Case Report

Focal mid-ventricular anterior ballooning: An unusual pattern of Takotsubo cardiomyopathy

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Summary Takotsubo cardiomyopathy (TC) or left ventricular apical ballooning syndrome is typically characterized by reversible systolic dysfunction of the apical- and mid-segments of the left ventricle. Symptoms are precipitated by intense emotional or physical stress, in the absence of obstructive coronary artery lesions. The most common presentation of TC is a transient left ventricular apical ballooning. However, recent case reports have described various patterns of TC associated with distinct regional left ventricular wall motion abnormalities. One of very rare these variants, referred to as a "mid-ventricular" type, is characterized by akinesis with or without ballooning of the mid-ventricular segment, together with a hyperdynamic base and apex. Using left ventriculography we describe an atypical form of TC with transient, focal mid-ventricular ballooning of the anterior segment, followed by complete resolution of ballooning, as observed by cardiac magnetic resonance (CMR) imaging.

Keywords: Atypical form of takotsubo cardiomyopathy, mid-ventricular anterior ballooning, diagnosis

1. Introduction

Takotsubo cardiomyopathy (TC) was first described by Japanese physicians in 1991 (1). It is generally characterized by transient systolic dysfunction precipitated by a stressful event, in the absence of obstructive coronary artery lesions. For this reason TC is also known as "stress-induced" or "brokenheart" syndrome. Diagnoses of TC are becoming more common, likely due to an increase in the awareness of interactions between critical brain activity and the heart. The most common presentation of TC is an apical ballooning pattern; however, several recent case reports have described variants of TC that present with differing patterns (2-4). These atypical forms are associated with distinct regional left ventricular wall motion abnormalities. Left ventriculography and CMR imaging is uniquely suited for the evaluation of patients with TC and its variants. Using these imaging

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techniques we present a case of atypical TC that fulfills all current diagnostic criteria for TC, excluding the classical contractile LV pattern.

2. Case Report

A 59-year-old female patient with a history of hypertension and dyslipidemia was admitted to the emergency department with retrosternal chest pain after learning of her son's death. Initial physical examination of the patient was normal. Arterial blood pressure was 130/80 mmHg with a heart rate of 80 beats per minute. Her first electrocardiogram (ECG) recording in the emergency room demonstrated normal sinus rhythm with T-wave inversion on leads V1-V4. Laboratory findings indicated a significantly elevated serum level of troponin I and CKMB (troponin I, 2.44 mcg/L and CKMB, 57.1 U/L, normal values < 0.1 mcg/L and < 25 U/L, respectively). Transthoracic echocardiography identified a regional wall motion abnormality. Akinesia was detected in the mid-anterior segment, with a 45% left ventricular ejection fraction. Due to the clinical presentation of symptoms an emergency coronary angiography was performed on the patient. Cardiac

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catheterization and angiography revealed no significant coronary artery disease and coronary artery spasm provocation tests were negative. Left ventriculography demonstrated akinesis of the mid-ventricular anterior segment with ballooning and a hyperdynamic base and apex (Figure 1). The patient was treated with aspirin, statins, beta-blockers and angiotensin-converting enzyme inhibitor and was discharged in stable condition on the third day. Four weeks following discharge CMR imaging was performed on the patient. We found a complete lack of ballooning together with an absence of scaring and no abnormalities with regards to wall motion (Figure 2).

3. Discussion

Takotsubo cardiomyopathy is typically characterized by reversible systolic dysfunction of the apical and mid-segments of the left ventricle, in the absence of obstructive coronary artery lesions. Clinical symptoms of TC mimic those of acute coronary syndrome (ACS), the most common of which are chest pain and dyspnea. The patient may also present with hypotension secondary to decreased stroke volume. Cardiac arrest, syncope and arrhythmias, with or without a prolonged QT interval, have also been described (5,6). Transient heart failure is the most common clinical complication, although the prognosis is generally favorable despite limited published data chronicling long-term patient outcome.

The majority of TC patients present with dynamic ECG changes and precordial ST segment elevation or diffuse T-wave inversions. These changes are typically associated with a modest rise in the level of serum cardiac enzymes. Wall motion abnormalities and functional mitral regurgitation are evaluated in patents using echocardiography, whereas coronary angiography is considered the gold standard to differentiate TC from ACS. The pathophysiology of TC is currently unclear; however, several theories have been proposed recently. A role for catecholamines in triggering TC appears feasible as high levels of circulating catecholamine can cause cardiotoxicity and coronary artery spasms, resulting in myocardial stunning (7,8). Low estrogen levels have also been implicated. It is known that estrogen provides cardioprotection in premenopausal females by downregulating beta-1 adrenoceptors. Accordingly, estrogen supplementation might protect postmenopausal females against the development of TC (8).

Diagnosis criteria for TC, as proposed by the Mayo Clinic, relate primarily to wall motion abnormalities, such as transient hypokinesis, akinesis, or dyskinesis of the LV mid segments, with or without apical involvement. These regional wall motion abnormalities extend beyond a single epicardial vascular distribution (9). Several novel variants of TC have been described



Figure 1. Left ventriculography demonstrated akinesis of the mid-ventricular anterior segment with ballooning and a hyperdynamic base and apex.



Figure 2. After four weeks cardiac magnetic resonance imaging showed complete lack of ballooning together with an absence of scaring and no abnormalities with regards to wall motion.

recently; however, "typical" TC remains the pattern observed most frequently. Atypical forms are associated with distinct regional left ventricular wall motion abnormalities, although reverse or inverted TC, midventricular type and biventricular (right and left ventricular) ballooning group have also been described (2-4). In reverse or inverted TC, the apex is hyperdynamic and the base is akinetic (2). Midventricular type is characterized by akinesis with or without ballooning of the mid-ventricular segment together with a hyperdynamic base and apex (3). Other variants described include akinesis of various left ventricle and right ventricle segments (4). In this case report we describe an atypical form of TC with transient, focal mid-ventricular anterior-segment ballooning.

Our report raises the intriguing question of why apical segments are predisposed to stunning, compared to the base, in patients with TC. Several potential explanations can be considered. First, the apex may be more responsive to adrenergic stimulation or demonstrate an increased density of catecholaminesensitive receptors. Second, the apex is structurally vulnerable. It possesses a relatively limited elasticity reserve and can easily become ischemic as a consequence of limited coronary vasculature (10). Interestingly, a recent case report identified different variants of typical and atypical forms in the same patient (11). Variation between individuals with regard to the local release of catecholamines or in the distribution of autonomic innervation of the LV wall may therefore contribute to the observation of different variants.

Coronary angiography is considered the gold standard for the diagnosis of TC. Left ventriculography is used to identify abnormalities in wall motion and is therefore key in differentiating the variants. CMR imaging is uniquely suited for the evaluation of patients with TC. Critically, it provides markers to identify reversible versus irreversible injury, which are important criteria when confirming a diagnosis of TC and excluding similar acute cardiac diseases, superficially ACS or myocarditis (12). In our case CMR imaging was utilized to demonstrate the reversibility of systolic LV dysfunction after the acute phase.

4. Conclusion

Several novel variants of TC have been described in recent case reports. Our case illustrates an unusual presentation of takotsubo cardiomyopathy. Coronary angiography and left ventriculography are considered the gold standard for the diagnosis of TC and its differentiation from ACS. Together with CMR imaging these methods are also effective at distinguishing between typical and atypical forms of TC. The pathophysiological mechanisms driving distinct LV wall motion abnormalities in different variants of TC remain unclear. Therefore, additional studies are warranted to investigate the role of genetic susceptibility and patient predisposition to physical and emotional stress in development of this disease.

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