VENTRICULAR FIBRILLATION AND SUDDEN CARDIAC ARREST IN APEX HYPERTROPHIC CARDIOMYOPATHY: TWO CASE REPORTS

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Abstract

BACKGROUND
Apical hypertrophic cardiomyopathy (HCM) is considered to have a benign prognosis in terms of cardiovascular mortality. This serial case report aimed to raise awareness of ventricular fibrillation (VF) and sudden cardiac death (SCD) in apical HCM.

CASE SUMMARY
Here we describe two rare cases of apical HCM that presented with documented VF and sudden cardiac collapse. These patients were previously not recommended for primary prevention using implantable cardioverter-defibrillator (ICD) therapy based on current guidelines. However, both received ICD therapy for the secondary prevention of SCD.

CONCLUSION
These cases illustrate serious complications including VF and aborted sudden cardiac arrest in apical HCM patients who are initially not candidates for primary prevention using ICD implantation based on current guidelines.

Key Words: Apical hypertrophic cardiomyopathy; Ventricular fibrillation; Implantable cardioverter-defibrillator; Case report

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Core Tip: Apical hypertrophic cardiomyopathy (HCM) is a rare form of non-obstructive...
HCM. It has a benign prognosis in terms of cardiovascular mortality. Here we describe two rare cases of apical HCM that presented as documented ventricular fibrillation (VF) and sudden cardiac collapse. Although apical HCM has a typically benign prognosis, clinicians must consider that VF can occur and lead to sudden cardiac arrest.

INTRODUCTION

Apical hypertrophic cardiomyopathy (HCM) is considered clinically benign, with an estimated annual mortality rate of 0.0-0.1% and no reports of sudden cardiac death (SCD) during follow-up[1]. Case reports of patients developing ventricular tachycardia (VT), mainly due to an apical aneurysmal segment and sudden cardiac collapse, have been reported[2,3]. However, documented ventricular fibrillation (VF) with sudden cardiac arrest without apical aneurysm is extremely rare in patients with apical HCM.

Here we report two cases of apical HCM who presented with documented VF and sudden cardiac collapse who were previously not candidates for implantable cardioverter-defibrillator (ICD) therapy based on current guidelines.

CASE PRESENTATION

Chief complaints
Case 1: A 41-year-old man was brought to the emergency department after sudden cardiac collapse.

Case 2: A 29-year-old man was brought to the emergency department after sudden cardiac collapse.

History of present illness
Case 1: The patient had known apical HCM; however, he did not receive regular follow-up or management. He presented to the emergency department after sudden cardiac collapse during sleep.

Case 2: The patient had known apical HCM, and he had received regular follow-up at the cardiology department for apical HCM and paroxysmal atrial fibrillation (AF) over the preceding 5 years. He was brought to the emergency department after sudden cardiac collapse while working.

History of past illness
Case 1: The patient visited the cardiology outpatient department with palpitations and chest discomfort 3 years prior. Echocardiography at that time revealed apical HCM (18.7 mm thickness at the apex) with a normal left ventricular ejection fraction (LVEF; 58%) and diastolic relaxation impairment with an elevated e/e’ ratio of 22 and enlarged left atrium (51 mm) (Figure 1A). Holter monitoring did not demonstrate relevant arrhythmia, and no paradoxical blood pressure response was observed during the exercise tolerance test at that time. He was prescribed a β-blocker; however, the patient did not complete follow-up.

Case 2: The patient was diagnosed with apical HCM and paroxysmal AF 5 years prior when he presented with chest discomfort. Echocardiography revealed apical HCM (20.1 mm thickness at the apex) without apical aneurysm and a normal LVEF (59%) and a diastolic relaxation impairment with an elevated e/e’ ratio of 16 and an enlarged left atrium (57 mm) (Figure 1B). Over the preceding 5 years, he had received regular follow-up at the cardiology department and was treated with aspirin and amiodarone.
Figure 1 Echocardiography images. A: Echocardiography in case 1 demonstrated apical hypertrophy (18.7 mm; white arrows) without apical aneurysm and a normal left ventricular ejection fraction (LVEF); B: Echocardiography in case 2 revealed apical hypertrophy (20.1 mm; white arrows) without apical aneurysm and a normal LVEF.

Personal and family history
Cases 1 and 2: Family history was unremarkable for structural heart disease, syncope, or SCD.

Physical examination
Cases 1 and 2: On admission, the patients were unconscious and pulseless.

Laboratory examinations
Case 1: The patient’s troponin I level (0.36 mg/mL) was within the normal range, while the CK-MB level (23.42 ng/mL) was remarkably elevated. His electrolyte levels were within the normal ranges.

Case 2: Troponin I level (1.78 mg/mL) and CK-MB level (9.77 ng/mL) were slightly elevated. His electrolyte levels were within the normal ranges.

Imaging examinations
Case 1: Initial electrocardiography (ECG) revealed VF (Figure 2A). Biphasic 200-J defibrillation restored sinus rhythm, and his cardiopulmonary function recovered without neurologic sequelae. ECG performed after stabilization showed sinus rhythm with deep T-wave inversion (Figure 2B). Coronary angiography revealed no significant stenosis in the epicardial coronary arteries.

Case 2: Initial ECG revealed VF (Figure 3A). Biphasic 150-J defibrillation restored sinus rhythm, and his cardiopulmonary function recovered without neurologic sequelae. His ECG after stabilization was similar to that before the cardiac collapse, showing sinus rhythm with a tri-fascicular block and T-wave inversion (Figure 3B). Coronary angiography revealed no significant stenosis in the epicardial coronary arteries.

FINAL DIAGNOSIS
Cases 1 and 2: The final diagnosis was VF and aborted sudden cardiac arrest in the apical HCM.

TREATMENT
Case 1: The patient was treated with carvedilol 6.25 mg twice daily and underwent ICD implantation for the secondary prevention of SCD.

Case 2: The patient underwent ICD implantation for the secondary prevention of SCD while maintaining his current medications.
Figure 2 The initial rhythm strip and electrocardiogram in case 1. A: The initial electrocardiogram (ECG) revealed ventricular fibrillation; B: The ECG after stabilization showed sinus rhythm with deep T-wave inversion.

OUTCOME AND FOLLOW-UP
Case 1: The patient was discharged uneventfully and remained free of VF for 3 years.

Case 2: The patient was subsequently discharged uneventfully. He experienced an inappropriate shock due to paroxysmal AF; however, he has remained free of VF for 10 years.

DISCUSSION
Apical HCM is considered clinically benign with an estimated annual mortality rate of 0-0.1% with no reports of SCD during follow-up[1]. Case reports have detailed patients developing VT mainly due to an apical aneurysmal segment and sudden cardiac collapse[2-5]. However, documented VF with sudden cardiac arrest without apical aneurysm is extremely rare in patients with apical HCM.

ICD implantation is recommended in HCM patients at high risk of SCD based on current guidelines[6,7]. Neither of our patients had any established risk factors, risk modifiers, or high-risk features. Neither met the criteria for ICD implantation according to the current guidelines. However, VF and sudden cardiac arrest occurred later despite the apical HCM, which is known to be clinically benign.

Although the risk factors for VF and SCD in apical HCM are rarely evaluated because of its significantly low incidence, several parameters affecting poor outcomes were reported previously. Patients of advanced age with hypertension, diabetes, or baseline AF have poor prognosis or decreased survival[2,8]. Patients with apical HCM and poor clinical outcomes have more advanced diastolic dysfunction, increased left atrial volume, reduced myocardial contraction/relaxation properties, and increased
LV filling pressure at presentation[2]. Impaired LV diastolic function is a proposed mechanism for progressive left atrial enlargement and subsequent AF development[1, 9]. Apical aneurysm and late gadolinium enhancement extent on cardiac magnetic resonance imaging (MRI) are also independent predictors of a poor outcome[10]. However, the association between VF and these parameters has not been evaluated until now.

The hypertrophied LV apex could predispose the myocardium to ischemia due to a limited coronary blood flow reserve. The foci of cellular disarray throughout the hypertrophied LV wall might impair the transmission of normal electrophysiological impulses and predispose that region to a disordered pattern of depolarization and repolarization, thereby serving as an arrhythmogenic substrate[11]. Our second patient had a trifascicular block and inverted T-waves on ECG in addition to LV hypertrophy prior to VF development. These findings indicate that adverse electrical remodeling had already progressed in the myocardium and may have predisposed the patient to developing VF.

CONCLUSION

Although apical HCM has a typically benign prognosis, VF can occur and lead to sudden cardiac arrest. Our case reports support the concept that clinical outcomes in patients with apical HCM are not always as benign as previously thought. We should be aware of serious complications, including VF, and aborted sudden cardiac arrest in apical HCM patients who are initially not candidates for primary prevention ICD implantation based on current guidelines. Risk factors such as diastolic dysfunction, late gadolinium enhancement on cardiac MRI, or electrical remodeling on ECG should be evaluated further for risk stratification for VF and SCD in cases of apical HCM.
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