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A Delayed Diagnosis of Pheochromocytoma Revealed by Tako-Tsubo Cardiomyopathy

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Abstract

Introduction: Pheochromocytoma is a rare cause of secondary arterial hypertension whose clinical presentation can be multifaceted. In particular, it may be revealed or complicated by cardiovascular manifestations independent of hypertension. These include Tako-Tsubo cardiomyopathy, an acute reversible dyskinetic cardiomyopathy associated with ballooning of the left ventricular apex. **Observation:** We present the case of a 32-year-old woman, diabetic for 2 years. Her history included untreated labile hypertension and emotional stress. She was admitted to a cardiac intensive care unit for left heart failure. Paraclinical investigations confirmed the diagnosis of Tako-Tsubo cardiomyopathy induced by pheochromocytoma. After a few days of medical treatment in the ICU, the outcome was favorable, with well-compensated heart failure, stable hemodynamics and restoration of left ventricular function. **Conclusion:** This observation raises the issue of delayed diagnosis of a pheochromocytoma revealed by a TTC. Despite its spectacular initial presentation, the evolution of the latter was rather favorable under appropriate treatment.

Keywords

Pheochromocytoma, Tako-Tsubo, Apical Ballooning

1. Introduction

Pheochromocytoma is a rare endocrine tumor (less than 1 case/100,000 people) that presents with the classic triad of headache, sweating and palpitations [1]. The clinical presentation can be multifaceted, leading to delayed diagnosis. Pheochromocytoma may be revealed or complicated by cardiovascular manifestations

independent of hypertension, such as rhythm disorders, Tako-Tsubo cardiomyopathy (TTC) or acute coronary syndrome (ACS) [2]. TTC is an acute reversible dyskinetic cardiomyopathy associated with ballooning of the left ventricular apex. It is a rare condition (1 case/36,000 adults), but probably underestimated due to a lack of awareness [3]. Despite extensive research, the pathogenesis of TTC remains incompletely understood. The proposed mechanisms of cardiac dysfunction are explained by the toxic effects of catecholamines on the myocardium between the apex and the base of the heart [4]. The initial management of patients with pheochromocytoma-induced TTC should be medical-surgical, including treatment of acute cardiomyopathy and removal of the endocrine tumor. In this report, we present a woman's care pathway reflecting the diagnostic difficulty of pheochromocytoma in its atypical form.

2. Observation

We present the case of a 32-year-old woman, diabetic for 2 years. Her 2-year history included untreated labile hypertension and emotional stress with marital problems and unemployment (informed patient consent obtained).

She was admitted to a cardiac intensive care unit (CICU) for recurrent choking with orthopnea. On admission, body temperature was 38.1˚C, the patient's blood pressure ranged from 95/62 - 165/95 mm Hg, and pulse rate fluctuated between 81 - 110 bpm. Examination revealed a left gallop sound and bilateral pulmonary crackles. Laboratory tests revealed elevated troponin (54 pg/ml) and pro BNP (9635 pg/ml). Blood glucose was 1.59 g/L. Blood catecholamines were at 5 times the upper normal limit: epinephrine 586.98 pg/mL, noradrenaline 921.04 pg/mL and dopamine 150.9 pg/mL. Electrocardiogram showed left ventricular (LV) hypertrophy. Emergency transthoracic echocardiography revealed systolic ballooning of the apical and middle portions of the LV (**Figure 1**).

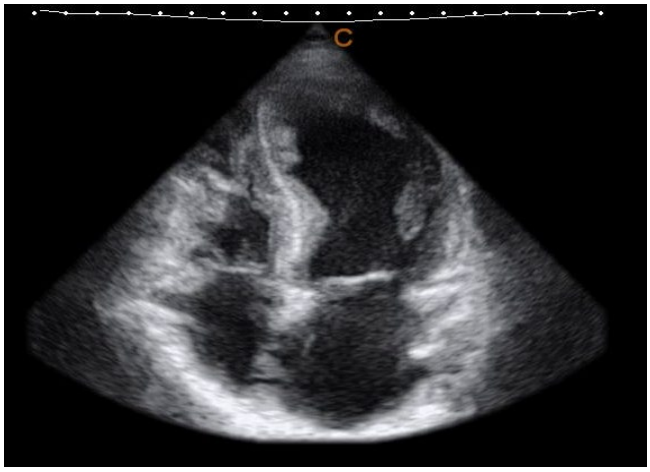

Figure 1. Systolic ballooning of the apical and middle parts of the LV.

Emergency management consisted of oxygen therapy combined with furosemide, spironolactone, ramipril and heparin. After a few days of medical treatment

in the ICU, the outcome was favorable, with well-compensated heart failure, stable hemodynamics and improved left ventricular systolic function. Coronary artery obstruction was excluded by subsequent coronary angiography. Abdominal computed tomography (CT) was performed using a volume acquisition technique without and then with iodinated contrast injection. It showed a homogeneously-enhanced tissue mass in the right adrenal gland measuring 59 × 32 × 97 mm. Paraclinical investigations confirmed the diagnosis of pheochromocytoma-induced TTC. She was discharged on day 14 on ramipril and labetalol. At 4 weeks, left ventricular function was fully restored. After 8 weeks, the patient underwent laparoscopic removal of the adrenal mass without complications. Pathological examination of the surgical specimen confirmed pheochromocytoma. Three months after surgery, the patient was asymptomatic. Repeated measurements of plasma metanephrines and ambulatory blood pressure were within the normal range.

3. Discussion

Our patient had an unusual presentation of pheochromocytoma. Pheochromocytoma classically presents with episodic headaches, palpitations and profuse sweating, accompanied by paroxysmal or permanent hypertension. Serious cardiovascular complications, such as cardiogenic shock, heart failure, cerebral hemorrhage and sudden death, can occur [5]. Our patient presented with cardiogenic acute pulmonary edema. TTC is a transient, reversible cardiomyopathy that occurs most often in postmenopausal women after intense emotional or physical stress. In the presence of new-onset cardiomyopathy and normal coronary arteries, the diagnosis must be made. The difficulty in diagnosing TTC lies in its close proximity to its main differential diagnosis, ACS. The diagnosis of TTC was made in our patient according to the international diagnostic criteria (interTAK criteria) proposed by the ESC 2018 consensus. Our patient was presented with transient left ventricular dysfunction with apical ballooning and an emotional trigger. Cardiac biomarkers (troponin, CPK mb and BNP) were increased. There was no evidence of myocarditis. Abdominal CT showed a right adrenal tumour consistent with a pheochromocytoma. The prevalence of TTC in patients with pheochromocytoma can be as high as 3%. Pheochromocytoma is now included as a risk criterion for TTC [6]. Despite the substantial improvement in our understanding of the pathophysiology of TTC, a number of gaps in our knowledge remain.

Extremely high catecholamine levels have been suggested as an etiological factor underlying ventricular dysfunction. Indeed, catecholamines play an essential role in triggering TTC [7]. Blood concentrations are extremely high and generally remain so for 7 to 9 days. Wittstein *et al.* compared blood levels of catecholamines in 13 TTC patients and 7 subjects hospitalized for acute myocardial infarction on admission to hospital. They found that catecholamine levels were higher in TTC patients [8]. This catecholamine surge leads, via multiple mechanisms (direct catecholamine toxicity, adrenoceptor-mediated injury, epicardial and microvascular coronary vasoconstriction and/or spasm, and increased cardiac workload) to myocardial injury, the functional counterpart of which is transient apical ballooning

of the left ventricle. The relative preponderance in post-menopausal women suggests that estrogen deprivation may play a facilitating role, probably mediated by endothelial dysfunction. The initial management of patients with suspected TTC should be that of ACS. Because of this left ventricular dysfunction, beta-blockers and ACE inhibitors are widely prescribed until contractile function is fully restored. The long-term prescription of beta-blockers to prevent recurrence is controversial [9]. After management of the acute phase, resection of the adrenal tumour should be considered. Alpha-adrenergic receptor blockers are the first choice for preventing perioperative cardiac and cardiovascular disorders [10]. The in-hospital mortality rate reported to date is only around 3% [11]. Recovery of LV systolic dysfunction is generally observed after 4 to 8 weeks [12]. In our patient, LV function was fully restored after 4 weeks.

4. Conclusion

This observation raises the issue of delayed diagnosis of a pheochromocytoma revealed by a TTC. Despite its spectacular initial presentation, the evolution of the latter is rather favorable under appropriate treatment. Initial management requires a meticulous diagnostic approach in order to eliminate differential diagnoses (ACS, myocarditis), establish the diagnosis of TTC and seek the link with pheochromocytoma. With regard to initial treatment, beta-blockers and ACE inhibitors are widely prescribed until full recovery of left ventricular contractile function. Removal of the adrenal tumour is considered after stabilization.

Conflicts of Interest

Authors declare no conflict of interest

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