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Transaortic and Transmitral Extended Myectomy and Concomitant Supracoronary Myotomy in a Girl with Hypertrophic Cardiomyopathy

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ABSTRACT

Concomitant idiopathic hypertrophic subaortic stenosis and disseminated myocardial bridging is an uncommon clinical entity with poor prognosis. We describe a symptomatic 19-year-old girl who had myocardial debridging and transaortic and transmitral extended septal myectomy in the same surgical session. An early and simultaneous surgical approach may prevent sudden cardiac death in these high-risk patients.

INTRODUCTION

The prevalence of idiopathic hypertrophic subaortic stenosis (IHSS) in the general population is 0.2% [Maron 1995]. It has been reported that 5%-10% of patients with IHSS had total occlusion of the left anterior descending coronary artery (LAD) due to myocardial bridging (MB) [Deanfield 1985]. Clinical symptoms may present any time from early infancy to the seventh decade. Patients are usually referred to a physician with symptoms of chest pain, effort dyspnea, syncope, and dizziness.

The coexistence of MB in IHSS worsens the prognosis and is one of the most common causes of sudden death in early adulthood [William 2005]. Urgent concomitant myectomy in the left ventricular outflow tract (LVOT) and myocardial debridging may prevent sudden cardiac death in patients with this condition.

CASE REPORT

A 19-year-old female patient was admitted to our clinic for progressive symptoms (dyspnea, chest pain, and palpitations) that were resistant to medical treatment. Physical examination revealed grade 2-3/6 systolic murmur in the aortic and mesocardiac area. Electrocardiograph revealed sinus rhythm and left-axis deviation. Transthoracic echocardiography (TTE) revealed a thickness of 35 mm in the interventricular septum (Figure 1A) and a left ventricular ejection fraction (LVEF) of 65%. The left ventricular outflow tract gradient was calculated as 120 mm Hg with heavy exertion during systolic anterior motion of the mitral valve. Coronary angiography demonstrated high-grade stenosis in the proximal section of the LAD and 90%-100% stenosis was also observed in the mid and distal portion of the LAD during systolic motion of the heart (Figure 2A). Grade 40%-50% stenoses attributable to MB were detected in the distal circumflex artery and the right descending posterior artery. Because the symptoms were resistant to medical treatment, surgery was scheduled quickly.

SURGICAL TECHNIQUE

After a median sternotomy was performed, cardiopulmonary bypass (CPB) was instituted. While the patient was under systemic hypothermia of 28°C, the aorta was cross-clamped and cardiac arrest was maintained with intermittent antegrade isothermic blood cardioplegia. Grossly hypertrophied myocardium (stone heart appearance) necessitated an aggressive myocardial protection policy. Therefore, additional cardioplegias were repeated every 15 min and delivered through both coronary ostiums. A supracoronary myotomy (debridging) was performed to the myocardial bridges on the LAD. A mosquito clamp was used to find a dissection plane between the overlying myocardial bridge and the LAD far distally where the LAD became visible close to the apex of the heart. Debridging was performed with a 30° Potts scissors through the whole intramural segment. To prevent restenosis, the divided muscle parts were tracted apart and fixed to the lateral side of the LAD with Teflon-supported sutures on different levels of the muscle layer (Figure 2B). Following aortotomy and left atriotomy, the hypertrophic septal myocardial tissues were resected through a transaortic and transmitral approach starting from the subaortic region and extending to the apex so no gradient would be left at the LVOT. Although effective myocardial protection measures were utilized, aortic declamping was followed by resistant ventricular fibrillation. Therefore, numerous ventricular defibrillation attempts were supported by high

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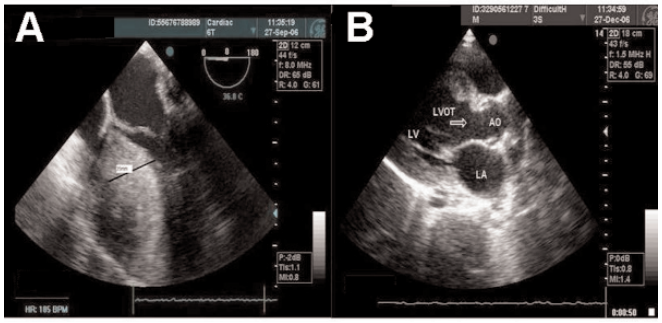

Figure 1. Transesophageal echocardiography (TEE). A, perioperative TEE image showing giant interventricular septum. B. postoperative TEE 3 months after surgery showing no obstruction on the left ventricular outflow tract TTE. LVOT indicates left ventricular outflow tract; AO, aorta; LA, left atrium.

doses of β-blocker and lidocaine treatment. Low cardiac output following cessation of CPB (due to latter explained measures) necessitated high doses of inotropic agents and intraaortic ballon pump support. Perioperative TEE revealed a septum thickness of 20 mm and no gradient left at the LVOT.

POSTOPERATIVE FOLLOW-UP

Transthoracic echocardiography on postoperative day 1 revealed an LVEF of 30%-35%, no gradient at the LVOT, and severe hypokinesia at the septum along with the apex. A control TEE on postoperative day 8 revealed an LVEF of 62%. Follow-up electrocardiographs during the patient's hospital stay recorded a sinus rhythm and left bundle-branch block. Holter monitoring for 24 h demonstrated no additional arrhythmias, ruling out the need for an implantable cardioverter defibrillator. After discharge, the follow-up TTE on postoperative day 90 demonstrated 22-mm septal thickness, 57% EF, and no pressure gradient at the LVOT (Figure 1B). After 3 months, the patient was still in normal sinus rhythm and was clinically asymptomatic.

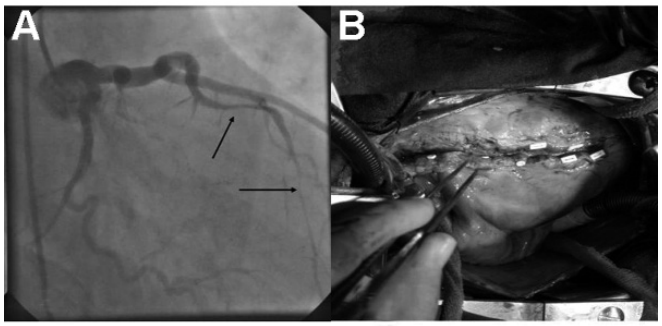

Figure 2. A, black arrows indicate left anterior descending coronary artery (LAD) stenosis in the systolic phase. B, divided muscle parts on both sides were fixed to the lateral side of the LAD with Teflon-supported 4-0 prolene sutures.

DISCUSSION

Myocardial bridges (with or without obstructive hypertrophic cardiomyopathy) were long thought to be a benign clinical entity [Downar 2004]. Mohiddin et al [1999] reported that compression of the LAD (isolated) in MB and concomitant IHSS could present a poor prognosis. Although Sorajja et al [2003] reported that MB was not linked to poor long-term outcome, including increased risk of sudden cardiac death, Yetman and coworkers [1998] reported a high prevalence of cardiac morbidity and sudden death in this cohort and recommended urgent myocardial debridging. The remarkable thickness of the septum, which typically occurs in obstructive HCM and concomitant MB, is associated with a poor prognosis compared to the absence of obstructive HCM [Fiorani 2005]. We could not find evidence in the literature of the same combination surgery that we performed in this patient, possibly because of low-grade stenoses and gradient in MB and IHSS, respectively, conditions that precluded the use of one of these surgical approaches.

Angiographic, echocardiographic, and hemodynamic studies have confirmed that the compression on the coronary artery still persisted during the diastolic phase, and the coronary flow diminished significantly in MB [Ge 1999]. Other studies have shown that supraarterial myotomy (myocardial debridging) led to improved coronary flow rate and myocardial perfusion, reduced incidence of arrhythmias, and improvement in symptoms [Downar 2004, Fiorani 2005]. In our experience, which includes 4 patients with HCM treated with off-pump or on-pump LAD bridging, supraarterial myotomy provided symptomatic relief in all patients. Beating-heart debridging performed without CPB is another treatment strategy for MB. We used the latter technique successfully to treat 3 patients with MB in the setting of nonobstuctive HCM. In the present case, long-segment stenosis in the LAD precluded stent placement. Although our patient had more than one MB on the coronary arteries, the lesions on the right and circumflex coronary arteries were distally located and caused only insignificant stenosis (40%-45%) and thus did not require debridging or revascularization. We did not use a stent or perform debridging with a beating-heart technique because we were already using CPB to resect the septal myocardium, which was causing severe LVOT obstruction.

Relieving obstruction in HCM is a technically challenging procedure because it is usually performed with a conventional approach through the aortotomy, which considerably limits surgical exposure. Such inadequate exposure may lead to inadequate surgical resection of the septal myocardium resulting in a residual gradient in the LVOT or extended resection resulting in iatrogenic ventricular-septal defect creation or complete heart block. Today, extended septal myectomy performed with the modified Morrow technique is the standard approach for IHSS, with good early and long-term outcomes [Sherrid 2003]. Casselman et al [2002] reported that the obstruction in IHSS could be treated with minimally invasive endoscopic techniques. Casselman et al used a port-access system with endoaortic clamping through the groin vessels (for CPB) and a small right thoracotomy

for a working port. To perform this procedure, however, they detached the anterior leaflet of the mitral valve from its annulus. After resection of the hypertrophied septum, a bovine pericardium was used to reattach the anterior mitral leaflet to the annulus. In our case patient, in addition to aortotomy, we used a transmitral approach through the left atriotomy to resect as much septal tissue as possible (without detaching the anterior mitral valve leaflet). We believe that the transmitral approach provides better midseptal and apical exposure and makes the myectomy in this remote area easier, safer, and more effective. Our previous experience with 2 patients in whom a mild midcavitary obstruction with 20-35 mm Hg systolic gradients occurred after transaortic myectomy prompted us to routinely perform additional transmitral myectomy. Because there was no additional mitral valve pathology we did not perform mitral valve replacement or plication of the anterior mitral valve leaflet.

Effective myocardial protection is very important in patients with HCM because the myocardium is thickened and stiff (stone like). In our limited experience in 9 patients with HCM, induction by antegrade blood cardioplegia followed by maintenance with intermittent ostial delivery every 15 min has provided successful myocardial protection. Retrograde cardioplegia has the advantages of simplifying myocardial protection and avoiding interruption of the surgical procedure, but a disadvantage is that low retrograde delivery pressure (ideally 25-40 mm Hg) may not be enough for myocardium that is 3.5 cm thick. Inadequate right ventricular protection with retrograde cardioplegia is another concern. In the present case, prolonged aortic cross-clamp time (125 minutes) and incomplete myocardial protection of gravely hypertrophied ventricles and ventricular arrhythmias occurring before the patient was removed from CPB may have been major contributors to myocardial deterioration. The stunning effect of multiple defibrillations for resistant ventricular fibrillation, however, and the administration of high-dose β-blockers, lidocaine, and amiodarone, which have negative inotropic effects on the myocardium, may be other important factors for early myocardial failure. Resistant ventricular fibrillations in the present case led us to give metoprolol (5 mg) a few minutes before aortic cross-clamp removal. Particulate emboli may be another reason for early myocardial dysfunction, a possibility that should be kept in mind. To prevent this dramatic complication in the case patient, we irrigated and aspirated the left ventricular cavity several times after myectomy.

CONCLUSION

The prevention of cardiac morbidity and sudden death is crucial in symptomatic patients with IHSS and MB resistant to medical treatment, and therefore these patients should undergo surgery as soon as the diagnosis is confirmed. A favorable long-term outcome is possible in the setting of concomitant treatment of obstructive IHSS and MB.

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