Surgical correction of hypertrophic obstructive cardiomypathy in a patient with severe hypertrophy and septal myocardial fibrosis

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Abstract

In patients with hypertrophic cardiomyopathy, myocardial fibrosis is an independent predictor of an adverse outcome. A new technique of hypertrophic obstructive cardiomypathy (HOCM) surgical correction in patients with severe hypertrophy and septal myocardial fibrosis has been proposed. This approach avoids mechanical damage to the heart conduction system, and for the surgeon it improves visual inspection of the area to be resected. We present a case report of a 33-year old female patient with biventricular obstruction, extreme hypertrophy, septal myocardial fibrosis and episodes of ventricular tachycardia who underwent surgical correction according to this novel procedure. The advantage of the approach is an effective surgical treatment of HOCM in patients with severe hypertrophy and septal myocardial fibrosis who cannot be treated with the current surgical techniques.

Keywords: Hypertrophic obstructive cardiomypathy • Severe hypertrophy • Myocardial fibrosis

INTRODUCTION

Hypertrophic cardiomyopathy is the most common cause of sudden cardiac death in the young. The mechanism of sudden death in hypertrophic obstructive cardiomypathy (HOCM) is ventricular tachycardia/fibrillation emanating from areas of fibrosis [1]. The classic Morrow technique for HOCM in patients with right ventricular outflow tract (RVOT) obstruction combined with extreme left ventricular hypertrophy and myocardial fibrosis is not effective. We describe a case of HOCM surgical correction by a new approach in a patient with severe hypertrophy and septal myocardial fibrosis.

CASE REPORT

A young woman was diagnosed with HOCM at the age of 31. She complained of dyspnoea on effort [New York Heart Association (NYHA) functional class II] and frequent palpitations. In the following 2 years, she developed worsening of her congestive symptoms (to NYHA class III) despite progressive increase of metoprolol dosage (to 190 mg/day). Furthermore, she experienced two syncopal episodes on effort. The echocardiography showed asymmetric hypertrophy of the left ventricle (LV), with a maximum septal thickness of 33 mm measured at the midventricular level. Doppler examination showed LV outflow tract (LVOT) obstruction with a systolic gradient of 101 mmHg, intraventricular gradient 79 mmHg and systolic gradient in RVOT at 46 mmHg. Septal myocardial fibrosis was detected by cardiovascular magnetic resonance with delayed enhancement imaging after gadolinium infusion. Delayed hyperenhancement magnetic resonance imaging demonstrated areas of scarring in the basal and midventricular septum (Fig. 1a). The image showed non-transmural fibrosis. A run of non-sustained ventricular tachycardia was present on 24-h Holter electrocardiogram (ECG). Cardiac catheterization revealed a 101 mmHg gradient at the level of the LVOT and a 44 mmHg at the level of RVOT. During invasive investigation of the right ventricle, ventricular fibrillation was registered. Ventricular fibrillation was interrupted by defibrillation shock. Coronarography was performed for identification of septal arteries as well.

This 33-year old woman with extreme hypertrophy, biventricular obstruction and septal myocardial fibrosis underwent surgical correction on November 2008. The follow-up period was 33 months. The operation was performed through a median sternotomy using cardiopulmonary bypass. A longitudinal incision in the conal part of RV (length 27 mm) was made. All attachments and additional trabecules that exist between the anterior part of ventricular septum and the RV anterior wall were divided. During direct visual inspection of the RV anatomy, severe hypertrophy of septomarginal trabecule, RV wall thickening and small size of the RV cavity were evident (Fig. 1c). Maximum RV anterior wall thickness was increased to 10 mm. The excision of the asymmetrical hypertrophied area of the interventricular septum causing simultaneous LVOT and RVOT obstruction was performed from the conal part of the RV corresponding to the zone obstruction of LV (Fig. 1b). This excision was carried out on the right side of the interventricular septum anterior of the Lancisi muscle. The resection zone was extended down to the midventricular level. The excision in the midventricular level. The excision in the middle part of the interventricular septum was performed correspondingly to...
the area of LV intraventricular obstruction. The areas of septal myocardial fibrosis as identified by delayed enhancement imaging were excised under direct visual inspection in the middle part of the right side of the interventricular septum and in the upper third part. This excision was done not through the whole interventricular septum thickness, that is, without penetration into the LV cavity.

The clinical state of the patient improved within 7 days after the operation. Significant relief of dyspnoea and its complete disappearance occurred during the first 14 days following the surgical correction. After 3 weeks, NYHA functional class improved from 3.0 to 1.0. The patient was free of symptoms (NYHA class 1) at the time of most recent contact (33 months after surgery) and had no syncopal or presyncopal episodes. As a prophylactic treatment, the patient received mild doses (23.7 mg/day) of metoprolol.

Follow-up echocardiographic data were obtained. The LV intraventricular gradient remained low at 14 mmHg and RVOT gradient at 3 mmHg after 33 months. The thickness of ventricular septum was substantially reduced. Preoperative and postoperative (follow-up) echocardiographic data are shown in Table 1. Systolic anterior movement (second degree) was noted before surgery and was absent after the operation. Holter monitoring during the follow-up period did not show ventricular tachycardia. The ECG showed a sinus rhythm without right and left bundle branch block.

Table 1: Preoperative and postoperative echocardiographic measurements in the 33-year old patient

<table>
<thead>
<tr>
<th>Measurement</th>
<th>Preoperative</th>
<th>Postoperative</th>
</tr>
</thead>
<tbody>
<tr>
<td>LVOT gradient</td>
<td>101</td>
<td>16</td>
</tr>
<tr>
<td>RVOT gradient</td>
<td>46</td>
<td>3</td>
</tr>
<tr>
<td>Intraventricular gradient</td>
<td>79</td>
<td>14</td>
</tr>
<tr>
<td>Interventricular septum thickness, middle part (mm)</td>
<td>33</td>
<td>18</td>
</tr>
<tr>
<td>Interventricular septum thickness, upper third part (mm)</td>
<td>19</td>
<td>12</td>
</tr>
<tr>
<td>LA (mm)</td>
<td>51</td>
<td>44</td>
</tr>
</tbody>
</table>

LVOT: left ventricular outflow tract; RVOT: right ventricular outflow tract; LA: left atrium.

DISCUSSION

The presence of a scar indicated by cardiovascular magnetic resonance in HCM patients is a reliable independent predictor of all cause and cardiac mortality [2]. Patients with a basal septal scar on cardiovascular magnetic resonance had higher ventricular tachycardia frequency compared with those without [3]. In our patient with severe hypertrophy and septal myocardial
fibrosis, ventricular fibrillation requiring defibrillation occurred during catheterization of the right ventricle. In the patient, the interventricular septum thickness was 33 mm before surgery and the patient also had history of syncope and ventricular tachycardia, which has been defined as a risk factor for cardiac death. The highest risk of sudden death was reported for patients with extreme hypertrophy (30 mm or thicker) [2]. In addition, our patient had simultaneous LVOT and RVOT obstruction and LV mid-ventricular obstruction. The removal of myocardial tissue by the Morrow approach is impossible if both left and RVOTs are obstructed. A biventricular resection is associated with a high risk [4]. The Morrow technique could be useful if muscular hypertrophy is present at only the LVOT site and myocardial fibrosis is present only in upper third part of the septum. However, it is easier to remove fibrosis tissue from the right of the interventricular septum side under direct visual inspection.

The presence and severity of fibrosis are associated with a greater risk of major adverse events [3]. Inappropriate implantable cardioverter defibrillator (ICD) shocks are the most common device complication and should be accounted for when counseling high-risk HCM patients for ICD implantation [5]. In the absence of generally accepted standards for surgical treatment of HOCM patients with severe hypertrophy and septal myocardial fibrosis, a new technique of HOCM surgical correction was proposed. After surgical correction of HOCM and precise removal of septal myocardial fibrosis, the patient had no ventricular tachycardia and no syncopal or presyncopal episodes. The presented excision of interventricular septum allows the avoidance of damage to the right branch of the His bundle. The removal of obstructing tissue causing simultaneous obstruction of the LVOT and RVOT using the same approach and the possibility of precise removal of areas of septal myocardial fibrosis are important advantages of the surgical technique.

Conflict of interest: none declared.

REFERENCES