Case report

Paradoxical renal embolism in a patient with congenital cardiac malformation

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Introduction

Paradoxical embolism is a rare phenomenon in which thrombotic material passes through a right–left shunt to the systemic circulation. This most often occurs across a patent foramen ovale.1 Usual sites of embolism are the brain and the extremities, paradoxical embolism into the kidney is much less common.2

Case report

A 35-year-old man was admitted with sudden new-onset pain in his right flank. He had been born with a complex cardiac malformation consisting of double outlet right ventricle with malposition of the great arteries, straddling of the tricuspid valve and large atrial and ventricular septal defects. During the newborn period, pulmonary artery banding was conducted. Three years later corrective surgery was attempted; however, de-banding of the pulmonary artery resulted in pulmonary congestion. Thus, pulmonary artery banding was restored and a palliative Mustard procedure was performed to achieve an increase in systemic arterial oxygen saturation by changing streaming and reducing the amount of venous blood that reaches the aorta (Figure 1). After this surgery, the patient developed relatively normal with only minor physical limitations. At the age of 33 years, cardiac function progressively deteriorated resulting in repetitive cardiac decompensation. Two months before the current admission, the patient presented with numbness and tingling of his left arm and leg, which was interpreted as a transient ischemic attack. Thromboembolism due to intermittent atrial fibrillation was assumed to be the most likely source, because the patient reported palpitations, and atrial tachycardia is a common late arrhythmia after the Mustard procedure.3

The patient was therefore started on phenprocoumon and the target International Normalized Ratio (INR) was set between 2 and 3. At the current admission, the patient described a progressive excruciating pain in his right flank that had abruptly started during the night before. Several days prior, he had doubled the dose of his diuretic since he had noticed increasing leg edema. At about the same time he had experienced a new episode of transient numbness and tingling of the left side of his body which ceased spontaneously after several hours. On physical examination, there was severe cyanosis of the face and hands and clubbing of fingers and toes. The pulse was 77/min and regular. The blood pressure was 105/75 mmHg. The jugular venous pressure was slightly increased. The lungs were clear. There was a holosystolic thrill over the anterior and posterior chest, which was maximal at the fourth left intercostal space. Heart sounds were normal. There was tenderness in the abdominal right upper quadrant and right costovertebral angle. All peripheral pulses were palpable. Routine
Laboratory examinations were remarkable for increased hemoglobin and hematocrit (20.2 g/dl; normal range 13.5–17.5 and 59.9%; normal range 41–53%), increased lactate dehydrogenase (LDH) (466 U/l; normal range <248) and increased aspartate aminotransferase (AST) (53 U/l; normal range <35). The patient had a subtherapeutic INR of 1.35 and elevated d-dimers (1.31 mg/l; normal range <0.5). Arterial blood gases at room air showed a pO2 of 46 mmHg, pCO2 of 35 mmHg, pH of 7.44 and HCO3 of 23.3 mmol/l. Since urolithiasis or renal infarction was suspected, a computed tomography was performed. It showed no signs of kidney stones, but a large area of nonperfused parenchyma in the middle and caudal pole of the right kidney (Figure 2). Treatment with unfractionated heparin was initiated; this resulted in symptomatic relief after 12 h. Heparin was then stopped and enoxaparin was started. Serum creatinine increased from 87 μmol/l at admission to a maximum of 116 μmol/l. In parallel, LDH increased to 558 U/l and AST to 71 U/l. A transthoracic echocardiography showed no signs of intracardial thrombotic material, but massive tricuspid valve regurgitation and a grossly enlarged right ventricle, with severe systolic dysfunction. As in all previous visits, the electrocardiogram showed a sinus rhythm with a complete right bundle branch block. The patient was instructed about the importance of maintaining a target INR of 2–3 and was discharged. An ambulatory 24-h electrocardiogram was recommended but was not realized until the patient presented again 3 months later. This time he suffered from severe pain in the region of his left kidney. The INR was 1.68 and d-dimers were slightly elevated (0.78 mg/l). Serum creatinine was 76 μmol/l. Renal infarction of the left kidney was suspected and confirmed by contrast-enhanced sonography revealing a normally perfused right kidney but a considerable perfusion defect in the cranial pole of the left kidney. No signs of embolism were found in other organs, including spleen and liver. The electrocardiogram at admission showed no new changes and sinus rhythm. On further questioning, the patient told us that he had recently increased his diuretic dose again because of leg edema. At that time, he had experienced cramps of his left calf. However, he had paid no attention to this symptom since he had been told that diuretics can cause muscle cramps due to electrolyte imbalances. On physical examination, we found a slightly increased circumference of his left calf. To rule out thrombosis, a
duplex ultrasound was performed showing massive deep vein thrombosis in the left popliteal and fibular veins. Compression stocking therapy was initiated and once the phenprocoumon dose was correctly adjusted, the patient was discharged home. His renal function remained stable with a serum creatinine of 80 μmol/l at discharge.

Discussion

Acute renal infarction can be due to a variety of predispositions, but atrial fibrillation is the most common cause. Accordingly, intermittent atrial tachycardia was the initially assumed diagnosis in our patient, who was predisposed to develop atrial arrhythmias. However, all electrocardiograms recorded showed a continued sinus rhythm and echocardiography revealed no signs of atrial thrombotic material. Retrospectively, it is much more likely that the recurrent ischemic events were all caused by paradoxical embolism of thrombotic material across the patient’s large ventricular septal defect (VSD).

Our patient presented with three factors predisposing to the development of deep vein thrombosis: (i) heart failure under diuretic therapy; (ii) insufficient anticoagulation; and (iii) erythrocytosis. Erythrocytosis is considered an important factor in the pathogenesis of ischemic events and it has been shown that an elevated hematocrit increases the risk of venous thromboembolism in the general population. Our patient had marked secondary erythrocytosis. However, it is important to note that secondary erythrocytosis in cyanotic heart disease is a physiological response to chronic hypoxia and is functionally different from primary erythrocytosis. Cumulative evidence suggests that phlebotomy in patients with cyanotic congenital heart disease does not only decrease exercise tolerance but can also increase ischemic events. Short-term fluctuations in hematocrit and blood viscosity may contribute to the generation of thromboembolic complications, and that this association may be higher in patients with chronic erythrocytosis. In this respect, it was remarkable that both episodes of renal infarction in our patient were associated with a sudden increase in diuretic intake. It is possible that this may have caused a relative volume depletion contributing to higher blood viscosity and thromboembolic events.

To our knowledge, this case is the first documentation of paradoxical embolism to the kidneys from a venous thromboembolic source in a patient with congenital cyanotic heart disease. Most commonly, paradoxical embolism occurs across a patent foramen ovale and in the majority of cases it leads to cerebral and not to peripheral embolism. The appropriate treatment of thromboembolism in patients with cyanotic heart disease is an efficient anticoagulation therapy. However, as illustrated in our patient, this simple treatment can be sometimes difficult to control.

Conflict of interest: None declared.

References