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Case Report

Stress-related cardiomyopathy, ventricular dysfunction, artery thrombosis: a hidden pheochromocytoma

Abstract

Clinical presentation of pheochromocytoma can vary, and it can sometimes mimic other diseases. Some patients with pheochromocytoma may have atypical presentations, such as clinical features consistent with an acute coronary syndrome, that only later suggest a classical picture of stress-related cardiomyopathy. To our best knowledge, pheochromocytoma has been incidentally revealed in a few cases of catecholamine-induced cardiomyopathy and in only 1 case of peripheral arterial thrombosis. This is the first case of pheochromocytoma revealed after left ventricular dysfunction caused by stress-related cardiomyopathy associated with inferior limb artery thrombosis in a patient with a complex cardiovascular history.

We report a case of a 63-year-old white woman with catecholamine-induced cardiomyopathy, mimicking acute coronary syndromes (ACSs), left ventricular dysfunction, peripheral arterial thrombosis, and an unrevealed pheochromocytoma incidentally diagnosed.

This case highlights the complexity of managing patients with atypical clinical presentation and multitude of symptoms mimicking other diseases. Pheocromocytoma remains often undiagnosed, and atypical presentation of stress-related cardiomyopathy could suggest a hidden pheochromocytoma, especially in a patient with an extensive cardiovascular history.

A smoker, obese, 63-year-old white woman with a medical history of hypertension, dyslipidemia, and previous ischemic stroke was referred to our emergency department with resting dyspnea and peripheral coldness. The patient presented with precordial pain, which started 24 hours before and lasted 30 minutes. Physical examination revealed a blood pressure of 120/75 mm Hg, heart rate of 100 beats/min, and oxygen saturation of 94%, as well as normal heart sounds and fine crackles over both lung bases. Chest x-ray showed typical findings of an acute pulmonary edema.

Electrocardiogram (ECG) showed ST-segment elevation in V_2 to V_4 and T-wave inversion in V_2 to V_6 precordial leads, suggesting a subacute myocardial infarction (Fig. 1). On admission to intensive care unit, echocardiography showed akinesia of all mid and apical segments with normal basal contraction and systolic function impairment (ejection fraction, 35%), typically consistent with takotsubo cardiomyopathy (TTC). Furthermore, she admitted to have a recent history of moderate psychological distress. Laboratory findings are shown in Table 1A.

A coronary angiography with a right radial approach showed normal coronary arteries (Fig. 2A, B).

Left ventriculography showed an apical akinesia of the anterior wall with enhanced contractility of basal segments (Fig. 2C, D).

Few hours later, the patient complained of right leg pain and paresthesia. The leg appeared cold and pale, with lack of tibial and

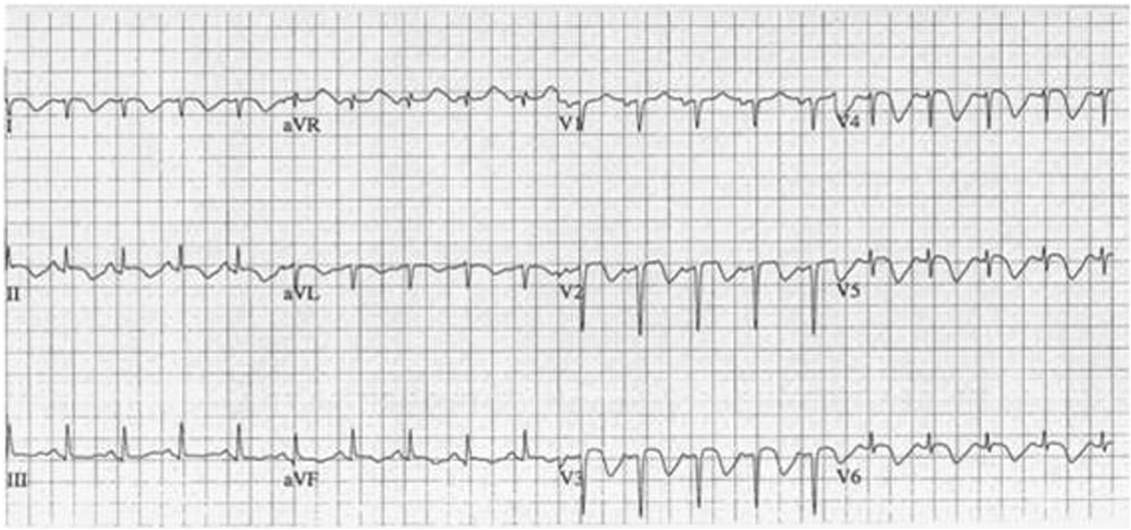

Fig. 1. On admission, 12-lead ECG showed segment elevation in V_2 to V_4 and T-wave inversion in V_2 - V_6 precordial leads.

Table 1
Laboratory values of the patient at hospitalization (A) and after CT (B)

	Values	Reference ranges
A		
Hemoglobine	16.1 g/dL	12-15 g/dL
Red blood cells	6.18 × 10^6/μL	3.8-5.0 × 10^6/μL
Hematocrit	50.20%	35 %-48 %
White blood cells	22.26 × 10^3/μL	4.5-10.00 × 10^3/μL
Platelets	433 × 10^3/μL	150-400 × 10^3/μL
Blood glucose	150 mg/dL	60-110 mg/dL
Creatinine	2 mg/dL	0.7-1.3 mg/dL
Troponin	24.30 ng/mL	0-0.05 ng/mL
Myoglobin	5668 ng/mL	0-116 ng/mL
Creatine kinase–MB	>300 ng/mL	0-3.8 ng/mL
Lactate dehydrogenase	1820 U/L	313-618 U/L
Brain natriuretic peptide	2917 U/L	0-160 U/L
B		
Plasma metanephrine	14.3 nmol/L	< 0.49 nmol/L
Plasma normetanephrine	24.1 nmol/L	< 0.89 nmol/L
Plasma cortisol (at 6 o'clock)	32.7 μg/dL	8 to 23 μg/dL
Urinary cortisol	1373 μg/24 h	50 to 200 μg/24 h

pedal pulses. Then, a computed tomography angiography (angioCT) was performed, revealing an abrupt interruption of posterior right tibial artery and a significant reduction of left posterior tibial artery flow (Fig. 3). Incidentally, a voluminous (7 × 6 × 5 cm) and inhomogeneous left adrenal gland mass was revealed (Fig. 4). These results indicated a percutaneous transluminal angioplasty to restore the perfusion of the right leg (Fig. 5). Right posterior tibial lesion was treated, achieving a suboptimal reperfusion. Moreover, the findings of angioCT led to suspect a pheochromocytoma and imposed more specific laboratory reports, which revealed significantly elevated levels of plasma metanephrine and normetanephrine, plasma cortisol, and urinary cortisol (Table 1B).

Three weeks later, elective surgery was performed.

Histopathologic findings of the removed lesion (Fig. 6) confirmed that the tumor was a pheochromocytoma.

Many patients with clinical features consistent with an ACS after a diagnostic workup revealed other diseases. Atypical presentations often delay correct diagnosis or lead to misdiagnosis. In 1.5% to 2.2% of patients presenting with symptoms that initially appear to be an ACS [1], TTC is revealed.

Our patient presented with many hallmarks of TTC, and until pheochromocytoma was found, the 4 "Mayo Clinic" diagnostic criteria [2] required for the diagnosis of TTC were satisfied (Table 2).

The literature indicates that the pathophysiology of TTC and pheochromocytoma-related cardiomyopathy is similar and mediated by catecholamines [3]. The left ventricular wall motion abnormality in patients with pheochromocytoma crisis is generally global, although apical-sparing and TTC-like wall motion abnormalities have been reported [4,5]. Thus, Bybee and Prasad [2] suggested a modified version of the diagnostic criteria for TTC (Table 3), in which the lack of proven pheochromocytoma does not appear.

This tumor is probably underdiagnosed and is often missed. Incidentally, in our patient, pheochromocytoma was diagnosed with angioCT performed for acute right lower limb ischemia occurred during hospitalization.

Catecholamine-induced cardiomyopathy has been ascertained in some cases of pheochromocytoma, and there are some reports of pheochromocytomas presenting as ACS [6].

In our case, clinical presentation was complex: ST-segment elevation myocardial infarction, stress-related cardiomyopathy, left ventricular dysfunction, and peripheral arterial thrombosis. During hospitalization, the patient showed thrombophilic diathesis, with the presence of right leg arterial thrombosis. The possibility of embolization after diagnostic coronary angiography was excluded because of use of transradial approach for cardiac catheterization. Thrombotic events have been reported rarely in patients with pheochromocytoma, and the exact mechanism of thrombosis is unclear [7].

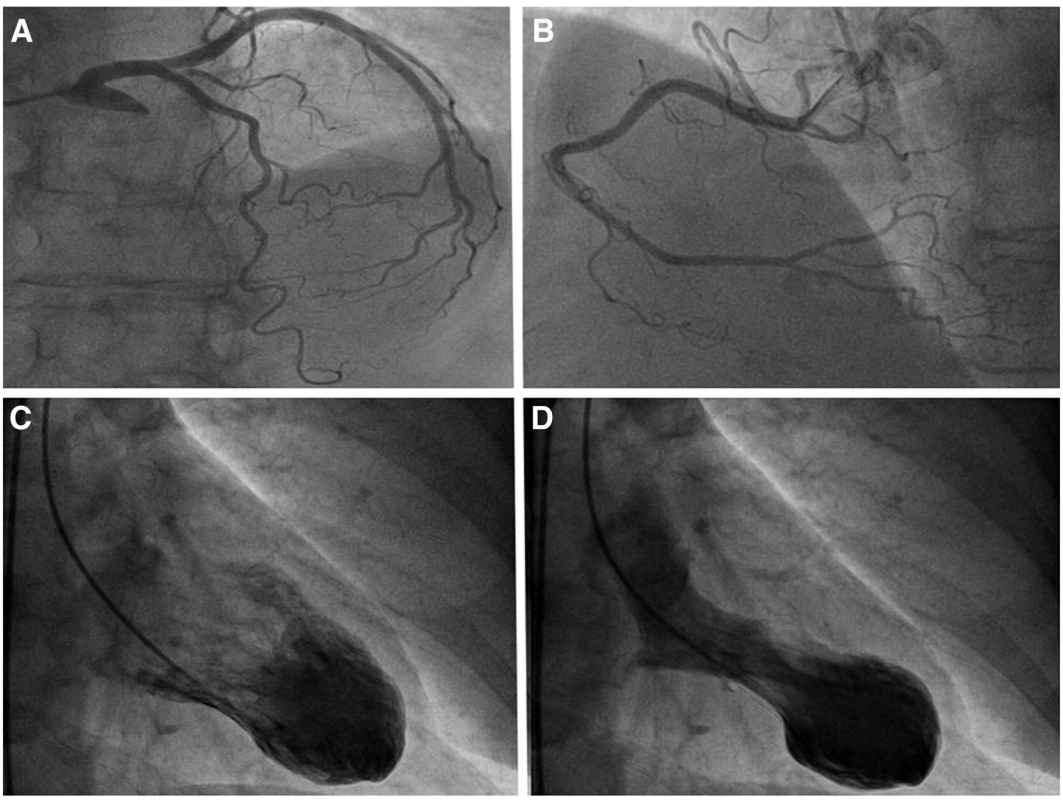

Fig. 2. Diagnostic coronary angiography: left coronary artery with branches to left anterior descending artery and left circumflex artery (normal [A]) and right coronary artery (normal [B]). Left ventriculography (in diastole [C] and in systole [D]) showing systolic dysfunction of the left ventricular apex and midventricle, with hyperkinesis of the basal left ventricular segments.