Proposal for bail-out procedures - Cardiac general

Apicoaortic conduit for the dilated phase of hypertrophic obstructive cardiomyopathy as an alternative to heart transplantation

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Abstract

We describe a 16-year-old suffering from the dilated phase of mid-ventricular obstructive hypertrophic cardiomyopathy with end-stage heart failure. Her plasma type-B natriuretic peptide level exceeded 8000 pg/ml, and she refused heart transplantation. She underwent an apicoaortic valved conduit as an alternative to the heart transplant procedure. This traditional procedure is still an ideal operation for very sick cardiomyopathy patients to avoid or delay heart transplantation.

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Keywords: Hypertrophic obstructive cardiomyopathy; Heart failure operation; Apicoaortic conduit

1. Introduction

Surgical myectomy with or without mitral valve surgery is the standard procedure for patients with drug-refractory hypertrophic obstructive cardiomyopathy (HOCM) [1, 2]. Treatment for the dilated phase of HOCM with severe heart failure is quite difficult however, and the only surgical option is usually heart transplantation. We report the case of a patient with dilated phase mid-ventricular obstructive hypertrophic cardiomyopathy with severe heart failure. The patient’s plasma type-B natriuretic peptide (BNP) level exceeded 8000 pg/ml, and the patient refused a heart transplant procedure.

2. Case

We describe a 16-year-old girl with Noonan syndrome suffering from hypertrophic cardiomyopathy since infancy. Her cardiac status became more complicated, with obstruction diagnosed at the age of ten years and a tendency toward the dilated phase of the left ventricular apical site from the age of 14 years. Her BNP level began to rise to over 2000 pg/ml at the age of 11, and now it is over 8000 (up to 12,300) pg/ml. She is mentally well developed. The patient’s BNP level began to rise to over 2000 pg/ml at the age of 11, and now it is over 8000 (up to 12,300) pg/ml. She is mentally well developed. The patient was advised to undergo a heart transplant procedure but refused this treatment. After spending more than a year at the National Cardiovascular Center for medical treatment, she was referred to the Kusatsu General Hospital for alternative surgical treatment. She was in the New York Heart Association functional class (NYHA) IV. Thoracic echocardiogram (Fig. 1) as well as reconstructed images of 64-slice computed tomography scanning (MDCT, Fig. 2a) revealed severe hypertrophy at the mid-portion of the left ventricle (LV) (ventricular septum = 29 mm; posterior free wall = 25 mm) and a thins, severe, hypokinetic to a kinetic wall at the severely dilated apical portion of the LV. The papillary muscles were also atrophic. No mitral regurgitation was detected, although slight systolic anterior movement of the chordae was noted. The ejection fraction (EF) was 16%. Cardiac catheterization demonstrated a pressure gradient of 100 mmHg between the LV apical high-pressure chamber and the aorta. With the patient in the right lateral decubitus position, the operation was performed through a median sternotomy with a continuous L-shaped anterior left chest incision. The thorax was entered through the 6th intercostal space. The heart was severely enlarged and tightly attached to the left anterior and lateral chest wall. As in usual cardiopulmonary bypass procedures, the LV apex was lifted up through the incision so that the descending aorta (15 mm in diameter) could be exposed. A second aortic cannulation to the descending aorta was placed just above the diaphragm. The descending aorta was totally occluded, and a hand-made valved conduit (20 mm Hemashield graft [Boston Scientific, Wayne, NJ, USA] with a 17-mm St Jude Medical Regent aortic valve (St Jude Medical, St Paul, Minnesota, USA]) was anastomosed in an end-to-side fashion. The postero-lateral wall 3 cm from the apex was selected for conduit implantation, and the proximal site of the conduit was anastomosed with pledged, interrupted sutures and secured with continuous sutures under fibrillation. Finally, the patient was weaned off of cardiopulmonary bypass, but her sternum was left open for five days, because the heart was enlarged after the cardiopulmonary bypass and the sternum was not able

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Fig. 1. The transthoracic echocardiogram in (a) diastole and (b) systole, showing mid-ventricular hypertrophic obstruction of left ventricle and massive dilation of the hypotrophic apical wall. Systolic anterior movement of the mitral valve was not obvious.

to be closed. The patient was placed on mechanical lung support for a while, underwent a tracheostomy, and finally was separated from the ventilator. Postoperative transthoracic echocardiography revealed significantly better movement of the apical site of the LV (EF = 40%) and a reduced LV diameter. The native trans-aortic valve flow was able to be detected with no significant pressure gradient at the obstructed site. The postoperative MDCT, which showed a 3-dimensional image of the conduit, did not reveal any problems (Fig. 2b). The patient’s BNP levels at six and 12 weeks after the operation decreased to 5600 and 2500 pg/ml, respectively. She was on warfarin and oral diuretics postoperatively. Her postoperative NYHA class increased to II, and the patient was discharged after the rehabilitation.

3. Comment

Originally, we planned to perform a surgical myectomy with mitral valve surgery (the standard procedure for HOCM) [1, 2]. However, this case was quite different from the usual surgical candidates suffering from HOCM. This patient exhibited the dilated phase of hypertrophic cardiomyopathy with severe mid-ventricular obstruction. The apical site of the LV wall had hypotrophied and showed severe hypokinesis and dilation. The muscle was still hypertrophic at the mid-portion of the LV, but it may shift to the dilated phase in the future. Although there was a high probability that the LV apex would contract better after the obstruction was resolved with the myectomy, we were reluctant to cut the properly functioning part of the
muscle. The patient had neither typical systolic anterior movement of the mitral valve nor mitral regurgitation. We did not think that mitral valve surgery, including papillary muscle surgery, would dramatically improve her condition. The aim of the surgical procedure was to extinguish the pressure gradient of the LV without damaging the contracting muscle.

The apicoaortic conduit has been one of the procedures of choice for correcting left ventricular outflow tract obstructions in high-risk patients with aortic stenosis [3, 4]. The apicoaortic valved conduit procedure was selected for this severe heart failure patient, whose BNP level exceeded 8000 pg/ml [5]. The operation was quite difficult due to the severely enlarged ventricle. An L-shaped incision was necessary to lift up the heart so that the descending aorta could be exposed, and this incision was also ideal because it established the cardiopulmonary bypass circuit. The 6th intercostal approach was chosen from the MDCT images, instead of normal 5th intercostal approach [3]. Although this patient required mechanical ventilation for a while, her NYHA class improved from IV to II.

The traditional apicoaortic valved conduit is still an ideal operation for very sick cardiomyopathy patients who wish to avoid and/or bridge to heart transplantation.

References


