

CASE REPORT

Syncope in a hypertrophic heart at a wedding party: can happiness break a thick heart? Takotsubo cardiomyopathy complicated with left ventricular outflow tract obstruction in a hypertrophic heart

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Abstract

A 70-year-old woman with known history of hypertension presented because of a syncopal episode during dinner at a wedding party, followed by chest pain. On physical examination a systolic murmur was noted, and her electrocardiogram showed ST segment elevation in anterior leads. She had elevated troponin levels while echocardiography showed a hypertrophic interventricular septum with dyskinetic apex and left ventricular outflow (LVOT) obstruction. Emergency coronary angiography excluded obstructive coronary artery disease and confirmed the presence of LVOT obstruction with a gradient of 90 mm Hg. A left ventriculography showed hypercontractility of the basal and mid segments with apical wall dyskinesia indicating Takotsubo cardiomyopathy. Patient was discharged after 6 days of hospitalization with normalization of left ventricular function and regression of the LVOT obstruction. This is an interesting case of Takotsubo cardiomyopathy complicated with severe LVOT obstruction in a patient with hypertensive heart disease and a sigmoid septum hypertrophy.

INTRODUCTION

Takotsubo cardiomyopathy (TCM) also called apical ballooning syndrome, broken heart syndrome and stress-induced cardiomyopathy is a syndrome characterized by transient regional systolic dysfunction of the left ventricle (LV), mimicking myocardial infarction, but in the absence of angiographic evidence of obstructive coronary artery disease or acute plaque rupture [1]. Some patients with TCM develop cardiogenic shock because of severe systolic dysfunction or left ventricular outflow tract (LVOT) obstruction. However, pre-existing dynamic LVOT obstruction is sometimes complicated with TCM [2] and their coexistence at presentation can create a diagnostic dilemma.

We present an interesting case of TCM complicated with severe LVOT obstruction in a patient with hypertensive heart disease and a sigmoid septum hypertrophy.

CASE REPORT

A 70-year-old female with a known history of hypertension was brought to the emergency department after an episode of sudden loss of consciousness followed by atypical chest pain, during dinner at a wedding party. Preceding the dinner, she recalls drinking 2–3 glasses of wine and enjoying plenty of dancing. Physical examination revealed a systolic

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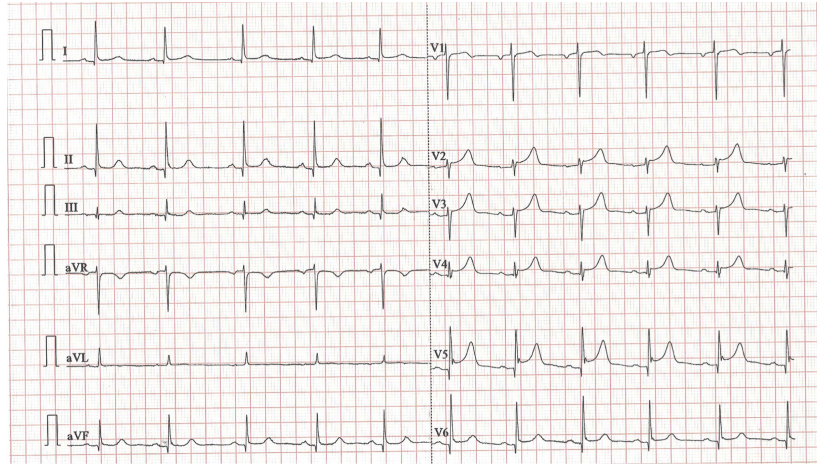


Figure 1: ECG at presentation.

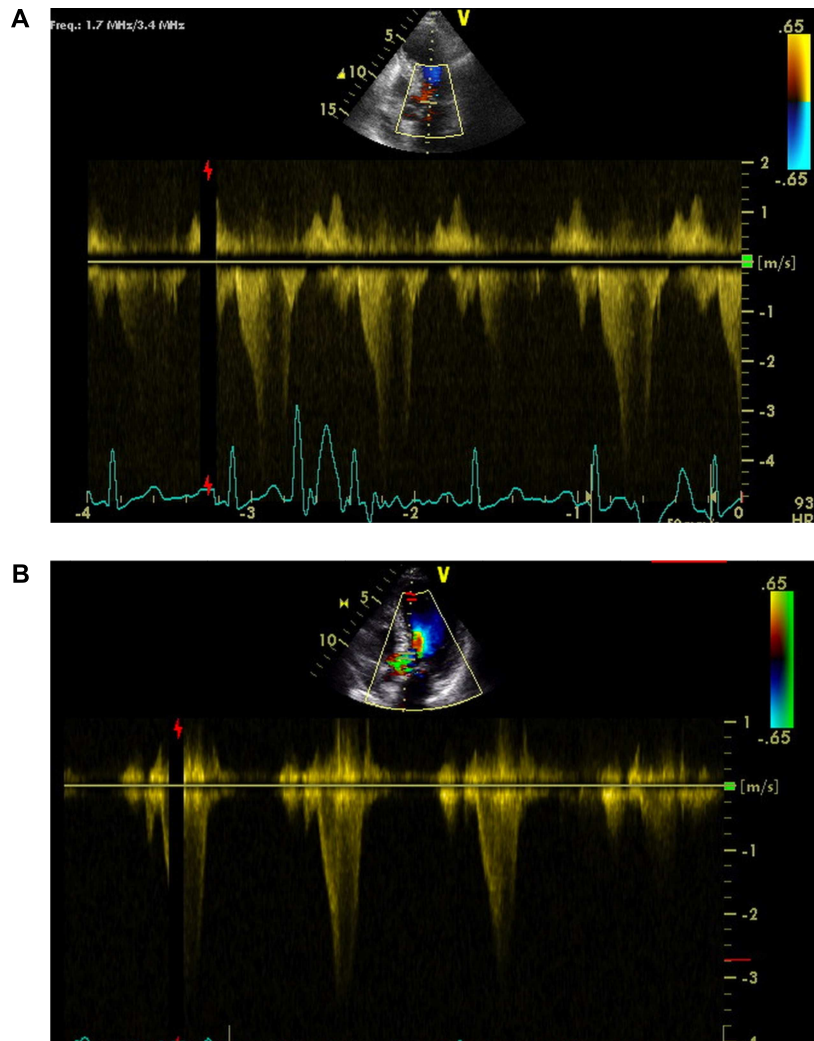


Figure 2: Echo with LVOT obstruction (a), echo with LVOT obstruction (b).

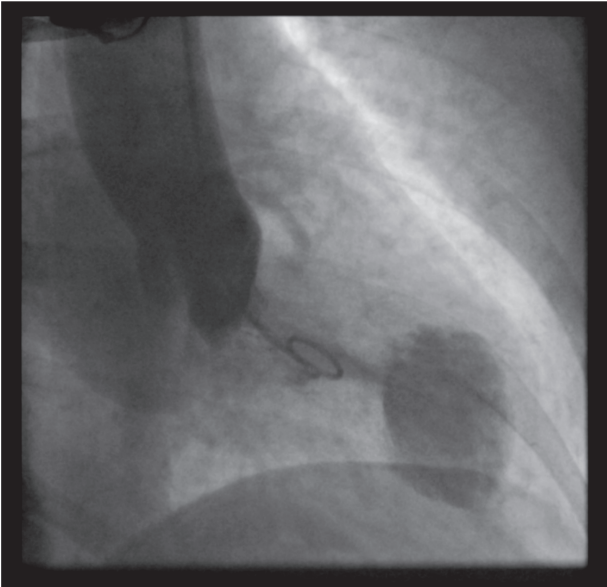


Figure 3: Left ventriculography.

murmur 4/6 in the left upper sternal border. The electrocardiogram (ECG) showed concave ST elevation in leads V2–V6, and Q waves in II, III and aVF (Fig. 1), while her troponin I levels were high (1.21 ng/ml, ‘negative’ < 0.2 ng/ml). Her echocardiogram showed asymmetric interventricular septal hypertrophy (1.5 cm), with severe apical dyskinesia with an ejection fraction (EF) estimated at 40–45%. A systolic anterior motion (SAM) of the mitral valve and severe LVOT obstruction (AV_{vmax} gradient 4 m/s) were also seen (Fig. 2a and b, Supplementary Video 1). A coronary angiography was performed to exclude acute coronary syndrome (ACS). The left ventriculography showed hypercontractility of the basal and mid segments with apical wall dyskinesia. There was also hypertrophy of the LV and moderate mitral valve regurgitation. The LV-aorta pullback gradient was 90 mm Hg. The coronaries were only mildly atheromatous without significant disease (Fig. 3, Supplementary Videos 2 and 3). The patient was admitted in the coronary care unit and was hospitalized for 6 days with the diagnosis of Takotsubo syndrome and LVOT obstruction. During her hospitalization she was hemodynamically stable and received supportive medical treatment with intravenous fluids and beta-blockers (b-blockers). Serial ECGs were performed, which showed the eventual formation of negative T waves in I, aVL, V3–V6 and biphasic T waves in II, III and aVF (Fig. 4). On Day 6, an echo was performed, which showed EF 55%, mildly hypokinetic apex and no LVOT obstruction with or without Valsalva maneuver. A month after her discharge the echo was repeated; a normal EF was seen with no regional hypokinesias and no LVOT obstruction (Fig. 5). A cardiac magnetic resonance imaging was also performed 2 months after discharge and demonstrated normal EF with a mildly hypertrophic LV and a sigmoid septum (measured up to 14 mm) and no LVOT obstruction. No areas of late gadolinium enhancement were seen. She remains well without any symptoms and her hypertension was finally controlled with b-blockers, diuretics and angiotensin converting enzyme inhibitors.

DISCUSSION

Up to 20–25% of patients with TCM may develop LVOT obstruction and/or acute mitral regurgitation (MR), which are severe complications that can lead to heart failure, shock and/or pulmonary edema [3]. In a paper from De Backer [4], it was shown that older age, septal bulging, SAM-induced MR and hemodynamic instability are associated with this condition and these patients are also more likely to present in cardiogenic shock as compared to their cohorts without LVOT obstruction.

The presence of SAM of the mitral valve and septal bulging appears to be a common feature and a predisposing factor causing the LVOT obstruction [3, 5].

In cases of TCM complicated with LVOT obstruction, vasodilation can exacerbate the pathology and should therefore be avoided. Inotropic therapy should also be avoided as it can increase circulating catecholamines, exaggerate basal hyperkinesia and worsen LVOT obstruction. Instead, these cases should be managed with fluid resuscitation and b-blockers with the use of intra-aortic balloon pump when necessary [3].

It is essential to look for a possible LVOT obstruction in TCM since it is a frequent finding that will help guide the appropriate therapeutic approach. The management of LVOT obstruction in TCM seems similar to the management of LVOT obstruction in hypertrophic cardiomyopathy. This includes the use of b-blockers that can aid in the resolution of the obstruction [6]. As catecholamine levels are increased in TCM, it may also be reasonable using b-blockers until full recovery and resolution of the EF but clinical trials are still required to support this hypothesis. Furthermore, an increase in preload will also help relieve the LVOT obstruction. This can be achieved by adequate hydration (IV fluids) or even leg elevation if the patient has no pulmonary congestion.

This case was very interesting in several aspects but mostly attributable to its clinical presentation, the diagnostic challenge and the LVOT obstruction caused by the combination of a hypertrophic septum along with TCM causing a diagnostic dilemma of a coexistence of Takotsubo with hypertrophic cardiomyopathy. It is interesting that the syncopal episode occurred during dinner at a wedding party, where most likely the increased vagal tone augmented the degree of LVOT obstruction because of decreased blood pressure, eventually leading to the syncopal episode. Thus, the combination of septal hypertrophy along with TCM can lead to a more severe clinical presentation [7–10].

CONCLUSION

TCM can be complicated with LVOT obstruction in older patients with septal bulging and early recognition of the situation will facilitate to appropriate treatment with IV fluids and b-blockers.

Learning points:

- It is important to keep in mind the differential diagnosis of syncope as well as chest pain with positive troponin and ECG changes.
- Suspect something other than ACS when the ECG has no typical characteristics (e.g. concave ST elevations, no mirror image)
- First exclude ACS to safely work on the rest differential afterwards. ECG changes, positive troponin values and motion abnormalities on echo mandate ACS exclusion.

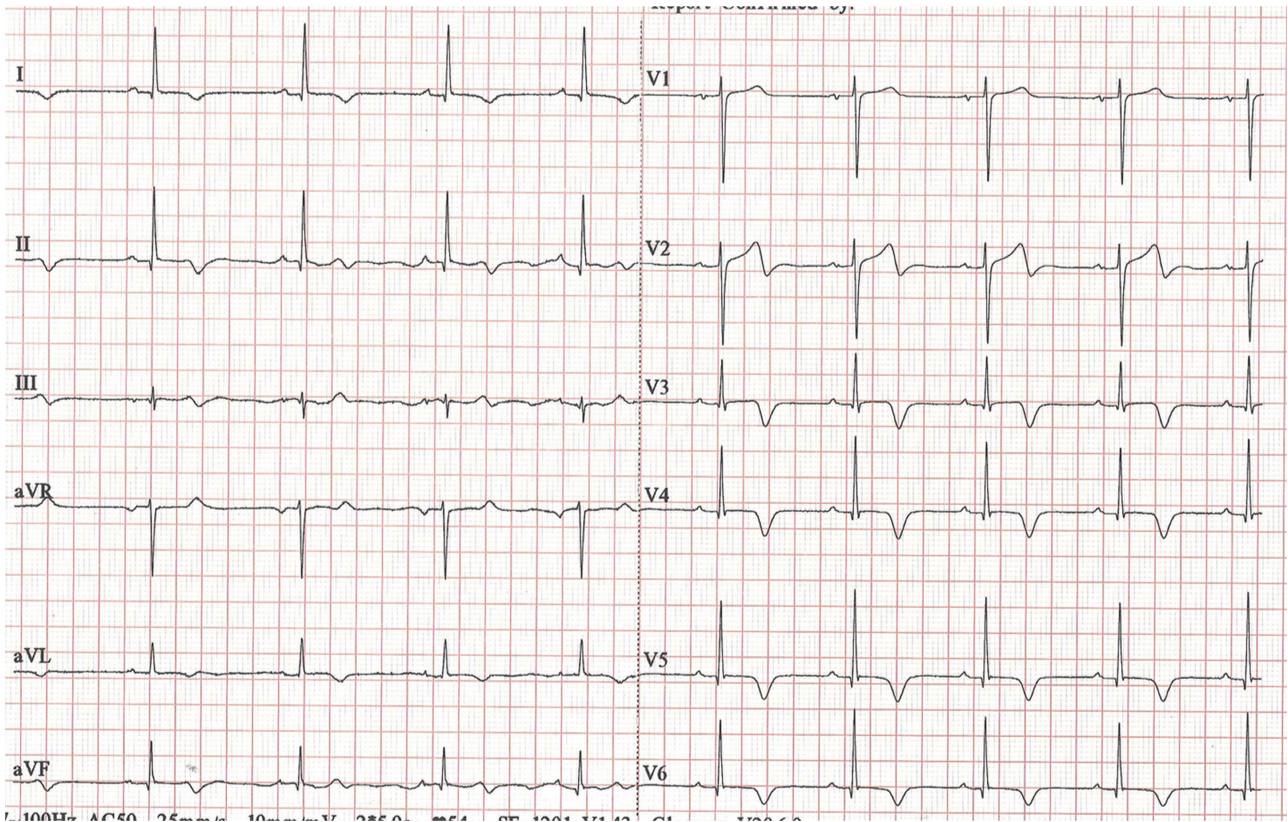


Figure 4: Follow-up ECG.

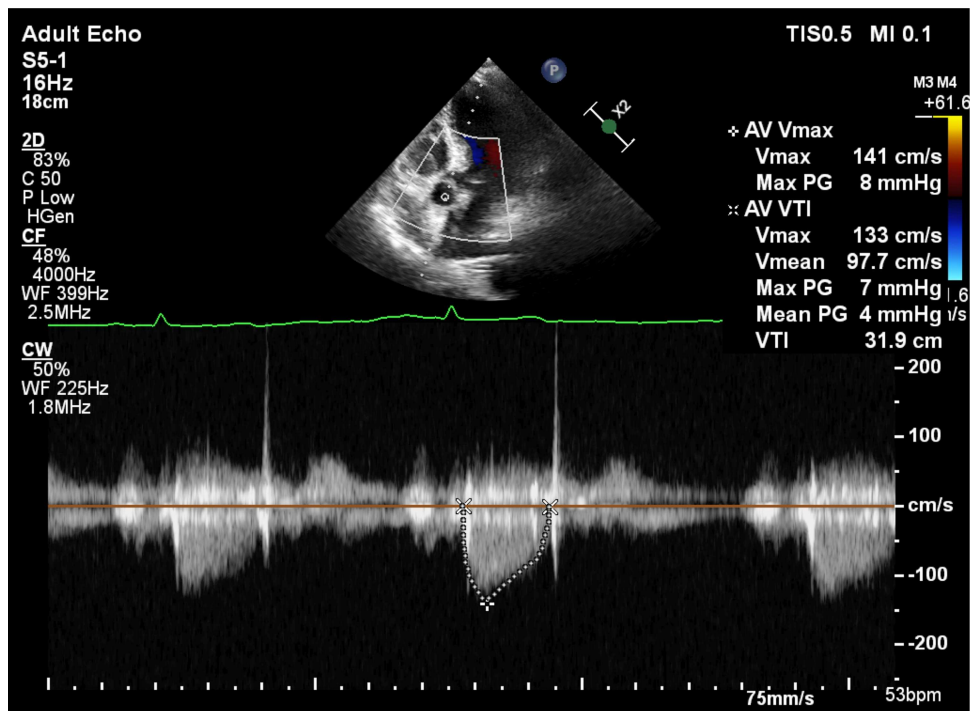


Figure 5: Follow-up echo with no LVOT obstruction.

SUPPLEMENTARY DATA

Supplementary data mentioned in the text are available to subscribers in OMCREP online.

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None.

CONFLICT OF INTEREST STATEMENT

None declared.

ETHICAL APPROVAL

Ethical clearance/informed consent obtained by the patient.

CONSENT

Patient consent was obtained.

GUARANTOR

Christos Eftychiou.

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