Internal Medicine
The Japanese Society of Internal Medicine

【 CASE REPORT 】

Bilateral Acute Renal Infarction Due to Paradoxical Embolism in a Patient with Eisenmenger Syndrome and a Ventricular Septal Defect

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Abstract:

A 52-year-old man who was diagnosed with Eisenmenger syndrome due to a muscular-type ventricular septal defect 30 years previously, visited our emergency room after experiencing six hours of severe left flank pain and vomiting. On laboratory examination, azotemia and microscopic haematuria were identified. Contrast-enhanced computed tomography also revealed pulmonary embolism (PE) and bilateral acute renal infarction. The flank pain resolved after heparin was administered for anti-coagulation and aspiration thrombectomy was performed. The patient was discharged on warfarin as anticoagulant therapy. In this case, a paradoxical embolism was considered to have been the cause of PE and bilateral acute renal infarction in a patient with Eisenmenger syndrome.

Key words: renal infarction, pulmonary embolism, Eisenmenger syndrome, paradoxical embolism

(Intern Med 60: 3937-3940, 2021)
(DOI: 10.2169/internalmedicine.7549-21)

Introduction

Renal infarction can occur due to a hypercoagulable state, renal artery injury, or cardiogenic problems, but bilateral acute renal infarction is rare (1, 2). Eisenmenger syndrome is a rare pathophysiological condition associated with severe pulmonary hypertension with shunt reversal in very different cardiac malformations (3).

Pulmonary embolism (PE) has been reported to be associated with Eisenmenger syndrome (4, 5), but not renal infarction. A paradoxical embolism can be an important clinical feature in patients with venous thromboembolism and cardiac or pulmonary shunts (6).

We herein report a rare case of bilateral acute renal infarction due to Eisenmenger syndrome associated with a paradoxical embolism.

Case Report

A 52-year-old man visited the emergency room with severe left flank pain and vomiting that had developed six hours prior to presentation. There were no other symptoms. His vital signs were stable. Left side costovertebral angle tenderness was observed on physical examination.

He was diagnosed to have Eisenmenger syndrome with large size (24-25 mm) muscular-type ventricular septal defect (VSD) (Fig. 1) 30 years previously and secondary erythrocytosis 20 years prior to this presentation. He had been awaiting heart-lung transplantation for one year. There was no history of nephrolithiasis. His medications included bosentan, valsartan, digoxin, and spironolactone for nine years since being diagnosed with pulmonary hypertension and heart failure.

A chest X-ray revealed cardiomegaly without pulmonary oedema. There were no abnormal findings on abdominal X-rays. Laboratory studies on presentation showed azotemia
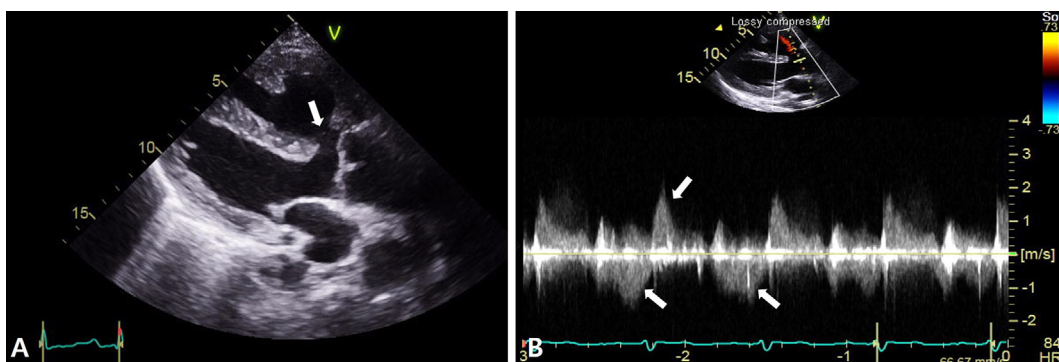
Figure 1. Transthoracic echocardiography. Muscular-type ventricular septal defect (A) with a bi-directional flow (B).

Table.   Initial Laboratory Findings.

	Value	Normal value	Unit
WBC	8.43	4.00-10.00	$\times 10^3/mm^3$
Haemoglobin	17.7	13.0-17.0	g/dL
Haematocrit	58	39-52	%
Platelet	138	130-400	$\times 10^3/mm^3$
BUN	15.9	6-20	mg/dL
Creatinine	1.67	0.6-1.2	mg/dL
Lactate dehydrogenase	>700	135-225	IU/L
Troponin-I	2.15	0-0.1	ng/mL
Uric acid	5.7	3.4-7.0	mg/dL
Prothrombin time	14.1	11.9-14.3	sec
Prothrombin time (INR)	1.1	0.8-1.2	
aPTT	35.3	29.1-43.5	sec
D-dimer	0.44	0.0-0.5	FEU μg/mL
Urine RBC	50-99	0-4	/HPF
Urine protein	2+	-	

aPTT: activated partial prothrombin time, BUN: blood urea nitrogen, INR: international normalized ratio, RBC: red blood cell, WBC: white blood cell

and microscopic haematuria. His haemoglobin level, troponin-I level, and lactate dehydrogenase level were elevated (Table).

Chest computed tomography (CT) revealed thrombi in the right middle lobe segmental and subsegmental pulmonary arteries. Abdominal CT demonstrated total occlusion of the left main renal artery and right renal arterial branch (Fig. 2). Immediately after diagnosing acute bilateral renal infarction, we administered anticoagulation therapy with intravenous heparin and performed aspiration thrombectomy by urokinase on the left main renal artery (Fig. 3). Right renal thrombectomy was not performed because the right renal perfusion was maintained even though the occlusion in right renal arterial branch. After the procedure, renal perfusion recovered, and his flank pain was resolved.

His coagulation factor assays were within the normal limits and there were no specific findings on 24-hour Holter monitoring. In addition, we could not find any deep vein thrombosis on ultrasonography. Intravenous heparin was followed by oral warfarin.

During hospitalization, the serum creatinine level was elevated to 3.05 mg/dL and then it decreased to 2.62 mg/dL on the 8th day of hospitalization. When azotemia showed an improvement, the patient was discharged. The serum creatinine level fell to 1.48 mg/dL while being treated on an outpatient basis. His PE decreased two months after starting treatment. However, he died eighteen months later due to end stage heart failure with cardiogenic shock.

Discussion

The causes of renal infarction are divided by cardiac disease, renal artery injury, and a hypercoagulable state. The most common cause of cardiac disease is atrial fibrillation followed by cardiomyopathy, artificial valve, endocarditis and thrombi from suprarenal aorta or left ventricle, in that order (1, 7).

Renal infarction is rarely reported in association with congenital heart disease (CHD). Dilatation or an aneurysm of the large vessels such as the pulmonary trunk in CHD may lead to thrombosis due to stasis of the blood flow and the formation of mural thrombi (8, 9). In addition, chronic hypoxemia and increased blood viscosity in CHD patients can lead to endothelium damage. This vessel wall damage can affect thrombus formation (10). The impaired fibrinolytic system and the release of procoagulant factors in Eisenmenger syndrome can also influence thrombus formation (11, 12).

Eisenmenger syndrome can lead to PE via biventricular dysfunction and a reduced pulmonary blood flow (4, 5). Anatomic right-to-left shunt can allow venous emboli to pass into the arterial circulation (13). Paradoxical embolism is rare clinical entity among patients with venous thromboembolism in the presence of intracardiac or pulmonary shunts. The clinical presentation is diverse and the condition is potentially life-threatening (6). The common presence of anatomic cardiac shunts is recognized as patent foramen ovale (14).

The symptoms of paradoxical embolism vary depending on the region of embolism. In the case of stroke caused by paradoxical embolism, the symptoms include hemiplegia and

Figure 2. Chest and abdominal enhanced computed tomography (CT). Chest CT showed thrombi in the right middle lobe segmental and subsegmental pulmonary arteries (A). Abdominal CT revealed thrombi in the right renal arterial branch (B, arrow) and left main renal artery (C, arrow).

Figure 3. Renal angiography. Renal angiography showed occlusion of the left main renal artery (A, arrow). The renal blood flow was restored in the left main renal artery (B, arrow) and arterial branches (C, arrow) by aspiration thrombectomy.

aphasia (15). Chest pain and electrocardiographic changes are associated with myocardial infarction (16). Symptoms such as acute abdominal pain, back pain, and haematuria can be caused by acute mesenteric ischemia or renal infarction (14, 17), and peripheral arterial occlusion due to embolism can seriously threatens the viability of the limbs (18). Although the serious nature and complications of paradoxical embolism have been recognized, this disease entity remains under-reported. As a result, we should consider further evaluation if there are ischemic symptoms and signs based on detailed history taking and physical examination.

The patient described herein had Eisenmenger syndrome due to VSD. His thrombogenic factors were suspected to be CHD and secondary erythrocytosis. We suspected that PE led to near-total renal infarction of both renal arteries via paradoxical embolism by Eisenmenger syndrome.

The treatments used for renal infarction include anticoagulation, percutaneous endovascular therapy like thrombolysis or thrombectomy with or without angioplasty or stent placement, and open surgery (19, 20). Thrombectomy is recommended if the renal blood flow is not restored within 6 hours and total obstruction of the renal artery is observed (20, 21). Anticoagulation is the standard therapy in thromboembolic diseases. The intensity and duration of the therapy depends on the underlying diseases (22). But anticoagulation for primary prevention in patients with Eisenmenger syndrome remains controversial (23, 24).

The patient arrived early after the onset of symptoms, and both sides were invaded, the range of renal infarctions were wide, and he showed severe azotemia, so we performed thrombectomy immediately and the patient's renal function thereafter successfully recovered.

This rare case report emphasizes the importance of considering paradoxical renal infarction in patients with PE underlying Eisenmenger syndrome. In addition, we should attempt to actively restore renal perfusion via intervention in patients with main renal artery occlusion which has significantly reduced the kidney function.

The authors state that they have no Conflict of Interest (COI).

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