Natural history of apical hypertrophic cardiomyopathy and novel surgical treatment

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Disclosures: Authors have nothing to disclose with regard to commercial support.

Received for publication Jan 28, 2016; revisions received March 1, 2016; accepted for publication March 5, 2016; available ahead of print April 5, 2016.

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J Thorac Cardiovasc Surg 2016;152:626-7

0022-5223/\$36.00

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<http://dx.doi.org/10.1016/j.jtcvs.2016.03.017>

A 47-year-old woman with hypertrophic cardiomyopathy was referred for further evaluation. Twenty-five years earlier, a left ventriculogram had shown a “spade-like” configuration of the left ventricular (LV) cavity in systole (Figure 1, A) and diastole (Figure 1, B), confirming the diagnosis of apical hypertrophic cardiomyopathy. The patient had done well until 4 years ago, when she noticed palpitations and exertional dyspnea. One year ago, she experienced 2 episodes of transient ischemic attack. She was subsequently found to have both atrial fibrillation and nonsustained ventricular tachycardia. She underwent radiofrequency ablation, restoring normal sinus rhythm. However, she continued to experience severe dyspnea and her cardiopulmonary exercise test revealed severe cardiac output limitation. Normal coronary arteries were present on coronary angiography.

Cardiac magnetic resonance imaging (MRI) revealed an apical aneurysm and complete obliteration of the midventricular cavity (Figure 2). For persistent symptoms and thromboembolic events, she underwent apical ventriculotomy, removal of an apical thrombus, resection of the apical pouch,

Apical hypertrophic cardiomyopathy and postmyectomy enlargement of the left ventricular cavity.

Central Message

We present the natural history of apical hypertrophic cardiomyopathy over a period of 25 years and the findings of a novel surgical treatment in 1 patient.

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and extensive midventricular myectomy. Postoperative cardiac MRI showed complete resolution of midventricular obliteration and resection of the apical aneurysm (Figure 3, A) and enlargement of the LV cavity in diastole (Figure 3, B). The patient had an improvement in symptoms with no further ventricular ectopy or embolic events.

These images illustrate the natural history of apical hypertrophic cardiomyopathy over a 25-year period. Apical aneurysms are present in 2% to 3% of patients with

FIGURE 1. Left ventriculogram in (A) systole and (B) diastole, showing the early stages of apical aneurysm formation in hypertrophic cardiomyopathy.

FIGURE 2. Cardiac magnetic resonance imaging showing the presence of an apical aneurysm with midventricular cavity obliteration in (A) systole and (B) diastole.

hypertrophic cardiomyopathy and are associated with worse long-term prognosis.^{1,2} Progression of apical aneurysm was believed to be due to continued subendocardial ischemia from high LV wall stress. This resulted in midventricular obstruction, which then led to higher levels of apical wall stress and ischemia. Resection of the apical aneurysm and midventricular obstruction is a novel surgical treatment for these patients³ to improve symptoms by enhancing diastolic filling and to prevent ventricular arrhythmias or thromboembolic complications. This surgical procedure has evolved and can be performed

by experienced surgeons with low operative risk and excellent outcomes.

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FIGURE 3. Postsurgical cardiac magnetic resonance imaging showing (A) complete resection of the apical aneurysm and (B) enlargement of the left ventricular cavity.