

KCH Lau 劉仲恒
 KKH Yiu 姚啟恒
 KL Lee 李麗芬
 RLY Ko 高立忻
 YM Lam 林銳明
 L Lam 藍連德
 SWL Lee 李偉聯

A case of takotsubo cardiomyopathy: transient left ventricular apical ballooning

一宗章魚壺心肌病病例：短暫左心室頂端脹大

A 78-year-old woman was admitted to hospital with central chest pain and the electrocardiographic and cardiac marker changes typical of acute anterior myocardial infarction. Coronary angiography revealed normal epicardial coronary arteries, and left ventriculography showed apical akinesis as well as basal hyperkinesis. This is a case of transient left ventricular apical ballooning or takotsubo cardiomyopathy, possibly attributable to catecholamine-mediated myocardial stunning.

一名78歲女性胸膈中央痛楚入院。心電圖和心臟病指標顯示為典型的急性前心肌梗塞。冠狀動脈造影顯示心外膜冠狀動脈正常，而左心室造影則發現左心室頂部失去運動能力，下部則運動過度。這是一宗短暫左心室頂端脹大的病例，又稱章魚壺心肌病，可能因為兒茶酚引致的心肌暈損導致。

Case report

A 78-year-old woman presented to the emergency department in August 2005 with a 1-day history of central chest pain that radiated to the jaw and left upper limb. She had visited her family doctor, and her electrocardiogram (ECG) demonstrated ST elevation in V2 to V5. The patient was referred to the emergency department. A close friend had died suddenly 3 days prior to onset of her symptoms. The patient had a history of hypertension, hyperlipidaemia and gout, all controlled by medication prescribed by her family doctor. She also had a 5-year history of intermittent non-exertional chest discomfort associated with dyspepsia. A previous upper endoscopy revealed no abnormality and there was no family history of heart disease or sudden death.

On admission, the patient's pulse was 62/min and her blood pressure 142/103 mm Hg. The cardiovascular and respiratory examinations were unremarkable. Routine laboratory tests revealed a normal blood count, but a troponin I level of 9.26 ng/mL (normal range, 0-0.4 ng/mL). Electrocardiography revealed a normal sinus rhythm with a 2 mm ST elevation seen over V2 to V5 (Fig 1a). Echocardiography revealed anterior wall and apical hypokinesia, left ventricular hypertrophy, and an ejection fraction of 40%.

In view of the delayed presentation and the ST elevation evidence of acute anterior myocardial infarction, she was prescribed standard treatment and no thrombolytic therapy. Three days later she developed atrial fibrillation and re-elevation of the ST segment over the precordial leads (Fig 1b). An urgent coronary angiogram performed for suspected re-infarction showed normal coronary arteries. Left ventriculography showed apical ballooning and basal hyperkinesis (Figs 2a and 2b). A diagnosis of 'transient left ventricular apical ballooning' or 'takotsubo cardiomyopathy' was made. On discharge she remained asymptomatic.

Serial ECGs and echocardiograms performed after discharge showed that the ECG and left ventricular contraction gradually returned to normal over a 12-week period. The ejection fraction was 55%.

Key words:

Cardiomyopathies;
 Catecholamines;
 Myocardial infarction;
 Ventricular dysfunction, left

關鍵詞：

心肌病；
 兒茶酚；
 心肌梗塞；
 心室機能失調，左面

Hong Kong Med J 2006;12:388-90

Department of Medicine, Queen Mary Hospital, Pokfulam Road, Hong Kong

KCH Lau, MB, BS
 KKH Yiu, MB, BS, MRCP
 KL Lee, MB, BS, FRCP
 RLY Ko, MB, BS, MRCP
 YM Lam, MB, BS, MRCP
 L Lam, MB, BS, FRCP
 SWL Lee, MB, BS, FRCP

Correspondence to: Dr KL Lee
 (e-mail: kllee@hku.uk)

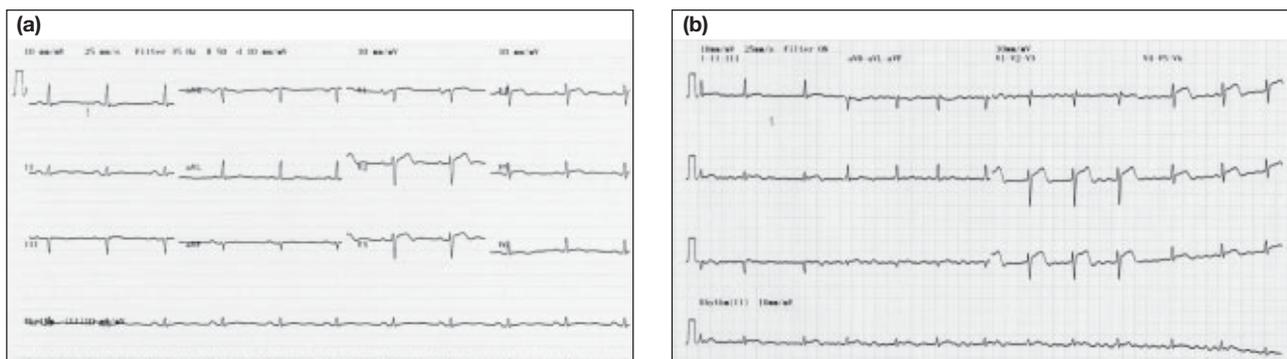


Fig 1. (a) Electrocardiogram (ECG) on admission at 24 hours after the onset of chest pain showing ST elevation over V2 to V5. (b) ECG on day 3 of hospitalisation showing increased ST elevation over V2 to V6

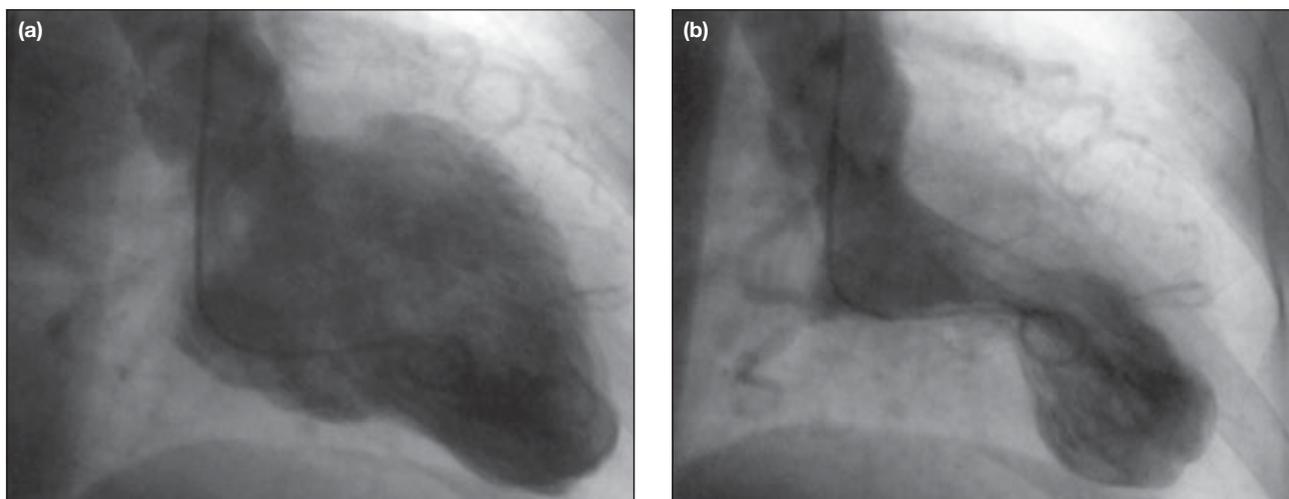


Fig 2. Left ventriculography in right anterior oblique projection in (a) diastole and (b) systole. There was marked apical akinesia but basal hyperkinesia of the left ventricle giving the typical appearance of apical ballooning

Discussion

A number of papers have reported cases of 'transient left ventricular apical ballooning' or 'takotsubo cardiomyopathy', so named because of its resemblance to a takotsubo: a fishing pot with a narrow neck and wide base used to trap octopus. The clinical presentation is similar to that of acute myocardial infarction in the absence of any obstructive epicardial coronary artery lesion. This type of left ventricular dysfunction is characterised by preserved basal function, moderate-to-severe dysfunction in the mid-ventricle, and apical akinesia.

People with takotsubo cardiomyopathy are typically elderly women over 60 years of age who have experienced emotional stress shortly before admission (usually the news of an unexpected death). Wittstein et al¹ reported that the median age of patients with takotsubo cardiomyopathy is 63 (range, 27-87) years, and they present to the emergency department within a median of 2 (range, 1-12) hours of symptom onset. The condition has also been associated with myocardial stunning,² subarachnoid

haemorrhage,³ pheochromocytoma,⁴ Guillain-Barré syndrome,⁵ and emotional stress.⁶ It has also been referred to as 'broken heart syndrome'.

Patients with takotsubo cardiomyopathy typically present with chest pain, pulmonary oedema, and cardiogenic shock. In the initial stages, electrocardiographic changes include a prolonged PR interval, prolonged QT interval, ST elevation mimicking acute myocardial infarction, T wave inversion, and pathological Q waves. In the 48 hours following symptom onset, most patients develop a prolonged QT interval that subsequently resolves.¹

Echocardiography typically shows a contractile pattern of apical ballooning with a reduced left ventricular ejection fraction (median value of 20%). The apical ballooning is characterised by preserved basal function, moderate-to-severe dysfunction in the mid-ventricles, and apical akinesia or dyskinesia. By the fourth day of presentation the left ventricular ejection fraction has usually improved (median value of 45%) with only mild hypokinesia of the left ventricle.¹ Magnetic resonance imaging confirms the

echocardiography findings, with no contrast enhancement; a finding consistent with the absence of myocardial necrosis.¹

The underlying mechanism of transient left ventricular apical ballooning or takotsubo cardiomyopathy remains unknown. One possibility is ischaemia as a result of epicardial coronary artery spasm. Angiographic study has revealed that 70% of affected patients have coronary spasm and electrocardiographic ST elevation at presentation.⁷ Another possible mechanism is microvascular spasm; abnormal coronary flow is possible in the absence of obstructive disease in patients with stress-related myocardial dysfunction.⁸ A third possible explanation is cardiotoxicity, caused by high levels of circulating catecholamines resulting in myocyte dysfunction.⁹ The apical myocardium is particularly sensitive to sympathetic stimulation, and this would explain the characteristic contractile pattern of changes seen on echocardiography and angiography.¹⁰

Treatment of takotsubo cardiomyopathy remains largely empirical with standard supportive care for congestive heart failure including diuretics and vasodilators.

In this patient, a coronary angiogram showed no evidence of coronary artery spasm. It is likely, however, that her pathology was catecholamine-induced cardiomyopathy, as her symptom onset was preceded by an emotional stressor. It is known that ST segment elevation is associated with a high concentration of plasma noradrenaline. Interestingly in this case, the emotional stressor occurred 3 days prior to the onset of chest pain, much longer than the median time previously reported for symptom onset.¹ In our patient, initial echocardiography also showed an ejection fraction of 40%, much higher than the median of 20% reported in a previous series (interquartile range from 15 to 30%).¹ In addition, in that series, the left ventricular ejection fraction measured on echocardiography returned to normal after a median 3-week follow-up. In our patient, the ejection fraction took 12 weeks to return to

normal.

It is now known that transient left ventricular apical ballooning is probably underdiagnosed and has diverse aetiologies and variable presentations. As primary coronary intervention is used more frequently in people with acute ST elevation myocardial infarction, more such patients will be diagnosed. We recommend carrying out a local epidemiological study of takotsubo cardiomyopathy to determine its natural history and pathogenesis.

References

1. Wittstein IS, Thiemann DR, Lima JA, et al. Neurohumoral features of myocardial stunning due to sudden emotional stress. *N Engl J Med* 2005;352:539-48.
2. Braunwald E, Kloner RA. The stunned myocardium: prolonged, postischemic ventricular dysfunction. *Circulation* 1982;66:1146-9.
3. Pollick C, Cujec B, Parker S, Tator C. Left ventricular wall motion abnormalities in subarachnoid hemorrhage: an echocardiographic study. *J Am Coll Cardiol* 1988;12:600-5.
4. Yamanaka O, Fujiwara F, Nakamura T, et al. "Myocardial stunning"-like phenomenon during a crisis of pheochromocytoma. *Jpn Circ J* 1994;58:737-42.
5. Iga K, Himura Y, Izumi C, et al. Reversible left ventricular dysfunction associated with Guillain-Barré syndrome—an expression of catecholamine cardiotoxicity? *Jpn Circ J* 1995;59:236-40.
6. Case records of the Massachusetts General Hospital. Weekly clinicopathological exercises. Case 18-1986. A 44-year-old woman with substernal pain and pulmonary edema after severe emotional stress. *N Engl J Med* 1986;314:1240-7.
7. Kurisu S, Sato H, Kawagoe T, et al. Tako-tsubo-like left ventricular dysfunction with ST-segment elevation: a novel cardiac syndrome mimicking acute myocardial infarction. *Am Heart J* 2002;143:448-55.
8. Bybee KA, Prasad A, Barsness GW, et al. Clinical characteristics and thrombolysis in myocardial infarction frame counts in women with transient left ventricular apical ballooning syndrome. *Am J Cardiol* 2004;94:343-6.
9. Mann DL, Kent RL, Parsons B, Cooper G. Adrenergic effects on the biology of the adult mammalian cardiocyte. *Circulation* 1992;85:790-804.
10. Mori H, Ishikawa S, Kojima S, et al. Increased responsiveness of left ventricular apical myocardium to adrenergic stimuli. *Cardiovasc Res* 1993;27:192-8.