Ventricular tachycardia and heart failure in a patient of mid-ventricular obstructive hypertrophic cardiomyopathy with apical aneurysm: A case report

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Received September 24, 2018; Accepted February 21, 2019

DOI: 10.3892/etm.2019.7796

Abstract. Mid-ventricular obstructive hypertrophic cardiomyopathy (MVOHC) with apical aneurysm is a rare type of cardiomyopathy. It is associated with an elevated risk of ventricular arrhythmias, thromboembolism, heart failure and sudden cardiac death. The present case study reports on a patient with MVOHC and apical aneurysm who developed ventricular arrhythmias and heart failure. The patient received an implantable cardioverter defibrillator for prevention of fatal arrhythmias. Ventricular tachycardia was terminated by increased doses of amiodarone and β-blocker. Transthoracic echocardiography indicated a mid-ventricular gradient of 64 mmHg. The patient refused surgical treatment and opted for alcohol septal ablation (ASA). At the 6-month follow-up, a rebound of the gradient following ASA was observed on echocardiography. In the present study, timely recognition of MVOHC with apical aneurysm led to prompt defibrillator implantation for prophylaxis regarding further malignant arrhythmias. Surgical management should be considered in symptomatic patients with MVOHC and apical aneurysm.

Introduction

Hypertrophic cardiomyopathy (HCM) is an inherited disorder of the cardiac muscle caused by a genetic defect and affects approximately one in 500 individuals (1). Patients with HCM present with multiple morphologic variants. Obstructive hypertrophic cardiomyopathy (OHC) is a classic pathophysiological feature of HCM, characterized by unexplained left ventricular (LV) hypertrophy in association with non-dilated ventricular chambers (1). OHC is associated with an increased risk of clinical deterioration and cardiovascular mortality (1). Obstruction can cause an increase in LV systolic pressure, which leads to prolonged ventricular relaxation, elevated LV diastolic pressure, mitral regurgitation, myocardial ischemia and decreased cardiac output (2-4). The presence of an aneurysmal apex in association with mid-ventricular obstruction is a rare morphological subgroup within the heterogeneous cardiomyopathy phenotypic spectrum (5). Mid-ventricular OHC (MVOHC) with apical aneurysm portends a wide spectrum of adverse outcomes, including ventricular arrhythmias, thromboembolism, heart failure and sudden cardiac death (6).

The present study presents a rare case of MVOHC combined with apical aneurysm and its management.

Case presentation

A 61-year-old Chinese male was admitted to his local cardiology department in June 2016 due to episodes of syncope after palpitation at rest. The patient had no family history of heart disease and his electrocardiogram indicated monomorphic ventricular tachycardia (Fig. 1). The initial troponin I level was 42.96 ng/ml and the patient was diagnosed with ST-segment elevation myocardial infarction. The abnormal electrocardiogram prompted invasive coronary angiography, which provided no evidence of obstructive coronary artery disease. The electrocardiogram that was then performed exhibited sinus rhythm, ST elevation and T wave abnormality (Fig. 2). The sinus rhythm was quickly restored but could not be maintained.

After 10 days, the patient was transferred to the Department of Cardiology of West China Hospital (Chengdu, China). The electrocardiogram revealed sinus rhythm, ST elevation (≥1 mm) and T wave abnormality in V2 through V6 (negative T waves). Stabilization of the patient was obtained after electric cardioversion and concomitant use of β-blockers and amiodarone. Coronary angiograms indicated no narrowing of any major epicardial coronary arteries. Transthoracic echocardiography revealed normal dimensions of the LV (48 mm) with mid-cavity hypertrophy (maximal wall thickness in the interventricular septum, 25 mm), left atrial dilatation (39 mm), a measured ejection fraction of 62% and an aneurysmal apex (Fig. 3). A subsequent cardiac acoustic contrast demonstrated clear evidence of HCM with an hourglass shape of the LV and a 35x32 mm apical aneurysm (Fig. 4). The patient then underwent dual chamber implantable cardioverter defibrillator (ICD) implantation for prevention of fatal ventricular...
arrhythmias (Fig. 5). Subsequently, the patient remained clinically and hemodynamically stable.

One year later, the patient experienced an electrical storm with 100 episodes of sustained ventricular tachycardia (VT) and multiple ICD shock (Fig. 6). Recurrent sustained monomorphic VT was terminated by increased doses of amiodarone and β-blocker. Furthermore, the patient experienced...
dyspnea on exertion. On transthoracic echocardiography, an apical aneurysm and a provoked mid-ventricular gradient of 64 mmHg were noted. The patient refused surgical treatment and but provided written informed consent to undergo alcohol septal ablation. Hemodynamic assessment revealed a significant intra-ventricular pressure gradient of 69 mmHg (LV apex, 169 mmHg; LV outflow, 100 mmHg; Fig. 7A). Repeated hemodynamic measurements after the procedure revealed a decrease of the post-extrasystolic gradient to 37 mmHg (LV apex, 135 mmHg; LV outflow, 98 mmHg) at the mid-ventricular level (Fig. 7B). The fourth and the fifth septal branches were dominant and echocontrast injection into target vessels resulted in the largest echocontrast view in the hypertrophic muscle segment. Therefore, the fourth and the fifth septal branches were assumed as target vessels and occlusion of these vessels resulted in a significant decrease in the intraventricular pressure gradient. The post-procedural echocardiographic examination indicated reductions in the left atrial dimension to 30 mm, intra-ventricular septum thickness to 20 mm and a mid-ventricular peak systolic gradient of 19 mmHg. There were no further VT episodes during treatment with amiodarone (200 mg, once a day) and metoprolol (47.5 mg, twice per day).

At the 6-month follow-up, the patient had developed dyspnea on exertion (New York Heart Association functional class III (7). Echocardiography indicated marked intracavitary LV obstruction at the mid-ventricular level, with a peak gradient of 80 mmHg. The LV ejection fraction was 45% (Fig. 8).

**Discussion**

According to the 2011 American College of Cardiology Foundation (ACCF)/American Heart Association (AHA)
guidelines, the clinical entity HCM is defined as a disease state characterized by unexplained LV hypertrophy associated with nondilated ventricular chambers in the absence of any other cardiac or systemic disease (8). Based on the echocardiogram at the onset of the disease, indicating persistent ST-segment elevation associated with T-wave inversion in the left precordial leads, the absence of obstructive coronary artery disease and clear evidence of HCM with an apical aneurysm on Doppler echocardiography, the patient of the present study was diagnosed with MVOHC and apical aneurysm.

Development of an apical aneurysm from MVO was observed in a single-center patient cohort during the 4-year follow-up period (9). Maron et al (5) hypothesized that apical aneurysm formation and the decrease in cardiac output may be secondary to increased LV wall stress as a result of mid-cavity LV obstruction and elevated intracavitary systolic pressure. Increased wall stress imposes an increased pressure load on the apical myocardium, increasing the oxygen demand, while simultaneously reducing the coronary flow through extravascular compression of the coronary artery, leading to chronic myocardial ischemia and aneurysm formation (5). MVOHC with apical aneurysm was reported to have a poor long-term outcome, with a high risk of sudden death and potentially lethal arrhythmic events (6,10). The conventional primary prevention risk markers in individual HCM patients are the following (1): i) Family history of ≥1 HCM-associated sudden cardiac death; ii) ≥1 episode of unexplained, recent syncope; iii) massive LV hypertrophy (thickness, ≥30 mm); iv) unsustained VT on serial ambulatory 24-h (Holter) electrocardiograms; and v) hypotensive or attenuated blood pressure response to exercise. ICDs following cardiac arrest or sustained VT in HCM patients for secondary prevention have achieved widespread acceptance (8,11). A previous study also reported that the presence of an apical aneurysm may be a novel risk factor necessitating ICD implantation (5). Considering the clinical context associated with the patient's hemodynamic instability, the patient of the present study received an ICD for prevention of fatal ventricular arrhythmias.

One year later, the patient experienced an electrical storm with 100 episodes of sustained VT and multiple ICD shock. Recurrent sustained monomorphic VT was terminated by pharmacologic therapy with increased doses of amiodarone and β-blocker. Catheter ablation is a controversial method for patients with MVOHC and apical aneurysm. VT recurrence in patients with non-ischaemic cardiomyopathy attributed to the myocardial tissue scarring of pathogenetic processes (12). A previous study reported on monomorphic VT in a case of MVOHC with apical aneurysm, and VT recurred on the next day after ablation (13). In addition, acute ablation failure is frequently associated with inability to identify the VT focus and control VT by catheter ablation (14). In the patient of the present study, ICD was implanted to prevent sudden death due to ventricular arrhythmias. Furthermore, amiodarone and β-blocker were administered to prevent arrhythmic recurrences. Therefore, the VT ablation was not performed.

According to the 2011 ACCF/AHA Guidelines for the Diagnosis and Treatment of HC, patients with marked LV outflow tract obstruction (peak instantaneous gradient at rest or with exercise, ≤50 mmHg) whose severe symptoms are refractory to maximum tolerant medical therapy and invasive relief of outflow tract obstruction should be considered (8). For HCM with subaortic obstruction and severe drug-refractory symptoms, surgical septal myectomy should be the gold standard for septal reduction therapy in centers with comprehensive experience (15). Alcohol septal ablation (ASA) has been proposed as a less invasive alternative to surgical myectomy in patients with advanced age, those that had previously received a pacemaker or defibrillator, as well as those with significant comorbidities and a strong aversion to surgery (16,17). Seggewiss and Faber (18) have reported on a case of mid-ventricular obstruction, which responded favorably to alcohol ablation. However, another previous study compared the effectiveness of ASA and transaortic extended myectomy (TEM) in HCM with MVO. The results indicated that TEM may provide a more reliable reduction in gradients compared to ASA, while serious ventricular arrhythmias were uncommon after either of the two therapies (19). Furthermore, a recent clinical study reported that surgical myectomy appeared to be protective against further malignant arrhythmias (20). Considering the high risk of surgery, the patient refused surgical treatment and underwent ASA for relief of intracavitary obstruction.

At the 6-month follow-up, rebound of the gradient following alcohol ablation was observed with an LV ejection fraction of 45% determined by echocardiography. Previous studies have reported that deterioration in LV function may due to aneurysm formation, leading to primary regional systolic impairment and cavity dilatation as a compulsory remodeling mechanism (5,21). Therefore, surgical management may be a consideration in certain patients with advanced heart failure to enlarge the LV cavity.

In summary, timely recognition of MVOHC with apical aneurysm may prompt prophylactic defibrillator implantation in clinical practice for further malignant arrhythmias. Surgical management should be considered in symptomatic patients with MVOHC and apical aneurysm.

Acknowledgements

Not applicable.

Funding

No funding was received.

Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Authors’ contributions

XL performed the operation and perioperative medical treatment. XL performed pre-operative examination and diagnosis. XZ collected the data and wrote the manuscript. All authors read and approved the final manuscript.

Ethics approval and consent to participate

Written informed consent was obtained from the participating patient.
Patient consent for publication

Written informed consent for publication was obtained from the patient.

Competing interests

The authors declare that they have no competing interests.

References