

Two Cases of Apical Ballooning Syndrome Masking Apical Hypertrophic Cardiomyopathy

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Apical akinesis and dilation in the absence of obstructive coronary artery disease is a typical feature of stress-induced (takotsubo) cardiomyopathy, whereas apical hypertrophy is seen in apical-variant hypertrophic cardiomyopathy. We report the cases of 2 patients who presented with takotsubo cardiomyopathy and were subsequently found to have apical-variant hypertrophic cardiomyopathy, after the apical ballooning from the takotsubo cardiomyopathy had resolved. The first patient, a 43-year-old woman with a history of alcohol abuse, presented with shortness of breath, electrocardiographic and echocardiographic features consistent with takotsubo cardiomyopathy, and no significant coronary artery disease. An echocardiogram 2 weeks later revealed a normal left ventricular ejection fraction and newly apparent apical hypertrophy. The 2nd patient, a 70-year-old woman with pancreatitis, presented with chest pain, apical akinesis, and a left ventricular ejection fraction of 0.39, consistent with takotsubo cardiomyopathy. One month later, her left ventricular ejection fraction was normal; however, hypertrophy of the left ventricular apex was newly noted. To our knowledge, these are the first reported cases in which apical-variant hypertrophic cardiomyopathy was masked by apical ballooning from stress-induced cardiomyopathy. (*Tex Heart Inst J* 2014;41(2):179-83)

Key words: Cardiomyopathy, hypertrophic/complications/diagnosis; heart ventricles/physiopathology; takotsubo cardiomyopathy/diagnosis/physiopathology; time factors; ventricular dysfunction, left

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Case Reports

Patient 1

In June 2011, a 43-year-old white woman with a history of alcohol abuse emergently presented with confusion, agitation, and shortness of breath of 2 days' duration. She had discontinued alcohol use and metoprolol succinate therapy 3 days before presentation. Her medical history included hypertension, previous tobacco use, and alcohol-related liver disease. She had no known coronary disease. Her temperature was 36.7 °C; heart rate, 125 beats/min; blood pressure, 100/64 mmHg; respiratory rate, 34 breaths/min; and oxygen saturation, 96% on room air. Physical examination revealed tachycardia, non-tender hepatomegaly, and mild pitting edema in the lower extremities. The patient was confused, disoriented, and irritable without focal neurologic deficits.

An electrocardiogram (ECG) showed sinus tachycardia with T-wave inversions and nonspecific ST-T-wave abnormalities in the anterolateral leads. Laboratory results included an elevated troponin T level of 0.27 ng/mL (normal, <0.03 ng/mL) and normal renal function. A transthoracic echocardiogram (TTE) showed an LV ejection fraction (LVEF) of 0.47 with hyperdynamic basal function but with a dilated, akinetic apex that suggested an apical variant of stress-induced cardiomyopathy (Fig. 1).

Results of coronary angiography were normal. On the 6th hospital day, TTE showed minimal improvement in LVEF and no change in the apical ballooning and akinesis. The maximal troponin T level was 0.29 ng/mL. The patient was discharged from the hospital on β -blocker and angiotensin-converting enzyme (ACE) inhibitor therapy, with an appointment for addiction counseling.

The patient returned 2 weeks later for repeat TTE. The apical wall-motion abnormalities had resolved, and the LVEF had returned to normal at 0.69. Newly apparent hypertrophy of the LV myocardium at the apex, with additional involvement of the right ventricular apex, was consistent with apical-variant HCM (Fig. 2A). A contrast agent was administered, and no apical pouches, thrombi, or gradients were observed (Fig. 2B). The maximal LV wall thickness was 13 mm at

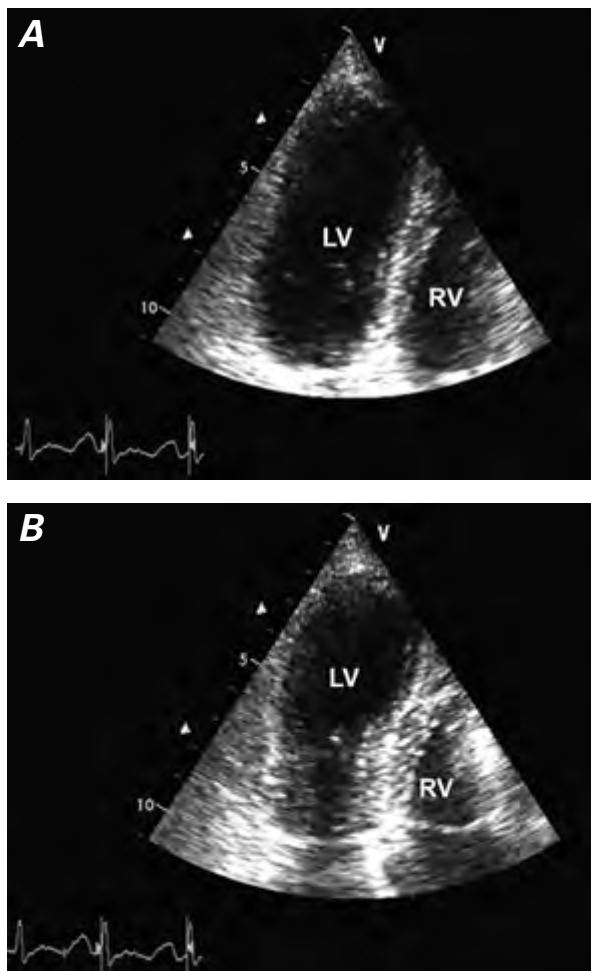


Fig. 1 Patient 1. Admission transthoracic echocardiogram (apical 4-chamber view) shows images at **A**) end-diastole and **B**) end-systole. At end-systole, apical ballooning with a small basal-cavity diameter is apparent.

LV = left ventricle; RV = right ventricle

Supplemental motion image is available for Figure 1.

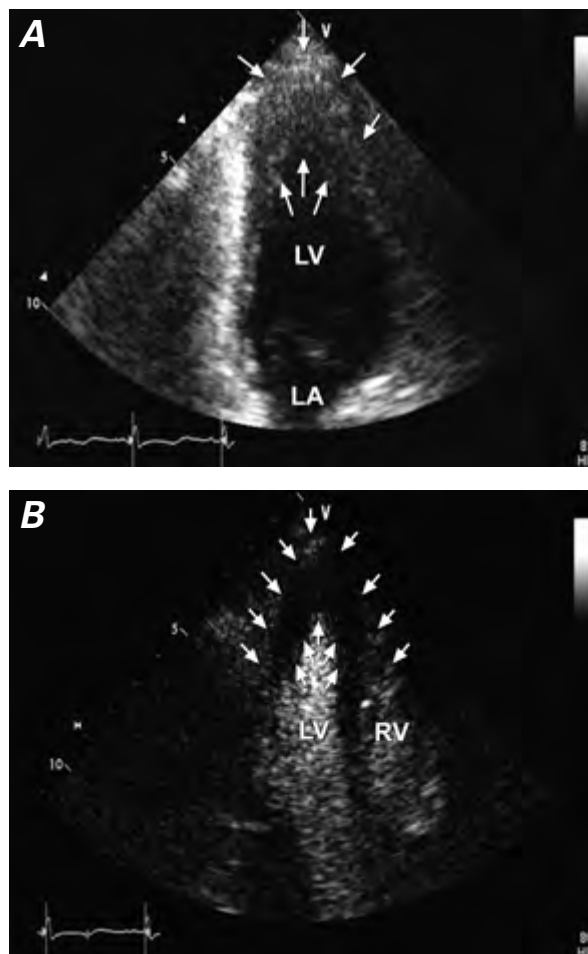


Fig. 2 Patient 1. Transthoracic echocardiograms. **A**) Apical 2-chamber view shows localized hypertrophy of the left ventricular (LV) apex (arrows) and a spade-shaped LV cavity. **B**) Apical 4-chamber view with contrast shows localized hypertrophy of the LV apex (arrows).

LA = left atrium; RV = right ventricle

end-diastole. The patient was counseled in regard to the diagnosis of apical HCM. At her 3-month follow-up examination, she was asymptomatic and doing well.

Patient 2

In July 2011, a 70-year-old white woman presented with epigastric and chest pain that radiated to her back and jaw. Associated symptoms included nausea, dry heaves, and mild shortness of breath. Her relevant medical history included stress-induced cardiomyopathy in 2006 with subsequent resolution, mild CAD, ongoing tobacco use, hypertension, hyperlipidemia, chronic abdominal pain of unclear cause, and cholecystectomy. Her medications included oxycodone, amitriptyline, and dexlansoprazole.

The patient's vital signs were as follows: temperature, 36.9 °C; heart rate, sinus tachycardia at 122 beats/min; blood pressure, 123/73 mmHg; and respiratory rate, 17 breaths/min. Results of a cardiopulmonary examination

revealed nothing unusual. She had mild epigastric tenderness without guarding or rebound. An ECG showed sinus tachycardia, T-wave inversion in the anterolateral leads, and poor precordial R-wave progression. Laboratory values included an elevated troponin T level of 0.3 ng/mL and a creatinine kinase-MB fraction of 10 ng/mL (normal, <3.8 ng/mL). Elevated levels of amylase (171 U/L), lipase (525 U/L), aspartate aminotransferase (183 U/L), and alanine aminotransferase (84 U/L) were consistent with acute pancreatitis. Other laboratory results were normal, including renal function.

The patient's admission TTE showed a depressed LVEF of 0.39 with akinesis and dilation of the mid and apical LV segments, suggesting an apical variant of takotsubo cardiomyopathy (Fig. 3). Cardiac catheterization yielded mild, nonobstructive CAD. Left ventriculographic results were consistent with apical ballooning syndrome (Fig. 4). The imaging studies and

clinical presentation confirmed acute and subacute pancreatitis. The patient was discharged from the hospital 5 days later on therapy with aspirin, a β -blocker, an ACE inhibitor, and a statin.

One month later, TTE revealed an LVEF of 0.65 without regional wall-motion abnormalities. However, the patient's LV apex was thickened, with an "ace of spades" cavity configuration that suggested apical HCM; the maximal wall thickness was 14 mm at end-diastole (Fig. 5). The patient was counseled in regard to



Fig. 3 Patient 2. Admission transthoracic echocardiogram (apical 4-chamber view) shows akinesis of the mid and apical segments in **A**) end-diastole and **B**) end-systole, typical of apical ballooning syndrome.

LA = left atrium; LV = left ventricle; RV = right ventricle

Supplemental motion image is available for [Figure 3](#).

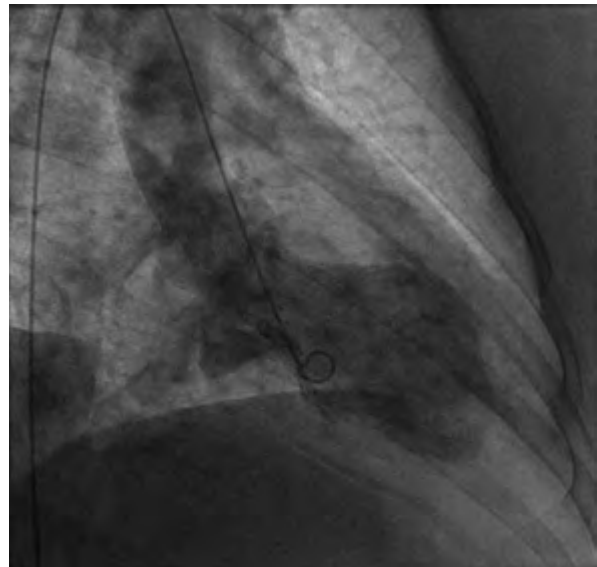


Fig. 4 Patient 2. Left ventriculogram at end-systole shows systolic apical akinesis with hyperdynamic basal function, concurrent with the echocardiographic findings.



Fig. 5 Patient 2. Transthoracic echocardiogram (apical 4-chamber view) one month after hospitalization shows localized hypertrophy of the left ventricular apex (arrows) and a spade-shaped ventricular cavity, typical of apical hypertrophic cardiomyopathy.

LA = left atrium; LV = left ventricle

Supplemental motion image is available for [Figure 5](#).

the diagnosis of apical HCM. At her 2-month follow-up examination, she was asymptomatic and doing well.

Discussion

The surprise in these 2 cases was that, after the patients' LV function recovered, the echocardiographic findings were consistent with apical HCM. No significant intracavitary gradient was noted in either patient on admission, while they were hospitalized, or during follow-up examination. To our knowledge, these are the first reported cases in which apical HCM was masked on presentation by takotsubo cardiomyopathy with apical ballooning. The apical HCM was not apparent until the apical myocardium had fully recovered and each patient underwent repeat TTE.

Takotsubo cardiomyopathy is characterized by reversible LV regional wall-motion abnormality in the absence of obstructive CAD, pheochromocytoma, and myocarditis.¹ Patients are typically postmenopausal women who present with clinical, ECG, and biochemical features of an acute coronary syndrome. The proposed pathogenesis is a sympathetic and catecholamine surge after a mental or physical stress that has caused ischemia and myocardial stunning.³ The condition is reversible, and myocardial contractility returns to normal within days to weeks. It is unknown whether ACE inhibitors hasten LV recovery and chronic β -blocker therapy prevents recurrences.¹

Hypertrophic cardiomyopathy is characterized by hypertrophy of the LV myocardium that cannot be explained by other systemic diseases and that occurs without dilation of the chambers, usually with an LV wall thickness of at least 15 mm.^{2,4} The condition is due to an autosomal dominant genetic mutation in one of the genes that encodes the myocardial contractile proteins.² Multiple genetic and morphologic forms of HCM, including apical HCM, were first described in

Japanese patients who had localized hypertrophy of the LV apex with a spade-shaped cavity on images and giant negative T waves in the left precordial leads on ECG.^{5,6} The diagnosis of apical HCM can be established with the use of echocardiography, left ventriculography, computed tomography, or cardiac magnetic resonance. Currently, most cases are diagnosed echocardiographically. Injecting a contrast agent for LV opacification helps to define the ventricular cavity's shape. Apical HCM is most often seen in Asian populations. Its prognosis is thought to be more benign than those of other forms of HCM, although there is still a substantial risk of cardiovascular morbidity and death.² Associated complications include atrial fibrillation, myocardial infarction, heart failure, and sudden cardiac death. Left ventricular outflow tract obstruction is not typically found, although apical-to-mid-LV obstruction might be seen, with the occasional formation of an infarcted apical pouch.² Patients are usually only mildly symptomatic; however, severe increases in wall thickness can evoke dyspnea, angina, and fatigue. Apical myectomy might be of benefit in markedly symptomatic patients.⁷

Our patients had apical myocardial stunning from different stressful precipitating factors. Patient 1 presented with acute, severe alcohol withdrawal and in an extremely hyperadrenergic state. Patient 2 had acute pancreatitis with severe pain. Of note, her previous episode of takotsubo apical stunning was precipitated by epigastric pain related to passing a gallstone, which led to cholecystectomy. Her LV function had returned to normal one month after that episode of stress-induced cardiomyopathy; however, apical HCM was not evident at that time because of suboptimal imaging and lack of contrast use to better delineate the apex.

Five reported cases of takotsubo cardiomyopathy with obstructive HCM were all in patients with asymmetric septal hypertrophy, not apical-variant HCM (Table I).⁸⁻¹² All 5 patients had systolic anterior motion

TABLE I. Reported Cases of Stress-Induced Apical Cardiomyopathy in Patients with HCM

Report	Age (yr), Sex	Clinical Presentation	HCM Variant	LV Wall Thickness (mm)	Resting LVOT Gradient (mmHg)
Jaber WA, et al. ⁹ (2006)	65, F	Chest pain and SOB	ASH	20	40
Singh NK, et al. ⁹ (2008)	79, F	Chest pain	ASH	22	20
Modi S and Ramsdale D ¹⁰ (2011)	54, F	Chest pain	ASH	19	25
Brabham WW, et al. ¹¹ (2011)	48, M	Chest pain and SOB	ASH	20	70
Daralammori Y, et al. ¹² (2012)	70, F	SOB	ASH	24	45
Current	43, F	SOB	Apical	13	None
Current	70, F	Chest pain	Apical	14	None

ASH = asymmetric septal hypertrophy; F = female; HCM = hypertrophic cardiomyopathy; LV = left ventricular; LVOT = left ventricular outflow tract; M = male; SOB = shortness of breath

of the mitral valve, which created an LV outflow tract gradient that would increase LV wall stress and possibly contribute to increased apical wall stress and ballooning.^{11,13}

Wall thickness in patients with HCM typically exceeds 15 mm in affected segments; in one series on apical HCM,² the average was 19 mm. However, wall thickness in HCM can vary considerably, and mild cases with thicknesses of 13 to 15 mm have been reported.^{14,15} In our patients, the wall thicknesses of 13 and 14 mm suggested a mild form of apical HCM. In addition, the degree of apical wall thickness was out of proportion to the basal segments, with spade-shaped apical cavities typical of apical HCM. Although our patients had less than the average apical wall thicknesses, these values are still within 2 standard deviations of thicknesses reported for apical HCM.² Patient 1 also had apical right ventricular involvement. It is unknown whether apical-HCM patients with greater wall-thickening at the apex would also show apical ballooning with stress-induced myocardial stunning. We suspect that apical ballooning is more likely to occur in patients such as ours, when the wall thickness is less severe and could allow for ventricular cavity dilation.

Irreversible apical myocardial infarction can occur in apical HCM in the absence of demonstrable CAD and stress-induced cardiomyopathy. Patients thus affected tend to have hypertrophy of additional myocardial segments basal to the cardiac apex and develop high-velocity gradients through a narrow tunnel just superior to the apical segments.¹⁶ The degree of apical wall-motion abnormality in such situations can vary from apical hypokinesis to apical pouches to large apical aneurysm.² This apical dilation (and often scarring) appears to be permanent, unlike the reversible stunning in our patients.

On the basis of our experience with these 2 patients, we think that anteroapical systolic akinesis and ballooning due to stress-induced takotsubo cardiomyopathy can mask apical-variant HCM and therefore render HCM unrecognizable on initial evaluation, to become apparent only after the apical myocardium has recovered fully.

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