A Case Report of Biventricular Takotsubo Cardiomyopathy as the Initial Manifestation of a Pheochromocytoma

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Abstract

Pheochromocytomas are an uncommon and contentious medical disorder that has been linked to TTC.1,3,4 Patients with pheochromocytoma-induced TTC provide a therapeutic challenge because to their unique clinical characteristics, varying patterns of ventricular impairment, and worse prognosis. 1, 2, 3 Here we furnish the case of a patient with pheochromocytoma-initiated transient ischemic attack (TTC) who likewise had extreme biventricular impedance on show, as identified via cardiovascular magnetic resonance (CMR).

Keywords: Pheochromocytoma, Cardiac Magnetic Resonance Imaging, Takotsubo Cardiomyopathy, and Stress-Related Cardiomyopathy.

1. Introduction

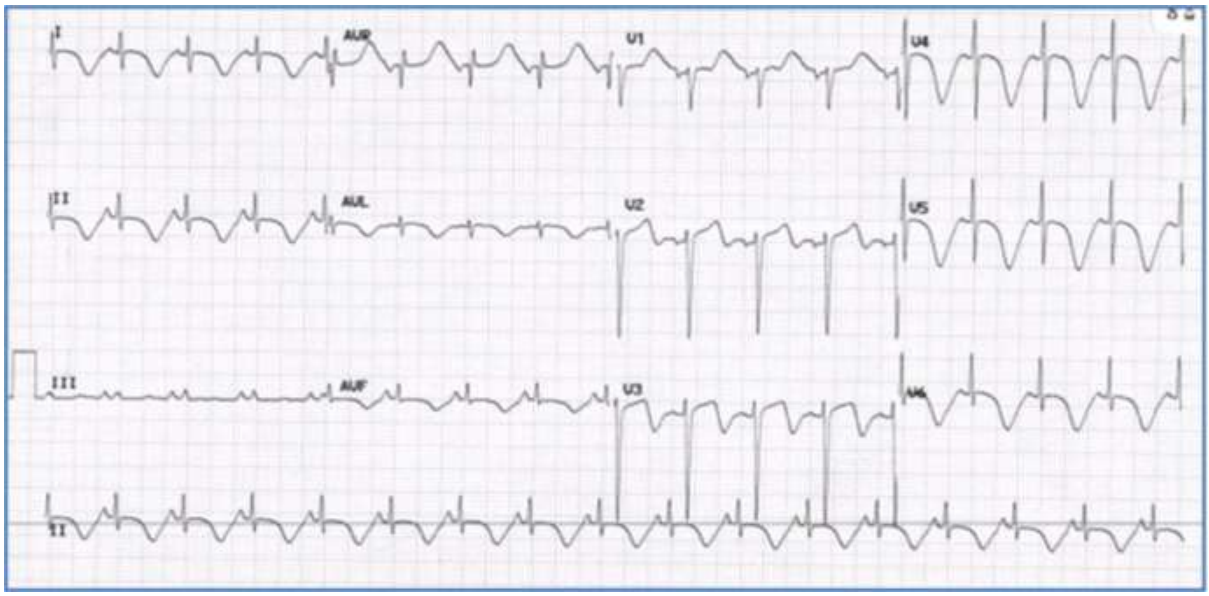
Acute and temporary myocardial damage, stress cardiomyopathy is characterised by localised systolic failure.1 Takotsubo cardiomyopathy (TTC) is characterised by a unique pattern of regional systolic dysfunction and occurs as a result of stress to the heart muscle. It is often brought on by stress, both mental and physical, and it may look a lot like a heart attack.1,2 Left ventricular (LV) dysfunction and regional wall abnormalities that extend beyond the coronary artery supply zone are hallmarks of TTC. This condition is also characterised by an apical and circumferential pattern of myocardial dysfunction that causes the LV to expand at its apex during systole.

2. Case Presentation

A 21-year-old lady who had been in good health before suddenly had acute, oppressive, substernal chest discomfort, along with diaphoresis and shortness of breath, and went to the emergency room. Exam results: oxygen saturation at 97% while breathing at a rate of 15 breaths per minute and a heart rate of 107 beats per minute in room temperature air. The heartbeats were rapid but otherwise normal. No gallops, rubs, or murmurs were heard throughout the cardiac assessment. There were no signs of neurologic dysfunction. There was nothing noteworthy in the patient's medical background. She claimed to have an active

lifestyle and not need any pharmaceutical aid. She admitted to smoking sometimes but said she didn't take drugs. There was no history of heart problems in the family.

Her quick development of chest discomfort, dyspnea, tachycardia, and hypertension was not likely due to coronary atherosclerotic disease because of her young age. Panic disorder, pulmonary embolism, acute coronary syndrome from a dissection or vasospasm of a coronary artery that occurred spontaneously, aortic dissection, and inflammatory disorders like pericarditis and myocarditis were other possibilities. Even though there was no history of drug misuse, an acute intoxication was also taken into account.

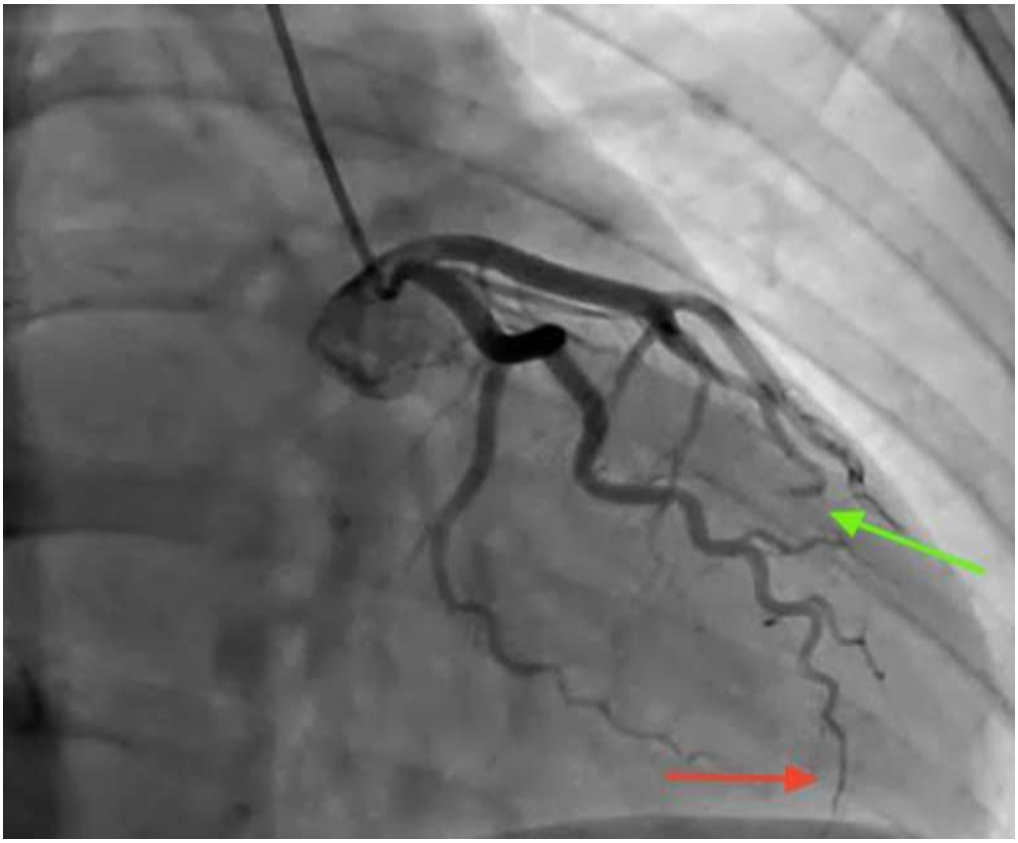


Complete blood count, electrolytes, and renal and hepatic function were all normal on first testing. The results of a toxicology test were negative. In the ECG taken in the hospital, sinus tachycardia and expansive symmetric T-wave reversal were both seen (Figure 1). High-responsiveness troponin I levels of 3.2 ng/mL (ordinary, 0.026 ng/mL) prompted a functioning determination of intense coronary disorder and an exchange to the coronary consideration unit. The severe T-wave inversion elevated the working differential diagnosis to include apical variant hypertrophic cardiomyopathy alongside acute coronary syndrome. Although coronary angiography revealed no stenosis, sluggish flow was found in the left anterior descendent area. Microvascular dysfunction is typified by this result.

There was modest systolic dysfunction, with a 40% left ventricular ejection fraction, as shown on a transthoracic echocardiography performed in the hospital (Video 2). With a fractional area change (FAC) of 32%, a tricuspid annular plane systolic excursion (TAPSE) of 1.5 cm, and a tricuspid annular S velocity of 18 cm/sec, tissue Doppler imaging (TDI; Video 3, Figures 3 and 4) revealed moderate systolic dysfunction and right ventricular apical akinesis.

4). After suspecting TTC, a cardiac magnetic resonance imaging (CMR) study indicated mid- and apical-segment LV akinesia and mid-segment dyskinesia of the right ventricular free wall (Video 4). No late gadolinium enhancement (LGE) was seen in the apical segments, and myocardial edoema was detected using the T2 short tau inversion recovery (STIR) sequence, as shown in Figure 5. Classically, biventricular TTC is diagnosed when both the left and right ventricles show mid and apical akinesis and there is also proof of apical edema without LGE

in the equivalent region. Focal LGE was seen in the basal septum, however it was unrelated to the present event. Given the lack of further evidence, we connected this to a history of myocarditis.



Clinically, the patient had substantial, paroxysmal increases in blood pressure despite intensive medical treatment. We suspected an underlying aetiology due to the existence of safe harmful hypertension and extensive pulse change in a youthful patient who had previously been healthy and free of risk factors. This led to additional investigation since secondary hypertension was suspected. Consequences of a 24-hour pee fractionated metanephrines test showing raised degrees of normetanephrine (141.3 mol/day; reference range, 0.48-2.42 mol/day) and metanephrine (76 mol/day; reference range, 0.26-1.73 mol/day) propose a hyperadrenergic state. Then, CT sweeps of the mid-region uncovered a 55 mm, 40 mm, and 37 mm mass, with heterogeneous improvement, starting in the right adrenal organ (Figure 6). Both the imaging and serologic exploratory results were solid with the finding of a biventricular TTC caused by an adrenergic crisis brought on by a pheochromocytoma. The patient was given immediate medical attention and then moved to another facility for further medical and surgical care.

3. Discussion

Since its first description by Sato and coworkers5 in 1990, Among women who lack traditional cardiovascular risk factors, TTC has become an inexorably perceived reason for short lived ventricular brokenness.The term "octopus trap" was first used to invasive left ventriculography because of the striking similarity between the LV during systole and a Japanese octopus trap.

Postmenopausal women with Takotsubo cardiomyopathy generally report with intense or subacute chest uneasiness in the wake of encountering huge close to home or actual pressure. Some individuals with TTC may have significant cardiovascular compromise, hypotension, cardiogenic shock, and the need for ventricular support, despite the fact that ventricular function and wall-motion anomalies are known to almost completely recover following the acute episode.2

Many different explanations for the pathophysiological processes and precipitating factors of TTC have been presented.2,6 According to the results of the worldwide Takotsubo Registry Study, 71.5% of patients who experienced a cardiovascular episode reported experiencing a profound or actual trigger in the weeks paving the way to it.2 The demise of a friend or family member, maltreatment at home, monetary misfortunes, or a shocking medical prognosis are all examples of major life events that may cause significant emotional distress. On the other side, acute critical illness, surgery, and extreme pain are regularly cited as sources of physical stress.6

Pheochromocytomas, tumours of the adrenal medulla that secrete catecholamines, are very uncommon, occurring in just 0.05% of people.Starting with tyrosine hydroxylase (TH), which controls the conversion of tyrosine to dihydroxyphenylalanine (DOPA), chromaffin cells create catecholamines. Dopa decarboxylase converts di-hydroxyphenylalanine to dopamine, which in turn is metabolised by dopamine -hydroxylase to provide norepinephrine. An enzyme called phenylethanolamine-N-methyltransferase (PNMT) converts norepinephrine to epinephrine. Adrenoreceptors modulate the effects of these catecholamines on the heart after they have been released from storage vesicles and into the bloodstream.[6]

Pheochromocytomas may produce catecholamines in a variety of ways, including intermittently, constantly, and in a hybrid pattern. Epinephrine is released in a paroxysmal manner that causes tachyarrhythmias, whereas norepinephrine is produced constantly and may cause chronic hypertension.[7][8] Both epinephrine and norepinephrine have different binding affinities for alpha (1,2) and beta(1,2) adrenoreceptors. Adrenoceptors may grow in number or have their affinity changed by hormones such glucocorticoids and thyroid hormones.[9][10][11]

Beta-1 adrenoceptors are found in the heart, and when stimulated, they cause adenylate cyclase to become active through the guanosine triphosphate protein-coupled receptor(Gs). By stimulating adenylate cyclase (AC), adenosine triphosphate (ATP) may be converted into cyclic adenosine monophosphate (cAMP). Protein kinase A (PKA) and HCN channels (stimulated by depolarization) are turned on as a consequence. This sequence of events may cause dromotropy in AV nodal cells and ionotropy in SA nodal cells to rise [12].

Calcium efflux into the cytosol is triggered by phosphorylation of ryanodine-2 receptors (RyR2) on the sarcoplasmic reticulum of cardiac myocytes by protein kinase A.[13] Cross-bridge cycling and inotropy are amplified as a result of calcium binding to the troponin-tropomyosin complex, which reveals myosin binding sites on actin.[14]

When stimulated by epinephrine or norepinephrine, beta-2 adrenoceptors in the peripheral blood arteries dilate the vessels. Activation of alpha(1)-adrenoreceptors in vascular smooth muscle cells by norepinephrine and epinephrine may cause vasoconstriction-induced

hypertension. Norepinephrine release is suppressed by alpha-2 adrenoceptors located on synaptic nerve terminals.

The lesion's histology often reveals zellballen nests of chromaffin cells that are highly positive for chromogranin, synaptophysin, CD56, and focally for S100.[15][16] The malignant type is only distinguished by the presence of chromaffin cells in the extra-adrenal tissue.[15]7 Pheochromocytoma is strongly suspected in cases of paroxysmal hypertension with accompanying episodes of headache, palpitations, and sweating. Patients who exhibit malignant hypertension, have difficulty controlling their blood pressure with antihypertensive medication, or whose blood pressure paradoxically rises in response to psychological or pharmaceutical stimulation or tumor manipulation (e.g., abdominal palpation) may benefit from a pheochromocytoma test. Pheochromocytoma must be detected using biochemical and imaging tests for an accurate diagnosis. Screening using plasma-free metanephrines is the gold standard. Further contrast-enhanced imaging of the abdomen and pelvis, such as computed tomography or magnetic resonance imaging, is advised if this test returns positive findings. In most situations, the tumour can be removed surgically, although there are exceptions.

Adrenergic crisis caused by pheochromocytoma has been linked to TTC, however this association has been the subject of much dispute and scepticism. 1,3,8,9 The presence of ventricular wall-movement irregularities with regards to an adrenergic emergency, even within the sight of the normal imaging discoveries of TTC, was recently delegated a different and unmistakable disorder by numerous specialists and recently proposed symptomatic models, precluding pheochromocytoma as a potential reason for TTC.10 However, provided that certain clinical and radiological criteria are satisfied, the current statement positions from the International Expert Consensus on Takotsubo Cardiomyopathy have validated the link between pheochromocytoma and TTC.1

Pheochromocytoma is infrequently reported when Takotsubo cardiomyopathy is the presenting symptom.2,4,10 Recent studies have shown that there are significant variations in demographic, clinical, and imaging characteristics between TTC caused by pheochromocytoma and TTC caused by other stresses. In contrast to patients with TTC caused by other causes, those with pheochromocytoma tend to be younger (mean age 46 vs. 66), more likely to be male (30% vs. 10%), and less likely to have the usual apical ballooning pattern (44% vs. 83%).2, 3, 10. Both global and basal ventricular compromise are more common in this population (20% versus 0% and 2.2%, respectively).They are also more likely to have cardiogenic shock (33% vs. 9.9%), one of several complications that are substantially more common in this cohort than in the whole TTC population (68% vs. 21.8%).The left ventricle (LV) is usually the sole organ affected by TTC,2,10 although biventricular impairment does occur. The right ventricle has been observed to be involved in 25%-42% of people with TTC.Biventricular compromise in pheochromocytoma-induced transient ischemic attack (TTC) is a rare complication, with just a few of instances recorded globally (11, 12).3,10 These patients often exhibit substantially impaired ventricular capability, early clinical decay, and an expanded risk of consequences, calling for a more extensive and aggressive approach to treatment.10 When compared to those patients, our

patient's hospitalisation did not progress towards respiratory or heart failure, pleural effusion, cardiogenic shock, or the need for any sort of advanced life support.

Multimodal imaging, and particularly CMR, was essential in our instance for ruling out other explanations of regional LV systolic failure, providing a thorough study of both ventricles, and bolstering a final diagnosis of TTC as the most probable aetiology. CMR has been considered as a useful technique in the diagnostic workup of unusual situations like this one. Gravina and coworkers hypothesised that this would make it possible to diagnose stress cardiomyopathy and pheochromocytoma with a single imaging test.

4. Conclusion

When atypical traits are present, such as a patient's young age, unique clinical symptoms, or irregular wall-motion, it's important for clinicians to be aware of rare aetiologies of TTC. It's important to be very suspicious from the outset of the investigation. Here, we show a youthful patient with threatening hypertension, imaging discoveries of biventricular brokenness, and the trademark segmental wall-movement irregularities of TTC. A youthful patient without risk factors was followed up on because they had a unique kind of Takotsubo and malignant hypertension at the same time. The existence of a pheochromocytoma was verified by imaging scans, which were consistent with the findings of the laboratory tests that had indicated a hyperadrenergic condition. A biventricular Takotsubo was suspected as the primary manifestation of a pheochromocytoma based on clinical and laboratory evidence, which was rather unusual.

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