Concurrent Pheochromocytoma, Ventricular Tachycardia, Left Ventricular Thrombus, and Systemic Embolization

Wei Zhou and Shi Fang Ding

Abstract

An obese 43-year-old female with a right adrenal pheochromocytoma is described. The clinical manifestations of this case included ventricular tachycardia, left ventricular thrombus, and elevation of serum myocardial enzymes. During the hospitalization, the left ventricular thrombus was detached, leading to renal infarction and embolic occlusion of the femoral arteries bilaterally.

Key words: pheochromocytoma, left ventricular thrombus, ventricular tachycardia, thromboembolism, amputation


Introduction

The main clinical manifestation of pheochromocytoma is paroxysmal hypertension, accompanied by arrhythmias, heart failure, and even sudden death due to catecholamine secretion (1). We report an unusual case of a pheochromocytoma, with concomitant ventricular tachycardia, a left ventricular thrombus, and a systemic embolism.

Case Report

An obese 43-year-old woman was admitted to the intensive care unit due to acute rapidly progressive dyspnea, tachycardia, and vomiting. Three months earlier, the patient was diagnosed with viral myocarditis based on an abnormal myocardial zymogram at a local hospital; there were no abnormalities of cardiac structure. About 5 days before admission, the patient presented with severe dyspnea, tachypnea, fever (39.6℃), and tachycardia. The physical examination showed a heart rate of 130 beats per minute (bpm), with no heart murmurs or an irregular rhythm. Her blood pressure was 120/80 mmHg, and the peripheral pulses were normal. Her respiratory rate was 40 breaths/min, and extensive moist rales were audible in both lungs, with no signs of systemic congestion. She had frequent vomiting, but her abdomen was flat and soft. The laboratory data showed the following: WBC, 18.7×10^9/L; serum potassium, 3.0 mmol/L; serum glucose, 18.6 mmol/L; cholesterol, 10.7 mmol/L; triglycerides, 10.34 mmol/L; the MB isoenzyme of creatine kinase (CK-MB), 112 U/L (normal, <25 U/L); and the cardiac troponin I (cTnI), 1.6 ng/mL (normal, <0.06 ng/mL). The electrocardiogram showed poor R wave progression in V2, Q waves in II, III, V₅, and V₇-V₉, and ST segment elevation in V₅-V₉ (Fig. 1A). A high-resolution CT scan showed signs of severe pulmonary infiltration. Subsequently, carbapenem and teicoplanin were administered intravenously to control pulmonary infection, and the hyperglycemia was treated with insulin.

Five hours after admission, the patient experienced non-symptomatic sustained monomorphic ventricular tachycardia (VT) with a pulse (Fig. 1C). However, the patient’s VT attacks were accompanied with a systolic blood pressure of about 200 mmHg, which dropped to about 90 mmHg within a few minutes without treatment. Amiodarone (150 mg) was administered intravenously and the VT converted to sinus rhythm. However, she continued to have frequent intermittent episodes of non-sustained or sustained repetitive monomorphic VT occurring several times per hour, despite intermittent intravenous amiodarone treatment (75 or 150 mg each administration). A 24-hour Holter monitor showed 70 runs of repetitive monomorphic VT, lasting for 30 seconds.

to 12 minutes (Fig. 1D). On the 2nd day, the Q waves in V3 and V4 disappeared (Fig. 1B). On the 3rd day, CK-MB and cTnI were 95 U/L and 6.27 ng/mL, respectively. At the same time, on the 6th day, the serum potassium decreased to 2.6 mmol/L, accompanied by diffuse symmetric widespread T wave inversion on the ECG (Figs. 1E, 1F). The administration of KCl (9 g/day) was started for the hypokalemia. Thereafter, the deeply inverted T waves improved (Figs. 1G, 1H). Amiodarone was given as an intravenous loading dose of 150 mg over 10 minutes, followed by an intravenous maintenance dose of 1 mg/min for 6 hours; then an intravenous maintenance dose of amiodarone at 0.5 mg/min for 48 hours was administered. Oral amiodarone was started at 600 mg/day for 1 week. The VT episodes were substantially diminished and terminated on the 8th day. On the 8th day, the 24-hour urine vanillylmandelic acid was 135.1 umol (normal, 0-68.6 umol). Phenoxybenzamine (5 mg, twice a day) was administered.

On day 2, the patient underwent cardiac duplex ultrasound because of VT attacks and a suspected acute myocardial infarction. Echocardiography revealed 28 mm×17 mm and 0.7 mm×0.8 mm thrombi in the left ventricular apical region, accompanied with inferior wall akinesis (ejection fraction, 50%), but there was no suggestion of hypertrophy or dilated cardiomyopathy. On the 3rd day, contrast-enhanced CT revealed a 4.0 cm×3.0 cm×3.0 cm heterogeneous right adrenal mass (Fig. 2A), a thrombus attached to the left ventricular apex (Fig. 2B), and a wedge-shaped filling defect area of the left kidney (Fig. 2C). A renal infarction of cardiac embolic origin was assumed. Antithrombotic treatment with subcutaneous dalteparin sodium (5,000 units/12 hours) and anticoagulation therapy with warfarin sodium
Figure 2. A: enhanced CT revealed a heterogeneous enhancement of right adrenal mass. B: enhanced CT showed a thrombus attachment to the apex of left ventricular; C: enhanced CT showed segmental infarction of the left kidney; D: digital subtraction angiography showed cardionic embolic occlusion of the distal segment of left femoral artery. E: wet gangrene developed in bilateral legs; F: magnetic resonance angiogram showed multiple embolic closures or severe stenosis in proximal peroneal, anterior and posterior tibial arteries on both sides.

(3 mg/per day) were initiated. However, thrombolysis was not performed and warfarin therapy was terminated 5 days later.

On the 5th hospital day, the patient complained of sudden right calf severe pain, accompanied by coldness and numbness. Gradually, the pain became increasingly severe, with erythematous lesion formation, and cyanotic toes, accompanied by pulseless bilateral dorsal arteries. Suspecting an acute arterial embolism, the patient underwent a duplex sonographic examination on the 7th hospital day. Duplex sonography revealed no stenosis or occlusion of the femoral arteries bilaterally. However, echocardiography revealed that the left ventricular thrombus had reduced in size to 13 mm × 11 mm, the inferior wall was moderately hypokinetic, and the left ventricular ejection fraction improved to 69%. Nevertheless, the skin of the toes progressed toward infarction.
On the 15th hospital day, emergency cardiovascular catheterization was performed. The coronary angiography was normal, the left ventriculography showed that the left ventricular apical thrombus had disappeared and the dyskinetic inferior wall had total resolved. Nevertheless, angiography showed complete occlusion of the distal segment of bilateral femoral arteries (Fig. 2D).

The patient underwent urgent transfemoral thrombectomy using a Fogarty balloon catheter. The dorsalis pedis pulses were successfully restored after a large amount of fresh and old mixed thrombus was removed. After surgery, local intraarterial fibrinolysis using urokinase (20,000 units/hr) was infused via the right femoral artery sheath and continued for 5 days. Oral anticoagulation therapy was initiated. On the 24th hospital day, repeated contrast-enhanced CT revealed several new occurrences of focal infarction in the kidneys bilaterally, and the left ventricular apical thrombus disappeared. The patient continued to experience severe pain after the thrombectomy. Pulsation of the bilateral dorsal arteries were gradually not palpable. Therefore, the ischemic changes progressed to wet gangrene in the lower limbs, which required amputation (Fig. 2E). On the 28th hospital day, a magnetic resonance angiogram showed multiple embolic closures or stenosis in the bilateral peroneal, and anterior and posterior tibial arteries (Fig. 2F). On the 31st hospital day, a high level below the knee amputation was performed bilaterally. Three months later, after preparation with alpha and beta adrenergic blockade, a right adrenalectomy was conducted. An approximately 5 cm x 7 cm mass was removed from the right adrenal gland without complications. Histopathologic examination revealed a pheochromocytoma. The patient was discharged in stable condition, with no recurrence of arrhythmia and normal blood pressure.

**Discussion**

It has been shown that catecholamines secreted from pheochromocytomas cause myocardial necrosis, focal myofibrillar degeneration, and subsequent fibrous scar formation (1). Although acute cardiitis as a primary manifestation of pheochromocytoma has been reported (2), acute myocardial infarction was not excluded in the present patient on the basis of ECG and cardiac biomarkers (3). Therefore, myocardial damage due to coronary artery spasm could not be ruled out since the patient had multiple risk factors, including left ventricular mural thrombus, hypertriglyceridemia, hyperglycemia, and catecholamine secretion. Myocardial lesions resulting from catecholamines and acute myocardial ischemia might have been the causative mechanism for the cTn I and CK-MB elevation (4).

Pheochromocytoma can present with life-threatening ventricular arrhythmias due to the sudden release of catecholamines (5, 6). However in this case, in the 1st week, VT was unsuccessfully terminated due to hypokalemia and non-standard amiodarone use (7). The patient developed hypokalemia due to vomiting, insufficient potassium administration, and insulin administration for hyperglycemia. T wave electrocardiographic abnormality is one characteristic of catecholamine-induced cardiomyopathy (3). However, hypokalemia was a precipitating factor for both serial abnormal T wave changes and incessant VT.

Duplex ultrasound examination is a first-line screening tool in arterial thromboembolic disease, but is limited by operator experience (8). There were four reasons responsible for the late diagnosis of arterial thromboembolism in the case described herein. First, we did not pay sufficient attention to the possibility of a left ventricular thrombus embolism. Second, the ischemic leg pain was subjectively attributed to arteriospasm due to catecholamine secretion. Third, the left ventricular thrombus was gradually detached and embolism developed in the distal lower extremity arteries. Fourth, we did not rationally use various imaging modalities in diagnosing arterial thromboembolism and monitoring the thrombectomy effect.

A left ventricular thrombus occurring in a patient with a pheochromocytoma has been rarely reported (3). For the present patient, we postulated that risk factors, such as myocardial damage, akinetic left ventricular wall, a hypercoagulable state resulting from catecholamine secretion, hypertriglyceridemia, and hyperglycemia, might have contributed to left ventricular thrombosis (3, 9). Echocardiography is the primary, cost-effective imaging modality in diagnosing an intracardiac thrombus (10). Patients with a left ventricular mural thrombus should be treated with anticoagulation therapy because of the risk of systemic thromboembolism. Mi-tsuma et al (11) suggested that anticoagulation therapy should be performed until cardiac wall motion abnormalities have improved. However, anticoagulant therapy in thromboembolic diseases is inappropriately underutilized in China because of concerns regarding hemorrhagic complications (12). For the present patient, thrombolysis and anticoagulant therapies were unreasonably prescribed (13). As a catastrophic result, the bilateral arteries of the legs were reoccluded and amputation had to be performed.

**References**