Case Report

Acute heart failure due to chronic juxtarenal aortic occlusion in a patient with antiphospholipid antibody syndrome

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Abstract

Abdominal aortic occlusions are rare, but occasionally life threatening. A 48-year-old man was hospitalized due to acute heart failure accompanied by acute kidney injury (AKI). Abdominal ultrasound revealed deteriorating blood flow in the bilateral renal arteries. Subsequent abdominal aortography showed abdominal aortic occlusion just below the right renal artery and an occluded left renal artery. Dilated superior and inferior mesenteric arteries functioning as collateral feeding arteries suggested chronic occlusion. A hypercoagulation workup led to a diagnosis of antiphospholipid antibody syndrome (APS). This case report describes rare chronic juxtarenal abdominal occlusion in a patient with APS.

<Learning objective: Abdominal aortic occlusions are rare, but occasionally life threatening. The proximal propagation of aortic thrombosis might cause prerenal AKI. In our case, the renal arteries’ involvement of chronic juxtarenal arteries in a patient with APS gave rise to the onset of acute heart failure associated with prerenal AKI.>

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Introduction

Juxtarenal aortic occlusion might cause prerenal acute kidney injury (AKI) due to the proximal propagation of aortic thrombosis [1]. Antiphospholipid antibody syndrome (APS) is characterized by clinical evidence of arterial or venous thrombosis [2]. In contrast, APS is rarely accompanied by chronic aortic occlusion [3]. The present report describes a patient with APS in whom acute heart failure associated with prerenal AKI was provoked by renal artery involvement in chronic juxtarenal aortic occlusion.

Case report

A 48-year-old man with a history of myasthenia gravis (MG) was admitted to the intensive care unit under endotracheal intubation for assisted ventilation with the diagnosis of acute heart failure. The patient had a surgical history of thymectomy about 15 years before the admission. After the operation, he had been taking a regular treatment as an outpatient with oral pyridostigmine, which he had discontinued about 6 years before admission. He had no history of hypertension, although his blood pressure after he stopped attending our institute had been unknown. He had been smoking 10 cigarettes per day for 28 years, and had been apparently well and active before the admission.

Blood pressure at admission in the upper and lower limbs was 200/120 mmHg and 160/90 mmHg, respectively. The ankle brachial index in the right side and the left side was 0.78 and 0.75, respectively. Laboratory findings showed mildly elevated serum creatinine of 1.52 mg/dl, obviously elevated N-terminal pro-brain natriuretic peptide of 4687 pg/ml, prolonged activated partial thromboplastin time of 38.2 s (control: 27.8 s), mildly increased d-dimer level of 2.0 μg/ml, normal platelet count of 21.5 × 10^3/μl, normal prothrombin time of 11.3 s (control: 11.8 s), increased fibrinogen of 637 mg/dl, and normal level of fibrin/fibrinogen degradation products of 6.2 μg/ml. Serum lipid profile was as follows: triglyceride was 193 mg/dl, and high- and low-density lipoprotein cholesterol were 39 mg/dl and 124 mg/dl, respectively. Plasma renin activity and serum aldosterone levels were 7.1 ng/ml/h and 90.8 pg/ml, respectively. Twelve-lead electrocardiography revealed sinus tachycardia and left ventricular...
hypertrophy (LVH). Echocardiography demonstrated 23% left ventricular ejection fraction (LVEF) with diffuse hypokinetic LV wall motion, and diffuse LVH with a wall thickness of 14 mm. Diastolic function could not be evaluated because mitral inflow E wave was fused with A wave due to sinus tachycardia.

The patient’s condition continued to deteriorate because of anuria. The fractional excretion of sodium was 0.46%, indicating prerenal AKI. Abdominal ultrasound including color Doppler revealed that the size of the kidneys was normal and non-pulsating waves with a velocity of 10 cm/s in both renal arteries. Subsequent abdominal aortography revealed complete occlusion of the aorta just below the right renal artery, and dilated collateral arteries originating from the superior mesenteric artery (SMA) through the inferior mesenteric artery that fed into the distal portion of the aorta. The left renal artery was undetectable.

Circulating fluid volume was controlled by continuous hemodiﬀusion (CHDF), followed by intermittent hemodialysis. This improved the lung congestion and assisted ventilation was withdrawn on hospital day 5. After introducing CHDF, the volume of urine gradually increased sufficiently to maintain an appropriate circulating fluid volume. Repeated echocardiography showed that LV systolic function improved with LVEF of 40%. Mitral inflow E’ wave velocity of 55 cm/s, A wave velocity of 44 cm/s, and deceleration time of 204 ms were normal, while mitral tissue E’ velocity of 6.5 cm/s was low, indicating mild diastolic dysfunction. Oral amlodipine (5 mg/day) and carvedilol (10 mg/day) decreased blood pressure in the upper limbs to around 130/80 mmHg. The absence of organic stenosis on coronary angiograms ruled out ischemic heart disease.

Magnetic resonance (MR) signal intensity in MR angiography indicated that the occlusion site was occupied with thrombus (Fig. 1). Table 1 shows the findings of a hypercoagulation workup.

![Fig. 1. Findings of abdominal magnetic resonance angiography. According to the signal intensity, the occlusion site was probably occupied by thrombus (white arrow).](image)

In addition to vascular thrombosis, recurrently positive lupus anticoagulant, anticardiolipin antibody of the IgG isotype, and anticardiolipin-β2 GP1 complex indicated a diagnosis of APS [2]. There was no evidence of arterial or venous thrombotic diseases such as renal infarction, arterial embolism in the lower extremities, deep vein thrombosis, or pulmonary embolism. Anticoagulant therapy with intravenous heparin followed by oral warfarin was started.

Although the patient remained clinically stable, serum creatinine remained around 2.5 mg/dL. Three-dimensional high-resolution computed tomography (CT) angiography showed that the ostium of the right renal artery was obviously narrowed. A left renal artery was not identified, but collateral vessels from the aorta to the left kidney were detected (Fig. 2). CT images indicated no evidence of aortitis.

Bypass surgery proceeded under general anesthesia on hospital day 44. The outside of the abdominal aorta appeared normal, without evidence of aortic coarctation or arteritis. An SMA-bilateral renal artery bypass preceded using 5-mm polytetrafluoroethylene (Gore Tex™, Flagstaff, AZ, USA) grafts. This strategy increased blood flow in the renal arteries; renal Doppler ultrasound showed pulsating waves in both renal arteries with a peak velocity of 55 cm/s in right and 16 cm/s in left. However, his renal function did not improve. The time course of the estimated glomerular filtration rate (eGFR) is shown in Fig. 3. He remains in good condition as an outpatient under treatment with diuretics.

Discussion

Solitary occlusion of abdominal aorta without iliac lesion as in our patient is rare [4]. The occlusion appeared to be chronic because the collateral artery was well developed.

The main cause of renal dysfunction appeared to be prerenal AKI because the fractional excretion of sodium was 0.46%. The patient had no proteinuria, and image modality findings including ultrasound, CT, and MR imaging showed normal size of kidneys, no evidence of ischemic renal disease, sclerotic kidney, or renal infarction. The etiology of LV systolic dysfunction is unknown, although hypertension might be one of the causes, and ischemic heart disease was ruled out. The cause of hypertension was likely renovascular hypertension. In chronic renovascular hypertension, systemic renin–angiotensin–aldosterone system activation appears to be transient, and blood pressure subsequently sustained by alternative pressor pathways, such as oxidative stress and endothelial dysfunction [5].

Aortic occlusion can be precipitated by a hypercoagulable state with APS in addition to a history of smoking, although the precise mechanism of occlusion was unclear.

There are several case reports of concomitant APS and MG as in the present case. Anticardiolipin antibodies of the IgG isotype were found in 25% of patients with MG, and significantly associated with

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Hypercoagulable profile.</th>
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<tr>
<td></td>
<td>Reference range</td>
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<tr>
<td>Antibodies to CLβ2-GP1 complex (U/ml)</td>
<td>0–3.5</td>
</tr>
<tr>
<td>aCL antibodies of IgG isotype (U/ml)</td>
<td>0–10</td>
</tr>
<tr>
<td>Lupus anticoagulant (s)</td>
<td>0–6.3</td>
</tr>
<tr>
<td>Protein C (%)</td>
<td>65–135</td>
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<tr>
<td>Protein S (%)</td>
<td>70–150</td>
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<tr>
<td>Antithrombin III (%)</td>
<td>80–130</td>
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CL, cardiolipin; GP1, glycoprotein-1; aCL, anticardiolipin.
Fig. 2. Findings of three-dimensional high-resolution computed tomography. (A) Anteroposterior view shows dilated superior and inferior mesenteric arteries functioning as collateral feeding vessels. Lower aorta distal to occlusion and iliac arteries appear free of lesions. (B) Enlarged right anterior oblique view around right renal artery shows obvious narrowing in ostium of right renal artery. (C) Enlarged left anterior oblique view around left renal artery shows collateral vessels from aorta to left kidney. Left renal artery is not evident.

thymic abnormalities such as thymoma or thymic hyperplasia [6]. Our patient had a history of thymectomy which might be responsible for the occurrence of APS, although the data are scarce and subject to controversy.

The enhanced CT gave us much important anatomical information, while the use of contrast medium was undesirable in the present case with renal dysfunction. Unfortunately, the image of MR angiogram was not clear. Therefore, we made a distressing decision to perform enhanced CT. Saline and sodium bicarbonate hydration were applied before the CT. Fortunately, the effect of enhanced CT on eGFR seemed to be minimum (Fig. 3). The focal aortic lesion in our patient together with the well-developed collateral circulation indicated underlying anomaly at the occlusion site. One possible anomaly was abdominal aorta coarctation [7], which was not identified in our patient during surgery. Takayasu’s arteritis, which is another possible underlying condition, was also ruled out on the basis of image modality findings and the appearance of outside wall during the operation. Small aorta syndrome is a congenital vascular anomaly characterized by highly bifurcated abdominal aorta (usually at the L3 level), hypoplastic iliofemoral artery system, and a maximum aorta diameter <14 mm with a distance of 10 mm from the bifurcation. None of these characteristics were met with our patient, denying small aorta syndrome.

Our patient was surgically treated with a SMA-bilateral renal artery bypass to increase tissue perfusion. There are several studies that showed a negative result of such an interventional strategy for renal function. The ASTRAL study is one of the most important studies [8]. However, the ASTRAL study has an important limitation that the patients with severe renal artery stenosis who should be treated with revascularization were unlikely to be enrolled. Because our patient had severe stenosis in the right renal artery, we considered that bypass surgery would be beneficial on the basis of a case-sensitive approach. Aorto-bifemoral bypass with renal artery reconstruction was a more fundamental therapeutic option that could be considered [9] and we selected this strategy because aorto-bifemoral bypass is more invasive and the patient lacked ischemic manifestation in the lower limbs. Thromboendoarterectomy is another option for surgery but was not done because the risk of restenosis appeared to be high in small arteries such as renal arteries, and abdominal aortic aneurysm can occur in the future due to thinning of aortic wall. Unfortunately, post-operative renal function did not improve. The renal damage appeared to be irreversible at the time of the surgery. However, blood supplied to the kidney through the bypass might prevent the progression of renal insufficiency if renal artery involvement had progressed due to the proximal propagation of aortic thrombus. In addition, this bypass surgery would be beneficial for our patient, in terms of prevention of recurrent cardiorenal syndrome [10].

Fig. 3. Time course of the estimated glomerular filtration rate (eGFR). The eGFR decreased rapidly after admission. After the fluid volume was controlled properly by continuous hemodialfiltration (CHDF) and hemodialysis (HD), the eGFR recovered to around 30 ml/min/1.73 m². Unfortunately, reduced renal function did not improve after the bypass surgery, while the effect of enhanced computed tomography appeared to be minimum. The eGFR has been stable around 20 ml/min/1.73 m² after the discharge.
Conflict of interest

The authors declare no conflict of interest.

References


