Rare disease

One heart, two cardiomyopathies

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Summary

A 63-year-old woman with no previous medical problems presented with acute chest pain and an ECG consistent with an acute anterior myocardial infarction. At emergency angiography, she was found to have smooth unobstructed coronary arteries. On invasive left ventriculography, overall poor systolic function was noted with apical hypokinesis and basal hyperkinesis, consistent with Tako-tsubo phenomenon. Echocardiography demonstrated a hypertrophic left ventricle and left ventricular outflow obstruction due to systolic anterior motion of the mitral valve and moderate mitral regurgitation. Following appropriate management, she was discharged 6 days later. An outpatient MRI confirmed normalisation of the left ventricular systolic function; however, there was still significant left ventricular hypertrophy and dynamic obstruction. Although most patients presenting with chest pain and an ECG with ST elevation will have an acute coronary event, our patient had normal coronaries but both Tako-tsubo and hypertrophic cardiomyopathies.

BACKGROUND

Tako-tsubo cardiomyopathy is an increasingly recognised diagnosis in patients with suspected ST elevation myocardial infarction. However, the combination of Tako-tsubo cardiomyopathy and previously undiagnosed hypertrophic obstructive cardiomyopathy has only been reported once before¹ and is therefore of interest. We feel that patients presenting with an ST elevation myocardial infarction, who are subsequently shown to have unobstructed coronaries, should be investigated for both Tako-tsubo and hypertrophic cardiomyopathies.

CASE PRESENTATION

A 63-year-old woman with no previous medical conditions presented acutely with cardiac chest pain. An ECG (figure 1) in the community confirmed an ST elevation myocardial infarction (STEMI) and she was transferred to our institution for emergency coronary angiography through the primary percutaneous coronary intervention pathway. Clinical examination revealed an ejection systolic murmur at the left parasternal edge and we proceeded to perform emergency coronary angiography to delineate coronary anatomy.

Figure 1 ECG of our patient at presentation showing anterior ST elevation.

Figure 2 Left ventriculogram image demonstrating basal hyperkinesis with apical akinesis consistent with Tako-tsubo cardiomyopathy and moderate mitral regurgitation.

INVESTIGATIONS

Coronary angiography demonstrated unobstructed coronaries. Tako-tsubo cardiomyopathy was suspected and confirmed on left ventriculography (figure 2). A 45 mm Hg gradient was noted on pull-back across the aortic valve and bedside transthoracic echocardiography confirmed left ventricular outflow tract (LVOT) obstruction due to systolic anterior motion of the mitral valve and moderate mitral regurgitation.

TREATMENT

The patient was in pulmonary oedema and incipient cardiogenic shock, which responded to treatment with a combination of intravenous diuretics and intravenous esmolol (to reduce LVOT obstruction). After 6 days she had persistent left ventricular dysfunction but was well enough to be discharged on oral administration of aspirin, bisoprolol, ramipril and diuretics.

OUTCOME AND FOLLOW-UP

At 3 months, the patient's ECG changes resolved and an MRI scan showed that her left ventricular function had normalised, which is characteristic of Tako-tsubo cardiomyopathy; however, there was still evidence of significant left ventricular hypertrophy and dynamic obstruction following low-dose dobutamine, consistent with hypertrophic cardiomyopathy (figure 3).

DISCUSSION

Although transient LVOT obstruction due to systolic anterior motion of the mitral valve in the context of Tako-tsubo is common affecting up to 10% of all cases, this type of LVOT obstruction is transient and disappears as the left ventricular function normalises.² However, coexistent *persistent* hypertrophic cardiomyopathy with obstruction in a patient presenting with Tako-tsubo cardiomyopathy is a much rarer phenomenon. To date, only one other case has been published in the literature of a patient admitted with a non-STEMI and shown to have both Tako-tsubo cardiomyopathy and a previously undiagnosed hypertrophic cardiomyopathy with obstruction.¹ Ours is the first case to report a patient presenting with a STEMI and shown to have unobstructed coronaries, Tako-tsubo

Figure 3 MRI (left rest, right dobutamine) coronal section during dobutamine testing. Left ventricular hypertrophy is demonstrated on the left and dynamic left ventricular outflow tract obstruction, following low-dose dobutamine infusion, consistent with hypertrophic cardiomyopathy is shown on the right.

phenomenon and persistent hypertrophic cardiomyopathy with obstruction. At present it is not clear whether these represent two separate and rare cardiomyopathies in one patient or a new linked entity.

The aetiology of Tako-tsubo still remains poorly understood. It is thought that possible pathophysiological mechanisms could include impaired coronary microcirculation, multivessel coronary spasm and catecholamine toxicity.³ A stressful emotional or physical trigger usually precedes Tako-tsubo cardiomyopathy, although it can be absent in as many as 20% of the cases. However, both in our case and in the case published by Brabham *et al*¹ a trigger factor was not evident supporting the notion that the previously undiagnosed hypertrophic cardiomyopathy contributes to Tako-tsubo cardiomyopathy. A potential mechanism could include a catecholamine surge exacerbating the resting LVOT gradient owing to a higher density of sympathetic innervation, resulting in a marked increase in intraventricular pressure, increased apical wall stress, reduced subendocardial blood flow leading to apical ballooning.⁴

We feel therefore that all patients with presumed STEMI but unobstructed coronaries should undergo further assessment for evaluation of the presence of both hypertrophic and Tako-tsubo cardiomyopathies.

Learning points

- ▶ Not every patient with acute chest pain and ST elevation on an ECG will have an acute coronary event.
- ▶ Tako-tsubo cardiomyopathy and hypertrophic cardiomyopathy should be considered in all patients with cardiac chest pain and abnormal ECG findings if coronary circulation is unobstructed.
- ▶ A new cardiac entity of patients with previously undiagnosed hypertrophic cardiomyopathy with obstruction and Tako-tsubo cardiomyopathy presenting as STEMI or non-STEMI can potentially be identified in the future.

Competing interests None.

Patient consent Obtained.

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