The "broken heart sydrome": you're likely to have it only once!

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ABSTRACT

Stress cardiomyopathy (SC) is also known as Takotsubo cardiomyopathy. The term transient left ventricular ballooning syndrome has also been used. These terms refer to the same phenomenon. The typical presentation of SC in a patient is a sudden onset of chest pain or heart failure associated with electrocardiographic changes of ST-segment elevation, which is suggestive of anterior ST-segment elevation myocardial infarction. When these patients are assessed, a bulging of the left ventricular apex with a hypercontractile base of the left ventricle is noted. It is this hallmark bulging of the apex with preserved function at the base that characterises this unique syndrome. The possible pathophysiology of its mechanism as well as the different morphological types are discussed.

Keywords: broken heart syndrome, coronary arteries, ST-elevation myocardial infarction (STEMI), stress cardiomyopathy, Takotsubo cardiomyopathy

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INTRODUCTION

Stress cardiomyopathy (SC) is also known as Takotsubo cardiomyopathy, but many still prefer to call it the 'broken heart syndrome'. The typical presentation of a patient with SC is a sudden onset of chest pain or heart failure associated with electrocardiographic (ECG) changes of ST-segment elevation, which is suggestive of anterior ST-segment elevation myocardial infarction (STEMI). A bulging of the left ventricular apex with a hypercontractile base of the left ventricle is often observed when these patients are assessed. It is this hallmark bulging of the apex with a preserved function at the base that has earned the syndrome its Japanese name 'tako tsubo', or octopus traps.[1] Some have termed this phenomenon as the transient left ventricle apical ballooning syndrome.

The syndrome is frequently observed in postmenopausal women, and is often precipitated by severe and intense emotional (grief, fear, extreme anger or even intense surprise) or physical stress. It has been reported worldwide and is not confined to any particular country or region. The pathophysiology of SC is not fully understood, but there are several postulations:[1-9] (1) Dysfunction of the microvasculature: this theory has the widest acceptance to date. It states that the dysfunction of the coronary arteries is most likely to occur at the level at which they are no longer visible on coronary angiography. As a result, the microvascular arteries fail to provide adequate oxygen and blood supply to the myocardium. While many SC patients have risk factors for coronary artery disease, the pathogenesis is thought to be dissimilar to that of microvasculopathy of diabetes mellitus;[1,9,10] (2) Transient vasospasm: this theory suggests that there are multiple simultaneous spasms of the coronary arteries that cause a reduced blood supply that is significant enough to cause myocardial stunning. This appears to occur less frequently than previously thought; (3) The 'encircling' left anterior descending (LAD) artery: the anterior wall of the left ventricle is supplied by the LAD artery in most people. However, if this artery also goes around the apex of the heart, it will provide the blood supply to the apex and the inferior wall of the heart. This theory, however, is not able to explain the aetiology of one of the documented variants of SC, where the mid-ventricular wall or the base of the heart does not contract; and (4) Effect of adrenaline: it is postulated that stressful situations increase adrenaline levels in blood. The exact mechanism as to how the adrenaline affects the heart in SC is not clear. It may cause a narrowing of the coronary arteries, thus reducing the blood flow. Alternatively, it may bind directly to the heart muscles, causing large amounts of calcium to enter the cells, rendering them temporarily dysfunctional.

With any of these postulations, the effects are known to be transient and completely reversible. As the presentation involves significant heart muscle weakness, patients can present with heart failure, hypotension, shock and arrhythmias. They do, however, improve quite quickly, but there have been reported cases where recovery of the left ventricular function may take up to two months.[1,8,9]

CASE REPORT

A 55-year-old postmenopausal Chinese woman

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presented at the Emergency Department (ED) with severe, compressive, central chest pain, which radiated to her left arm and left upper back. There were associated cold sweats and giddiness, as well as shortness of breath. The patient had felt faint before her arrival at the ED. All these symptoms appeared within ten minutes after she had a heated argument with her family members. The patient had a history of hyperlipidaemia and was compliant with her medication.

Physical examination did not show any evidence of an irregular pulse, heart failure or other abnormalities. The patient was not pale and her vital signs were stable. Her chest pain score was five out of a maximum score of ten. Immediate 12-lead ECG showed ST-segment elevation in leads V2–V6. The patient was managed using the standard STEMI acute coronary syndrome protocol. Blood tests were conducted and sent STAT to the laboratory. The full blood count, urea and electrolytes, glucose level and coagulation profile were all normal. The troponin T level was elevated at 1.16 (normal: < 0.01) µm/l. The patient's chest radiograph was normal and did not show any evidence of heart failure or cardiomegaly.

Aspirin and clopidogrel were administered, and nitroglycerine infusion was commenced for the chest pain. The cardiac catheterisation laboratory and interventional staff were activated immediately, while informed consent was being obtained for balloon angioplasty.

The patient's door-to-ECG and door-to-balloon times were 4 minutes and 23 minutes, respectively. Her coronary angiography showed normal coronary arteries, but a pathognomonic wall motion abnormality was noted. Although the base of the left ventricle was contracting normally, the apical portion had 'ballooned' out and was akinetic. Significant wall motion abnormality without any significant coronary plaque or obstruction was noted. The patient's ejection fraction (EF) was 48% at that time.

The procedure was completed and the patient was sent to the intensive care unit for monitoring. She recovered well and her troponin levels decreased gradually. She was discharged three days post operation. Repeat echocardiography after three weeks showed the EF to be 56%.

DISCUSSION

Our patient is a good illustration of the SC syndrome. She presented as a classical case of acute myocardial infarction, precipitated by an emotionally stressful situation and with classical ST-elevation on her 12-lead ECG. The patient was also postmenopausal, among whom the described phenomenon appears to be more common. The initial treatment rendered in the ED was similar to that in any case with ST-elevation acute coronary syndrome. The diagnosis was made when the patient underwent angiographic studies, which also confirmed an absence of obstructive coronary artery disease. Mortality from this condition is low, and no cases of recurrent attacks have been reported in the literature.[7,9]

In a systematic review by Pilgrim and Wyss, the transient left ventricular apical ballooning syndrome was observed in 0.7%–2.5% of patients with suspected acute coronary syndrome. It affected women in 90.7% of the cases (95% confidence interval [CI] 88.2%–93.2%), with a mean age of 62–76 years. The most common presentations included chest pain (83.4%, 95% CI 80%–86.7%) and dyspnoea (20.4%, 95% CI 16.3%–24.5%). The ECG on admission showed ST-elevation in 71.1% (95% CI 67.2%–75.1%) of cases and was usually accompanied by mild elevation of the troponin levels in 85% (95% CI 80.8%–89.1%). The left ventricular function and ejection fraction improved from 20%–49.9% to 59%–76% within a mean time of 7–37 days.[9]

In a study on 107 SC patients by Singh et al, it was observed that different regions of the myocardium were affected by the ballooning process: postero-basal 1%, basal and mid-ventricular 1%, diaphragmatic 2%, localised apical 2%, anterolateral 11%, complete mid-ventricular 29% and the classical apical variant (called Takotsubo) made up 54%. The localised and complete mid-ventricular variants had a younger median age at presentation (64 vs. 71 years, $p = 0.008$) and a higher median left ventricular ejection fraction (45% vs. 35%, $p = 0.006$) compared to the classical variant, with a similar baseline exposure to stressors, risk factors and in-hospital complications.[10]

Emotional stress often precipitates SC. At presentation, patients with SC tend to have higher plasma cathecholamines levels than those with Killip Class III acute myocardial infarction (AMI) (median levels 1,264 pg/ml vs. 376 pg/ml, $p < 0.005$) in a series by Wittenstein et al.[11] This information, however, has been derived from a small observational case series.

Focal myocytolysis is believed to be the origin of SC. No microbiological agents have been found to be associated with it. The pathological changes in the myocardium have not been demonstrated in 'stunned myocardium', and the infiltration of small mononuclear cells has been documented. This makes SC likely to be a type of inflammatory heart disease rather than a coronary artery disease.[6,8]

In conclusion, patients with SC have a presentation similar to those with an AMI, including chest pain, shortness of breath, congestive heart failure and hypotension. As SC involves heart muscle weakness, it can cause complications such as hypotension, heart failure, shock and even potentially life-threatening arrhythmias. However, these are infrequent, and patients do recover quickly when SC occurs. Theoretically, it appears that this syndrome can recur, but to date, there have been no reports of patient having had the attack twice. Patients often make a full and complete recovery. The long term prognosis appears to be excellent.

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