Electrical storms in patients with apical aneurysms and hypertrophic cardiomyopathy with midventricular obstruction: A case series

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Video clip is available online.

Ventricular arrhythmias commonly are associated with apical aneurysms in patients with hypertrophic cardiomyopathy (HCM) and midventricular obstruction (MVO). Electrical storms are a severe manifestation of ventricular arrhythmias, defined as 3 or more episodes of sustained ventricular tachycardia (VT), ventricular fibrillation, or appropriate shocks from an implantable cardioverter-defibrillator (ICD) during a 24-hour period. In this report, we describe 3 patients with HCM and apical aneurysms complicated by electrical storms.

CASE 1

A 46-year-old man with HCM developed recurrent VT and was referred to our clinic for surgical opinion. Three years before presentation, he had a thromboembolic stroke. On transthoracic echocardiography (TTE), a large apical aneurysm and provoked midventricular gradient of 34 mm Hg were noted. At the time, he was managed medically with warfarin, amiodarone, mexiletine, and metoprolol. However, 2 years later, the patient experienced a cardiac arrest, and after resuscitation, an ICD was placed. In the following year, his ICD fired approximately 60 times. One electrical storm was triggered by vigorous exercise, during which he experienced 18 shocks within 30 minutes.

The patient was referred for apical aneurysmectomy in hopes of controlling the ventricular arrhythmias and eliminating a source of thromboembolism. At operation, we performed a transapical myectomy to relieve the obstruction at the midventricle. We encountered extensive subendocardial scarring, which was resected together with the apical aneurysm (Video 1).

Transthoracic echocardiogram of an apical aneurysm and MVO.

Central Message

Resection of apical aneurysms and associated endocardial scarring might reduce the risk of debilitating electrical storms in patients with hypertrophic cardiomyopathy and midventricular obstruction.

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The patient had an uncomplicated postoperative course. Eight years after surgery, however, the patient experienced another electrical storm. He is now 12 years postsurgery and has not experienced any further ventricular arrhythmias.

VIDEO 1. Surgical resection of an apical aneurysm and midventricular myectomy via a transapical incision. Video available at: [http://www.jtcvsonline.org/article/S0022-5223\(17\)31161-3/addons](http://www.jtcvsonline.org/article/S0022-5223(17)31161-3/addons).

FIGURE 1. Preoperative transthoracic echocardiogram of the left ventricle (*long-axis view*). A, Apical aneurysm and MVO. B, Same view with contrast. MVO, Midventricular obstruction.

CASE 2

A 57-year-old man with MVO and recurrent VT was referred for surgical opinion.¹ An ICD was placed due to

multiple syncopal episodes, and subsequently, he experienced an electrical storm with 5 episodes of sustained VT and multiple ICD shocks. Before surgical consultation,

FIGURE 2. Intraoperative exposure and resection of the apical aneurysm. A, Thin-walled aneurysm at the apex of the left ventricle. B, Incised aneurysm can be seen. C, Left ventricle after aneurysm resection and midventricular myectomy. D, The left ventriculotomy is closed in 2 layers.

medical management with amiodarone, mexiletine, and dofetilide had been attempted but was unsuccessful at relieving the recurring ventricular arrhythmias. On preoperative TTE, marked hypertrophy of the midventricle and a small apical aneurysm were seen.

A transapical ventriculotomy was made to resect the aneurysm and relieve MVO. The apical aneurysm had a large amount of subendocardial scarring, which was excised together with the aneurysm. The anterolateral papillary muscle was markedly hypertrophied and was shaved to reduce risk of midventricular obstruction.

The patient had an uneventful postoperative course. At follow-up 3 years after surgery, he reports no recurrence of the ventricular arrhythmias and had no further ICD shocks.

CASE 3

A 74-year-old woman was referred to our clinic with a history of recurrent VT refractory to medical treatment with amiodarone, mexiletine, and carvedilol. She had a background of HCM with midventricular obstruction. An ICD was placed, but ventricular arrhythmias led to frequent shocks and multiple electrical storms. Her most recent storm resulted in 13 successive ICD discharges. On preoperative TTE, a resting midventricular gradient of 38 mm Hg was noted, and a large apical aneurysm was seen (Figure 1), which was thought to be responsible for the recurring ventricular arrhythmias.

Through a transapical incision, a midventricular septal myectomy was performed to relieve intracavitary obstruction, and the large apical aneurysm and underlying scar were excised (Figure 2). Postoperatively, the patient did not experience any ventricular arrhythmias. She has remained asymptomatic and had no further VT or ICD shocks at 2-year follow-up.

DISCUSSION

Apical aneurysms are thought to arise from high midventricular gradients, causing increased wall stress in the left ventricle and subendocardial ischemia and scarring.^{2,3} All 3 of our patients had marked obstruction at the midventricle, which we believe was a significant risk factor leading to aneurysm formation. Ventricular arrhythmias commonly are associated with apical aneurysms in patients with HCM and midventricular

obstruction. In these patients, VT often is refractory to medical treatment and may result in electrical storms.¹

In a recent clinical study, Rowin and colleagues⁴ recommended conservative treatment of patients with apical aneurysms using anticoagulation for prevention of thromboembolism and ICD for patients with malignant ventricular arrhythmias. These authors specifically cautioned against surgical treatment of apical aneurysms, as they rarely cause heart failure and do not rupture. Catheter ablation may be acceptable in patients with small apical aneurysms in whom a localized arrhythmogenic area is suspected. However, midventricular obstruction and the increased risk of thromboembolism are not managed adequately by catheter-based treatment. Surgical resection of apical aneurysms in patients with MVO and/or nonobstructive apical hypertrophic cardiomyopathy may be indicated if there is recurrent VT refractory to medication, and arrhythmia is thought to arise from extensive endocardial scarring, often associated with sizable aneurysms.^{1,5} Electroanatomic map-guided surgical ablation and cryoablation of apical aneurysms have been reported in the literature.⁵ However, in our experience, mapping and cryoablation are not necessary, and we prefer linear resection of the aneurysm and wide excision of surrounding scar tissue.¹

CONCLUSIONS

In patients with apical aneurysm and electrical storm, resection of the aneurysm and endocardial scarring might reduce the risk of further ventricular arrhythmias, especially recurrence of debilitating electrical storms.

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