

Multimodality Cardiac Imaging in the Evaluation of a Patient with Near-Fatal Arrhythmia

A previously well 64-year-old man presented with 2 episodes of non-vertiginous giddiness during exercise. The patient had no history of drug use, prior history of syncope or family history of sudden cardiac death. Systolic blood pressure was 55 mmHg on presentation at the emergency department, with the presenting electrocardiography

(ECG) shown (Fig. 1). Synchronised cardioversion was successfully performed and the postcardioversion ECG is as below (Fig. 1). The patient underwent a coronary angiogram which showed minor coronary artery disease, followed by transthoracic echocardiography and cardiac magnetic resonance imaging (MRI) (Figs. 2 and 3).

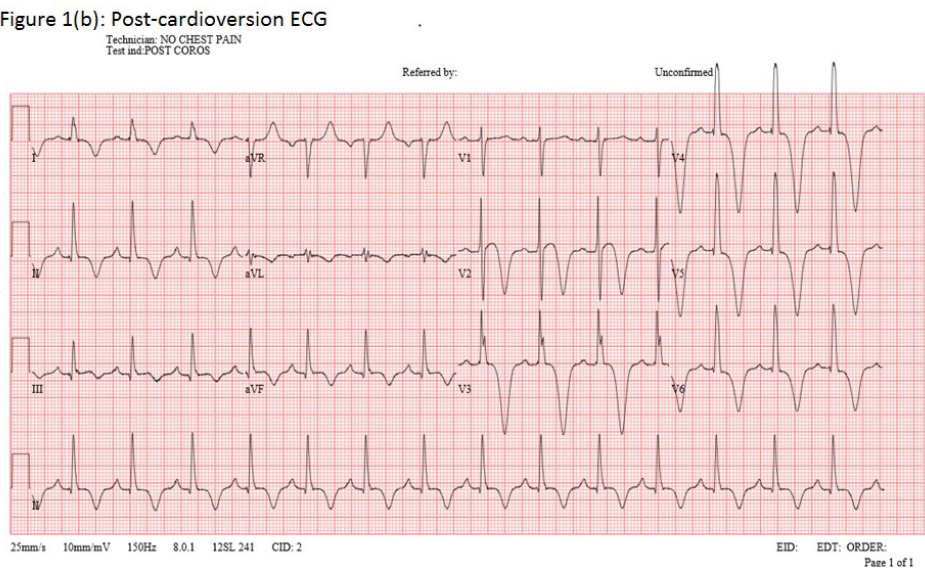


Fig. 1. ECG at initial presentation and postcardioversion ECG monomorphic ventricular tachycardia originating from the LV apex converting to sinus rhythm with deep, symmetrical T wave inversions suspicious for apical hypertrophic cardiomyopathy.



Fig. 2. Transthoracic echocardiography 4-chamber view showing left ventricular (LV) apical aneurysm with mid-ventricular and apical hypertrophy. Doppler interrogation showed increased gradient, consistent with narrowing at the entrance of the LV aneurysm.

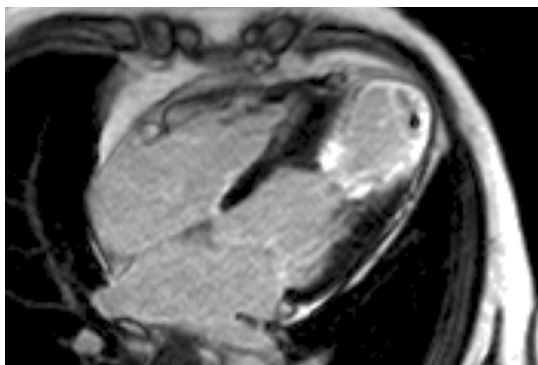


Fig. 3. Cardiac magnetic resonance imaging with late gadolinium enhancement sequence demonstrating subendocardial fibrosis within left ventricular (LV) aneurysmal wall, as well as the presence of a thrombus within the LV apex.

Given the above clinical presentation and multimodality imaging findings, what is the likely underlying diagnosis?

- A. Dilated cardiomyopathy
- B. Tachycardia-induced cardiomyopathy
- C. Apical hypertrophic cardiomyopathy with apical aneurysm
- D. Cardiac amyloidosis
- E. Takotsubo cardiomyopathy

The clinical presentation and imaging findings are consistent with apical hypertrophic cardiomyopathy (ApHCM) with apical aneurysm, presenting with unstable ventricular tachycardia. The postcardioversion ECG demonstrated precordial T-wave inversions associated with ApHCM. The 4-chamber view on transthoracic echocardiography (TTE) demonstrated hypertrophy that was localised to the left mid-ventricular and apical region, with aneurysm of the apical portion of the left ventricle. Figure 4 shows an example of a different patient with ApHCM without aneurysm formation for comparison. Subsequently, late gadolinium enhancement sequences of the cardiac MRI showed subendocardial fibrosis of the apical wall of the left ventricle and additionally morphologically demonstrated

the apical hypertrophy and aneurysm, with an LV thrombus contained within.

The expanded and aneurysmal LV apex and hypertrophy on TTE was more consistent with ApHCM with apical aneurysm rather than Takotsubo cardiomyopathy or dilated cardiomyopathy. In Takotsubo cardiomyopathy, the ballooned LV apex is classically reversible (usually over weeks or months), so subsequent follow-up imaging should show normalisation of LV ejection fraction and LV dimensions. In dilated cardiomyopathy, the entire LV may be dilated and the walls uniformly thinned or normal in thickness. Tachycardia-induced cardiomyopathy is usually a diagnosis made after excluding other causes of cardiomyopathy, and patients may show improvement in cardiac function after treatment of the tachyarrhythmia. Furthermore, atrial tachyarrhythmias are also more commonly associated with tachycardia-induced cardiomyopathy compared to ventricular tachyarrhythmias.¹ Lastly, the cardiac MRI findings are not consistent with cardiac amyloidosis. In cardiac amyloidosis, diffuse increased left ventricular (LV) wall thickness may be expected, with the late gadolinium enhancement images showing generalised gadolinium uptake representative of amyloid infiltration, which was not the case in this patient.

Discussion

ApHCM is more common in East Asians and typically has a more benign clinical course compared to non-ApHCM.² When associated with apical aneurysm formation, these patients may, however, experience more significant complications.³ Although its incidence is unclear, in 1 study, 11 out of 46 patients with ApHCM had apical aneurysms, and this was associated with increased severity of cavity obliteration.⁴

We present the case of a premorbidly well middle-aged man who presented with 2 episodes of non-vertiginous giddiness during exercise. The patient had no relevant history of drug use or family history. He presented with severe hypotension at the emergency department, with

Answer: C

ECG showing monomorphic ventricular tachycardia originating from the LV apex (Fig. 1). Synchronised cardioversion was successfully performed. The postcardioversion ECG demonstrated deep T-wave inversions (Fig. 1).

Subsequently, the patient underwent emergency coronary angiogram, which showed minor coronary artery disease. TTE performed raised the suspicion of ApHCM with an apical aneurysm (Fig. 2). There was associated LV mid-cavity obstruction as well. The LV ejection fraction was mildly reduced at 45%. Cardiac MRI confirmed these findings, and further demonstrated the presence of a thrombus within the LV apical aneurysm (Fig. 3). For ease of comparison, a different patient with ApHCM without aneurysm is shown as well (Fig. 4).

The patient was started on anticoagulation, and an automatic implantable cardioverter-defibrillator (AICD) was implanted prior to discharge.

The mechanism of apical aneurysm in hypertrophic cardiomyopathy (HCM) remains unclear. Previous authors have suggested that it may be related to the presence of mid-ventricular obstruction that led to increased intracavitary pressures and wall stress, consequently resulting in ischaemia, fibrosis and aneurysmal dilatation.⁵ Imaging in our subject favoured this pathophysiological process. Other potential factors may include genetic predisposition and left anterior descending artery myocardial bridging, but were not consistently reported in all cases of apical aneurysm.⁵

Although studied in conventional HCM, the incidence and prevalence of apical aneurysm in ApHCM remains unclear.⁶ Though typically having a more benign course, ApHCM when coexisting with an apical aneurysm likely predisposes patients to more significant complications. These include: a) malignant arrhythmias, where the myocardial fibrosis in the aneurysm wall (as shown by the gadolinium enhancement pattern on cardiac MRI) may

form an arrhythmogenic substrate, b) thrombus formation within the aneurysm, and c) poorer LV systolic function in view of the aneurysm.⁶

Our case demonstrated the important role of cardiac imaging in the management of patients with ApHCM. We also demonstrated apical aneurysm to be an important comorbidity to consider in such patients. Apical aneurysm appears to be a novel risk factor to consider in ApHCM, but it remained unclear if the presence of an aneurysm in ApHCM without prior arrhythmia or thrombus warranted AICD placement for primary prevention or prophylactic anticoagulation. Of note, pharmacologic therapy does not provide protection against malignant arrhythmia but may be used for symptom control. For patients with frequent sustained ventricular arrhythmias that require AICD shocks, adjunctive anti-arrhythmic therapy may be considered.⁷ Failing which, radiofrequency catheter ablation may also be successful in terminating the arrhythmia.⁵

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Nicholas Ngiam,¹^{MBBS}, Nicholas Chew,¹^{MBBS}, Ping Chai,²^{MBBS}, Kian Keong Poh,^{2,3}^{MBBChir, FACC}

¹Department of Medicine, National University Health System, Singapore

²Department of Cardiology, National University Heart Centre Singapore, National University Health System, Singapore

³Yong Loo Lin School of Medicine, National University of Singapore, Singapore

Address for Correspondence: A/Prof Poh Kian Keong, Department of Cardiology, National University Heart Centre, National University Health System, Singapore, 1E Kent Ridge Road, NUHS Tower Block, Level 9, Singapore 119228.

Email: kian_keong_poh@nuhs.edu.sg

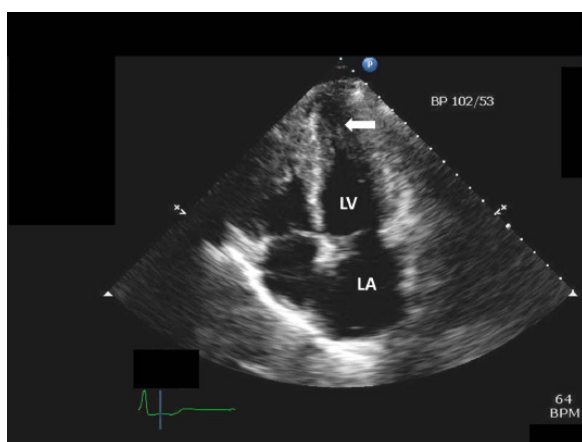


Fig. 4. Transthoracic echocardiography of a different patient at the 4-chamber window, showing hypertrophy of the left ventricular (LV) apex (arrow), consistent with apical hypertrophic cardiomyopathy (without aneurysm). LA: Left atrium.