibodies, and the anticardiolipin immunoglobulin G was positive. The patient underwent covered endovascular reconstruction of aortic bifurcation (CERAB). Twelve months after the procedure, he demonstrated no signs of limb ischemia, and the CTA showed excellent stent patency as well as sufficient blood flow in the infrarenal aorta and both lower limbs.

INTRODUCTION

Antiphospholipid syndrome (APS) represents an autoimmune disorder characterized by recurrent thromboses in arterial and/or venous circulation.[1-2] Although arterial thrombosis preferentially occurs in small and medium size vessels, aortic thrombosis has been also reported.[3] Unfortunately, little is known about the optimal treatment strategy for this rare condition. We report a case of symptomatic aortic thrombus due to APS, which was successfully treated by the covered endovascular reconstruction of aortic bifurcation (CERAB) technique.

CASE PRESENTATION

A previously healthy 58-year-old man presented to the emergency department with nine days history of right lower extremity rest pain (class Fontaine III, Rutherford IV). His medical history was unremarkable, and he also denied any tobacco use, cardiac arrhythmias, coronary artery disease or prior embolic events. The ankle-brachial index (ABI) in the right and left leg was 0.67 and 0.75, respectively.

Computed tomography angiography (CTA) revealed a heavy burden of thrombus in the infrarenal aorta starting one centimeter below the renal arteries with occlusion of the right common iliac artery (Figure 1, red arrow). Due to the unremarkable medical history, a rheumatological workup was performed. Antiphospholipid antibodies detected by enzyme-linked immunosorbent assay were 640 U/mL (positive, n.v. 0-0.9 GPL U/ml), and the anticardiolipin immunoglobulin G detected by lupus anticoagulant assays was >160 mg/mL (positive, n.v: 0-8 GPL U/ml). Additionally, an extensive hypercoagulopathy investigation (Protein C, S, Antithrombin, Factor V Leiden, Homocysteine, fibrinogen etc) was conducted. The levels of fibrinogen were slightly elevated (>400mg/dl). The patient had history of deep vein thrombosis, which had been diagnosed by Doppler ultrasound, and ischemic heart disease. He was diagnosed with symptomatic thrombus in the infrarenal aorta and occlusion in the right common iliac artery, co-occurring with antiphospholipid syndrome. The patient immediately received anticoagulant (rivaroxaban 2.5mg, twice daily) and antiplatelet (aspirin 100mg o.d) regimen. However, these conservative treatments were insufficient to resolve the patient's symptoms. A multidisciplinary team of internists, haematologists, rheumatologists, anesthesiologists and vascular surgeons recommended a surgical approach (thrombectomy or bypass grafting) or an endovascular approach (CERAB technique) to manage his condition. The patient, however, was unwilling to undergo surgical treatment, so he was scheduled for endovascular treatment (EVT) of the thrombotic lesion of the infrarenal aorta and the right common iliac artery.

The patient was placed in a supine position. The procedure was performed percutaneously using a bilateral transfemoral approach. The patient received 100 units/kg of heparin intraoperatively. A short 6Fr sheath was placed in both femoral arteries. In the right leg, the lesion was crossed intra-luminally using a 0.035 glidewire. A 12 Fr sheath was inserted via the left femoral artery, while in the right femoral artery, the 6 Fr

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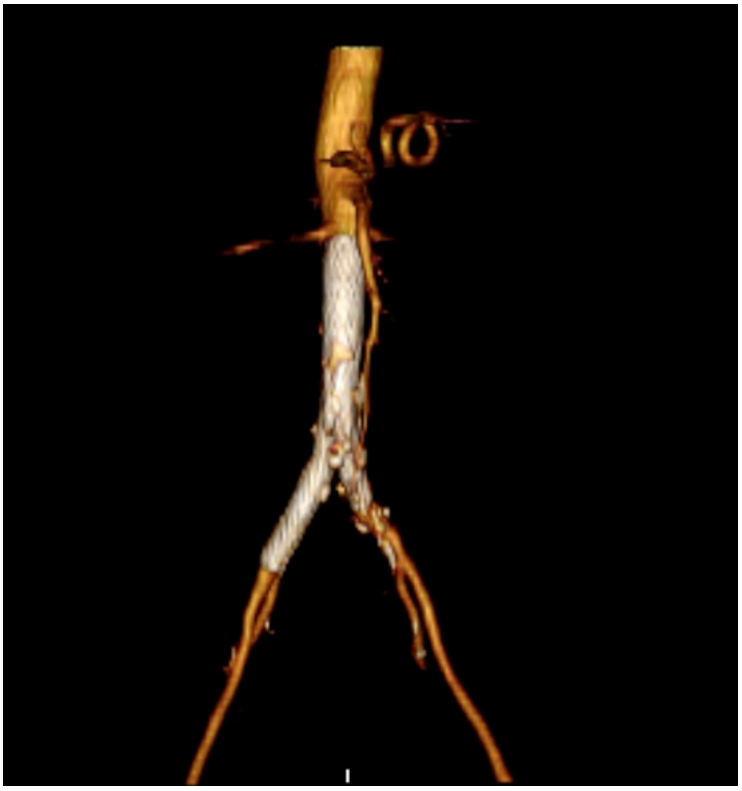

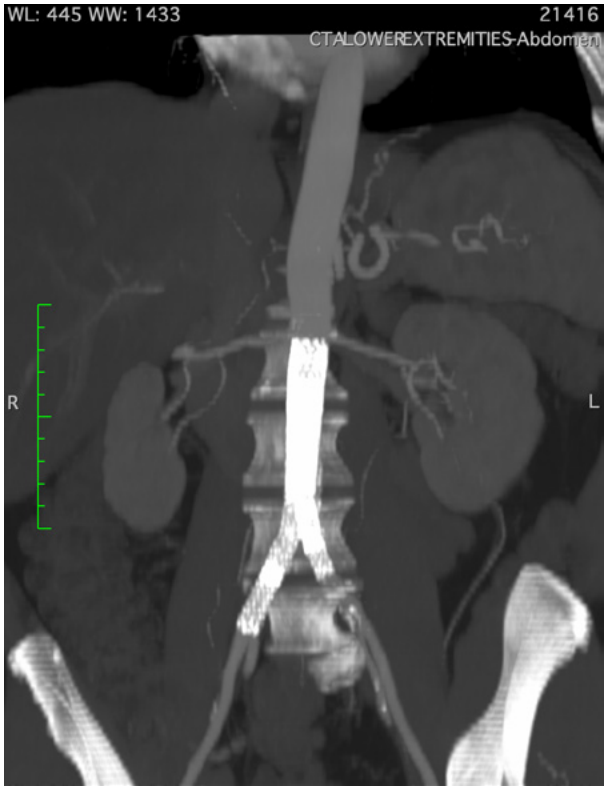

sheath was replaced with a 7 Fr sheath. Angiography through the 12 Fr sheath detected an irregular surface of the infrarenal aorta and occlusion of the right common iliac artery. A 20 mm in diameter and 48 mm in length covered balloon-expandable stent (BeGraft, Bentley, Hechingen, Germany) was deployed below the renal arteries ending approximately 20 mm above the bifurcation. The aortic stent was then molded with short semi-compliant balloons to ensure optimal aortic wall apposition.

Thereafter the iliac stents (10 x 57 mm, BeGraft, Bentley, Hechingen, Germany) were positioned in a kissing conformation, overlapping with the aortic stent for 15mm. Then ballooning with two compliant balloons was performed to adapt the parallel stents to the aortic one. Completion angiography was performed at the end of the procedure to verify the correct deployment of the stents, the apposition of the kissing stents to the aortic one, the patency of renal arteries and CERAB reconstruction.

After the removal of catheters and sheaths, a closure device (Perclose ProGlide Abbott Scientific, Abbott Park, IL) was used to close the arteriotomies in both common femoral arteries.

Postoperatively, the patient was treated with rivaroxaban (2.5mg, twice daily) and acetylsalicylic acid. Twelve months after the procedure, he demonstrated no signs of limb ischemia, with a normal ABI, whilst the CTA showed excellent stent patency as well as sufficient blood flow in the infrarenal aorta and the bilateral lower limbs. (Figure 2).

DISCUSSION

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 preeclampsia. The syndrome occurs due to the autoimmune production of antibodies against the cell membrane phospholipid.[1-2] When the aorta is affected, this scenario often leads to thrombosis. The management of this rare complication varies in the literature, from conservative treatment [4-5] to open [6-7] or endovascular approaches.[8] We experienced an extremely rare case of aortic thrombosis due to primary APS, which was successfully treated by the CERAB technique. According to the Sapporo criteria,[8] in patients with triple factors positivity and first unprovoked venous thrombosis, vitamin K antagonists are recommended. In our case, we preferred lifelong rivaroxaban because the patient had two positive factors. To the best of our knowledge, 11 cases have been reported in the literature (table 1), and only one was treated by endovascular means.[9] Four cases have been conservatively treated, and their late outcomes are unknown. However, these conservative treatments were insufficient to resolve the patient's symptoms, according to scarce date in the literature.[7]

The CERAB technique was introduced in 2013 for treating patients suffering from aortoiliac occlusive disease. The main goal was to reduce some negative impacts of the kissing stents technique, such as the discrepancy between the stented lumen and the aortic lumen ("radial mismatch"), that may affect their patency rate.[9] Additionally, this procedure moves

Table 1. Case reports of aortic thrombosis due to antiphospholipid syndrome

Author	Year	Journal	Age/Sex	Symptoms	Location of Thrombus	Treatment Strategy
McGee et al	1992	Arch.Surgery	26/F	Claudication	Infrarenal Aorta	Aortobifemoral grafting +Anticoagulation and Antiplatelet therapy
Poux et al	1996	Am J. Kidney Dis.	35/M	Abdominal pain	Pararenal aorta	Anticoagulation therapy
Dupont et al	2001	Nephrol Dial Transplant	46/F	Acute Ischemia	Suprarenal aorta	Aortic endarterectomy
DiCenta et al	2002	Annals of Vascular Surgery	46/F	Subacute Ischemia of the lower limbs	Infrarenal aorta	Aortobifemoral grafting +Anticoagulation therapy
Alfayate et al	2002	Vascular and Endovascular Surgery	38/F	Claudication	Pararenal aorta	Aortobifemoral grafting +Anticoagulation therapy
Letang et al	2005	Lupus	46/F	Lumbar pain	Pararenal aorta	Anticoagulation therapy
Ryu et al	2009	J Thorac Cardiovasc Surgery	57/M	Dyspnea	Ascending Aorta	Surgical removal of thrombus
Shroff et al	2011	The Journal of Reumatology	39/F	Abdominal pain	Pararenal aorta	Anticoagulation therapy
Toffon et al	2013	Case Report Surgery	68/F	Paresthesia and Pseudoclaudication in the lower limbs	Infrarenal Aorta	Aortobifemoral grafting +Anticoagulation therapy
Hsieh et al	2016	J Microbiol Immunol Infection	52/M	Claudication	Infrarenal Aorta	Anticoagulation therapy
Kadoya et al	2019	Vascular and Endovascular Surgery	60/M	Claudication	Infrarenal Aorta	Endovascular treatement

the aortic bifurcation proximally, which mimics the mechanics of a bifurcated graft used in open surgery.[10]

Results from the largest CERAB series have reported a primary patency rate of about 80% at one year and no peri-procedural mortality.[10] Additionally, in cases of isolated infrarenal aortic stenosis, endovascular stenting was reported to have good long-term patency and reduced perioperative mortality rates compared to surgical options.[9] As a result, the CERAB technique seems to be a feasible treatment option for APS-associated aortic thrombosis. However, another important aspect to underline is that treating surgeons must be aware that thrombus might dislodge into the renal arteries during treatment of juxta-renal aorto-iliac occlusions or in patients with a distance shorter than 2 cm between the ostium of the renal arteries and the beginning of the aortic lesion. In this perspective, some vascular surgeons have advocated the use of protective measures such as an extra dose of heparin and the position of two guidewires in both renal arteries through the brachial route to be ready to perform prompt angioplasty and stenting if needed.[10]

CONCLUSION

We reported a rare case of symptomatic thrombus of the infrarenal abdominal aorta due to APS. Although the relationship between APS and an isolated infrarenal aortoiliac lesion is still unclear, we speculate that APS might be one of the underlying causes of such conditions. Screening tests should be conducted in this group of patients, especially if their aorta contains a large amount of thrombus and low atherosclerotic burden.

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