SUDDEN CARDIAC DEATH CAUSED BY AN OBSTRUCTIVE LEFT ATRIAL BALL THROMBUS

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Abstract

Systemic embolization due to left atrial (LA) thrombus in patients with rheumatic mitral stenosis (RMS) and atrial fibrillation (Af) is common. However, a large ball thrombus in the LA in RMS patients is rare. It has potentially catastrophic consequences such as total occlusion of the narrowing mitral valve orifice, often resulting in sudden cardiac death. We herein report a case of 68-year-old woman, who died suddenly due to an obstructive LA thrombus.

Key words: Mitral stenosis, Large ball thrombus, Echocardiogram

Introduction

About 10-25% of patients with rheumatic mitral stenosis (RMS) have left atrial (LA) thrombi.¹ Among those RMS patients with atrial fibrillation (Af), the incidence of LA thrombi was even higher.² Mobile LA thrombi have a high rate of systemic embolization.³⁴ Although a large ball thrombus in the LA in RMS patients is rare, it has potentially catastrophic consequences such as total occlusion of the narrowing mitral valve orifice, often resulting in sudden cardiac death. We herein report a case of 68-year-old woman, who died suddenly due to an obstructive LA thrombus.

Case Presentation

A 68-year-old woman was admitted to the hospital because of shortness of breath and progressive leg edema of one week duration. The patient had RMS and Af, which had been diagnosed about six years before the current admission. She also had a six-year history of hypercholesterolemia. Her medications consisted of aspirin, amiodarone, pravastatin, and a fixed-dose combination antihypertensive drug moduretic (amiloride and hydrochlorothiazide). Five years before admission, she suffered from an episode of left-side limb weakness and slurred speech. A computed tomographic scan of the brain, obtained without the administration of contrast material, showed a right lobe lacunar infarction. Warfarin therapy was begun. The prothrombin time was prolonged, with the international normalized ratio (INR) kept around 1.7 when she was taking anticoagulant medication. Warfarin therapy had been stopped 10 days before admission because of hemorrhagic erosive gastritis by a gastroenterologist documented by panendoscopy. Approxi-
Immediately one week earlier, the woman noticed increased shortness of breath and progressive leg edema. She came to this hospital and was admitted.

On physical examination, the blood pressure was 120/70 mm Hg, the heart rate was irregular, with 108 beats per minute, the respiratory rate 24 breaths per minutes, and the temperature 37°C. An examination of her neck showed a jugular venous pressure of 6 cm of water above sternal angle at 45°. The carotid pulses were equal and there were no bruits. There were wheezing and crackles in both lower lung fields. On auscultation, a loud first heart sound, opening snap, and a grade I/VI diastolic rumbling murmur at apex was noted. Abdominal and neurologic examinations showed no abnormalities. There was +++ peripheral edema.

The hemoglobin was 12.8 g/dl. The prothrombin time was 9.9 seconds (control 8.0-12.0 seconds); the INR was 0.99. The activated partial thromboplastin time was 24 seconds (control 23.9-35.5 seconds). An electrocardiogram (EKG) showed atrial fibrillation at a rate of 108 beats per minute without Q-T interval prolong. A chest radiograph showed increased pulmonary infiltrates, suggesting of pulmonary edema. A transchachoracic echocardiogram revealed moderate mitral stenosis (mitral valve area = 1.6 cm², estimated by pressure half-time method), trivial mitral regurgitation, and adequate systolic left ventricular function. However, no definite LA thrombus was found (Figure 1A).

Intravenous furosemide was administered. Amiodarone and pravastatin were continued, and intravenous nitrate and diltiazem were added. No anticoagulant was administered. The patient’s congestive symptoms improved gradually. On the 5th hospital day, a large frail, mobile, ball-shaped thrombus (3.0 x 3.2 cm in size) attached to the LA free wall was demonstrated by a follow-up echocardiogram (Figure 1B). Intravenous unfractionated heparin of 800 units per hour was given and emergency surgical removal of the thrombus was strongly recommended. However, the patient and her families rejected it. Unfortunately, the patient died suddenly on the 6th hospital day. EKG monitor didn’t show polymorphic ventricular tachycardia or ventricular fibrillation before asystole during this hospitalization. A bedside emergency echocardiogram confirmed that the mitral valve orifice was stuffed by the dislodged LA thrombus, resulting in total obstruction of blood flow.

![Figure 1A,1B. Parastenal long-axis view (A and B), recorded in the present patient with rheumatic mitral stenosis. A: Typical diastolic doming of the mitral valve with focal thickening at the tips of both the anterior and posterior leaflets were demonstrated on admission, no thrombus formation obvious. B: A large ball thrombus (3x3.2cm, arrow) attached to the free wall of left atrium was demonstrated on the 5th hospital day.](image-url)
Discussion

Atrial fibrillation is a common complication of RMS due to the elevation of LA pressure and consequent LA enlargement.\(^5\) The prevalence of Af in patients with RMS is related to the severity of valve obstruction, age, and the presence of other valvular abnormalities, such as tricuspid regurgitation. Atrial fibrillation predisposes to LA thrombus formation in RMS patients. Systemic embolism due to LA thrombus in patients with RMS and Af is common. The most common site for clinical evidential embolism is the cerebral circulation, but any organ may be involved. Although a large thrombus in the LA in RMS patients is rare, it may be potentially lethal because that the fragmentation or dislodgement of the thrombus may produce systemic embolization of vital organs or total occlusion of the mitral valve orifice, resulting in sudden death.\(^6,8\)

Current guidelines recommended that anticoagulant therapy is indicated in prevention of systemic embolism in RMS patients with Af, especially in those who have prior embolic events and those patients with documented LA thrombus. Anticoagulant therapy may also be considered in patients with severe RMS and sinus rhythm when LA diameter of more than 55 mm or spontaneous contrast were demonstrated on echocardiogram. The international normalized ratio should be maintained between 2.0 and 3.0 when the patient is taking anticoagulant therapy.\(^9\)

However, maintaining the safety of anticoagulant therapy is a substantial challenge because of problems with compliance, concurrent medical or surgical conditions, drug interactions, and variability in laboratory measurements. Inadequate anticoagulation may allow thrombus formation. In this patient, warfarin therapy had been stopped 10 days before admission because of hemorrhagic erosive gastritis. The INR was 0.99 on admission, when she was not taking anticoagulant medication. Thrombosis in the LA developed on the 5th hospital day.

Once echocardiographic evidence of frail, mobile, or free-floating ball thrombus is established, prompt surgical removal of the thrombus, often combination with mitral valve repair or replacement, is advocated as the treatment of choice.\(^8,10\) Thrombolysis with urokinase, streptokinase, and tissue plasminogen activator or anticoagulation with high-dose unfractionated heparin or low-molecular-weight heparin has been reported, but the risks of hemorrhagic or embolic complications should be taken into consideration.\(^7,11-13\)

Therefore, anticoagulation and thrombolytic therapy for RMS patients with large mobile or free-floating LA thrombus should be reserved only for those who in extremely high-risk for perioperative morbidity and mortality or those patients refusing surgery. In the present case, even though emergency surgical removal of the thrombus was strongly recommended, the patient declined it. Unfortunately, she died suddenly the next day owing to dislodgement of the LA thrombus, resulting in total obstruction of the narrowing mitral valve orifice.

In conclusion, RMS patients with large mobile or free-floating thrombi are at high risk of embolism and sudden death. Emergency surgery with valve replacement or repair has been the treatment of choice for these patients. Based on the limited data of clinical experiences, thrombolytic therapy or high-dose anticoagulant therapy remain controversial and should only be considered in selected patients with high surgical risk, or those refusing surgery.

References

4. Tsioufis CP, Stefanadis CI, Tsiamis EG, ea al. A free
左心房球狀血栓塞住二尖瓣開口引發猝死：一病例報告

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摘要

左心房血栓在風濕性二尖瓣狹窄合併慢性心房顫動的病人相當常見，且容易引起全身性栓塞。雖然風濕性二尖瓣狹窄病人發生左心房球狀血栓很清楚，但一旦掉落卻有很高的機率造成狹窄的二尖瓣出口完全堵塞，導致猝死。我們在此報告一位68歲女性因暫停抗凝藥物導致左心房球狀血栓掉落堵塞二尖瓣開口導致猝死的病例。

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