Case Report

Combined Cox Maze Procedure, Septal Myectomy, and Mitral Valve Replacement for Severe Hypertrophic Obstructive Cardiomyopathy Complicated by Chronic Atrial Fibrillation

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Atrial fibrillation (AF) has been reported to be an important prognostic indicator for clinical deterioration particularly in patients with hypertrophic obstructive cardiomyopathy (HOCM). A 66-year-old female complained of severe exertional dyspnea and tachycardia, which were resistant to medical treatments. Doppler echocardiography demonstrated a peak left ventricular outflow tract (LVOT) gradient of 117 mmHg at rest. A catheter examination revealed left ventricular end diastolic pressure of 34 mmHg, a cardiac index of 1.94 L/minute/m^2, and a peak LVOT gradient of 70 mmHg at rest. A transaortic septal myotomy/myectomy was performed first, and Cox maze III procedure was performed through the right and left atrium followed by mitral valve replacement. The patient recovered dramatically except for temporary complete atrioventricular block. One year after operation, the patient is doing well with sinus rhythm and the echocardiogram revealed a peak LVOT pressure gradient of 7.6 mmHg at rest. This surgical approach might be recommended for the treatment of AF in HOCM. (Ann Thorac Cardiovasc Surg 2003; 9: 323–5)

Key words: maze, septal myectomy, mitral valve replacement (MVR), atrial fibrillation (AF), hypertrophic obstructive cardiomyopathy (HOCM)

Introduction

Hypertrophic obstructive cardiomyopathy (HOCM) causes a left ventricular outflow tract (LVOT) obstruction resulting from the combination of left ventricular (LV) hypertrophy, usually septal, and systolic anterior motion (SAM) of the mitral valve. Atrial fibrillation (AF) has been reported to be an important prognostic indicator for clinical deterioration in patients with hypertrophic cardiomyopathy (HCM), particularly in those with LVOT obstruction. In this case, combined septal myectomy, mitral valve replacement (MVR),[1)] and Cox maze procedure,[2)] were employed to treat severe symptomatic LVOT obstruction complicated with chronic AF.

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Case

A 66-year-old female consulted a local practitioner for palpitation. She was diagnosed as HOCM with paroxysmal AF and was treated medically. From one year before operation, the paroxysmal AF became chronic. Since her condition deteriorated with exertional dyspnea and tachycardia [New York Heart Association (NYHA) functional class III], which were resistant to medical treatments with propranolol, diltiazem, and disopyramide, she was referred to our hospital for surgery. A physical examination revealed a grade 4/6 systolic murmur along the left sternal border. Electrocardiogram showed AF rhythm with LV hypertrophy and inverted T in I, aVL, V5, V6. Doppler echocardiography demonstrated LVOT obstruction at rest, with a peak pressure gradient of 117 mmHg. The interventricular septum thickness was 20.2 mm and posterior wall thickness was 22.5 mm. Mitral regurgitation of 1+ with severe SAM was also seen (Fig. 1). A catheter examination revealed pulmonary wedge pressure of 17 mmHg, pulmonary artery pressure of 29/18 (23) mmHg,

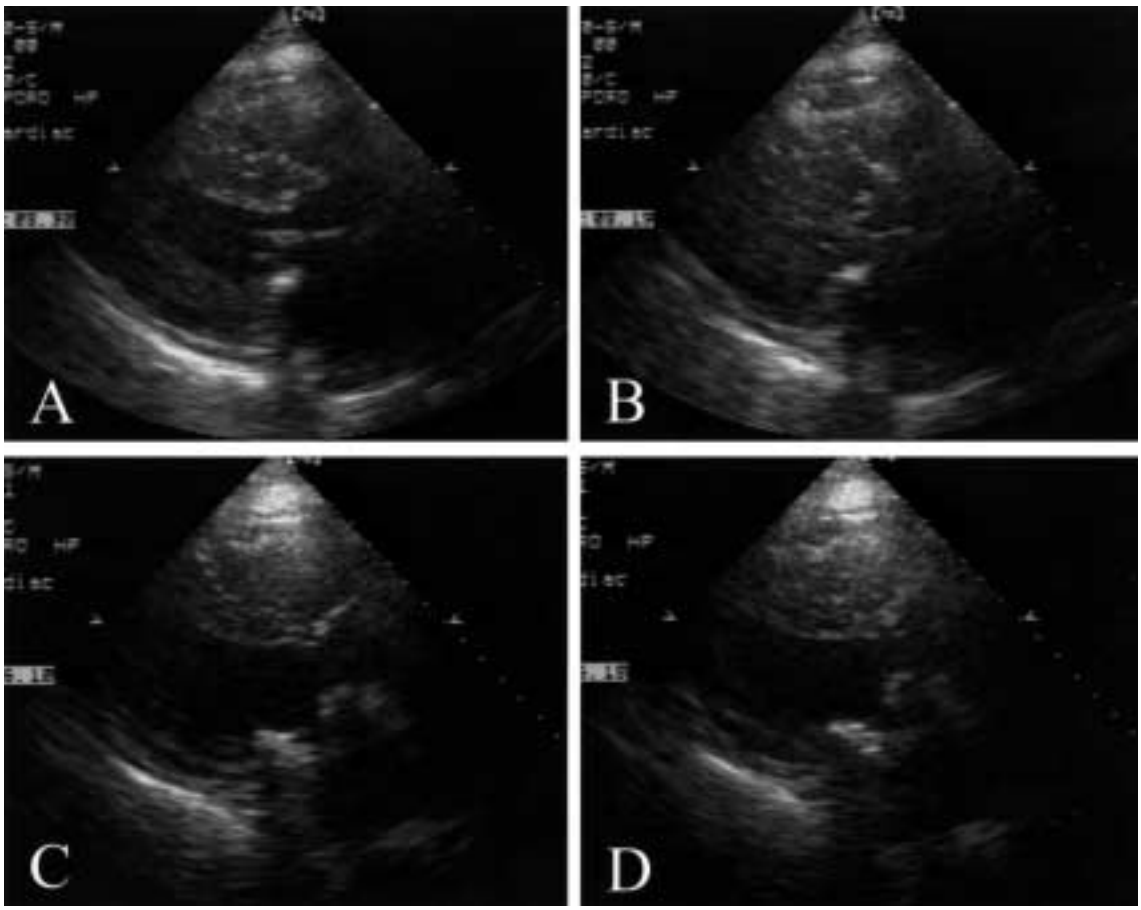

Fig. 1. The echocardiography before and after surgery.
Diastolic (A) and systolic (B) phase of LV longitudinal view before operation. In systolic phase, LV inner volume looked completely diminished.
Diastolic (C) and systolic (D) phase of LV longitudinal view one year after operation. Note that wall thickness decreased to normal level.

LV end diastolic pressure of 34 mmHg, a cardiac index of 1.94 L/minute/m^2, and a peak pressure gradient between the left ventricle and the ascending aorta of 70 mmHg at rest. Coronary angiogram showed normal coronary arteries. After admission, AF tachycardia occurred frequently in the early morning, which had to be treated by propranolol even when blood pressure fell to 60 mmHg.

Following these examinations, the patient underwent surgery. A transaortic septal myotomy/myectomy was performed first, and Cox maze III procedure was performed through the right and left atrium. An MVR was performed using a Saint Jude Medical (SJM) 27 bileaflet valve with preservation of the posterior leaflet. Aortic cross-clamping time was 184 minutes and extracorporeal time was 291 minutes. The patient recovered uneventfully without inotropic supports by cathecholamine, except for temporary complete atrioventricular block treated by DDD pacing. Postoperative echocardiography before discharge revealed a peak LVOT gradient of 5.7 mmHg at rest.

One year after operation, the patient is now doing well with NYHA functional class I. Electrocardiogram showed sinus rhythm with non specific intraventricular conduction block (Fig. 2). Echocardiogram showed a peak LVOT pressure gradient of 7.6 mmHg, and the interventricular septum thickness decreased to 10.1 mm and posterior wall thickness also decreased to 9.1 mm (Fig. 1).

Discussion

HOCM is a disorder characterized by asymmetric hypertrophy of the interventricular septum associated with SAM of the mitral valve leading to LVOT obstruction. Generally, the LVOT pressure gradient and subjective symptoms can be improved by medications such as beta-

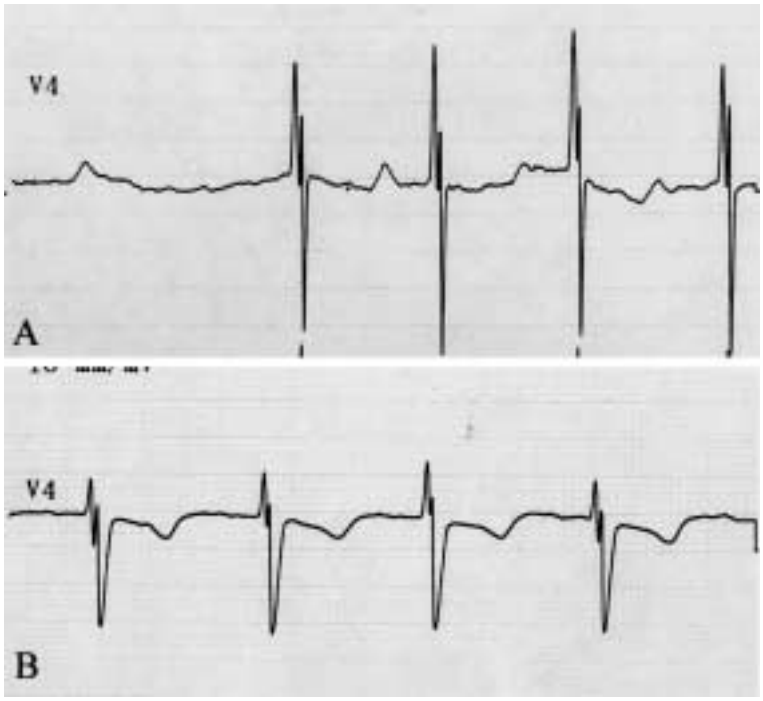

Fig. 2. The electrocardiogram before and after surgery.
A: ECG before operation showed AF.
B: ECG one year after operation showed sinus rhythm.

blockers, calcium antagonists, disopyramide or amiodarone, DDD pacing and the injection of absolute alcohol into the first major septal artery. Surgery should be performed in patients with HOCM resistant to the above treatments. LV myotomy/myectomy or MVR by resecting the mitral valve and papillary muscle to eliminate SAM are usually employed to treat LVOT obstruction. However, combined MVR and septal myotomy/myectomy should be employed when typical asymmetric septal hypertrophy is absent.[1)]

In our case, both the interventricular septum and posterior wall were severely thickened and LV volume was severely decreased to cause syncope when tachycardia occurred. The patient had to have proplanolol to relieve LVOT obstruction and control tachycardia even when blood pressure fell to 60 mmHg. Theoretically tachycardia causes a decrease in LV volume in HCM due to poor volume inflow to LV. Development of AF has been also reported to be a poor prognostic sign in HCM patients.[3)] Olivotto et al. reported that HCM with AF (22% prevalence over nine years) was associated with substantial risk for heart failure-related mortality, stroke, and severe functional disability, particularly in patients with outflow obstruction, those under 50 years of age, or those developing chronic AF.[4)]

From this point of view, we employed a combined septal myectomy, MVR, and Cox maze procedure. In the literature only two papers of maze procedure for paroxysmal AF in HOCM[5,6)] were reported, but no report for chronic AF in HOCM. This successful surgical case might be very rare for treating chronic AF in HOCM.

Although long-term observation is important in examining the validity of this operative procedure, this surgical approach might be recommended for the treatment of AF in HOCM.

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