An Unusual Presentation of Persistent Dyspnea Following Alcohol Septal Ablation For Obstructive Hypertrophic Cardiomyopathy

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Abstract

Residual or recurrent symptoms after septal reduction therapy are most often related to inadequate relief of left ventricular outflow gradients. We recently encountered a 71-year-old woman with hypertrophic cardiomyopathy (HCM) and prior alcohol septal ablation who had a unique constellation of findings causing her symptoms. She was found to have four potential causes for her symptoms, residual midventricular obstruction, apical distribution of hypertrophy reducing end-diastolic volume, constrictive pericarditis, and marked arterial stiffness, as reflected by aortic atherosclerosis. She underwent complete pericardiectomy, transaortic septal myectomy, transapical myectomy, and replacement of a heavily calcified ascending aorta.

Introduction

Persistence or recurrence of symptoms early after septal reduction by surgical septal myectomy or alcohol septal ablation (ASA) is most often due to residual or recurrent left ventricular outflow tract obstruction.[1,2] After obtaining written consent from the patient, we describe in the present report that the patient had persistent exertional dyspnea for six months following ASA and was found to have multiple potential cardiac causes for her disability.

Patient presentation and management

A 71-year-old woman with the previous diagnosis of obstructive HCM presented to our Clinic with complaints of exertional breathlessness and chest fullness for three years. Because of progressive symptoms that were not relieved with medical therapy, she underwent ASA at another institution. Complete heart block complicated her post-ablation course, and she received a dual-chamber transvenous pacemaker-defibrillator. Post procedure, she continued to experience persistent limiting symptoms of exertional shortness of breath and chest fullness.

There were several important findings on her clinical examination. She had severe systemic hypertension with blood pressure of 180/90 mmHg. Her jugular venous pressure was markedly elevated with rapid X and Y descents. She had a 2/6 systolic ejection murmur at the left sternal border which increased from the squat to stand position.

Transthoracic echocardiography demonstrated septal hypertrophy and systolic anterior motion of the mitral valve producing severe left ventricular outflow tract (LVOT) obstruction with a peak Doppler gradient of 77 mmHg. There was, in addition, midventricular obstruction with gradient of 31 mmHg. Extensive calcified plaques in the ascending aorta and aortic arch were seen on CT angiography (**Figure 1**). Cardiac catheterization demonstrated a large gradient between the LV apex and aorta from combined midventricular and subaortic left ventricular outflow obstruction. The gradient decreased during inspiration (**Figure 2A**) with a Brockenborough response after a PVC (**Figure 2B**), indicating a dynamic outflow obstruction. There was also elevation and end-equalization of diastolic pressures in all four cardiac chambers to 25 mmHg,

and the cardiac index was reduced to 2.3L/min/M^2; she had evidence of enhanced ventricular interaction all consistent with the diagnosis of constrictive pericarditis **(Figure 2C)** . Additional medical problems included systemic hypertension, diabetes, obesity (BMI > 37kg/m2), paroxysmal atrial fibrillation, and chronic obstructive pulmonary disease.

At operation, the pericardium was found to be thickened and intensely constrictive. Further, severe calcification of the proximal aorta prevented an adequate aortotomy for myectomy. Prior to bypass, the gradient from the left ventricle to the aorta was 80 mm Hg at rest, and this increased to 143 mm Hg following a premature ventricular contraction (PVC). After myectomy, the gradient was 7 mm Hg at rest and 20 mm Hg following PVC.

After cannulating the proximal aortic arch for arterial inflow, a two-stage cannula was placed in the right atrium. During the initial phase of bypass, a complete pericardiectomy was performed including the anterior portion and the diaphragmatic pericardium. The aorta was cross-clamped, and the calcified midportion of the ascending aorta was excised. We then performed an extended transaortic septal myectomy. A 22-mm Hemashield replacement graft was trimmed to the correct length and sewed to the proximal and distal ends of the aorta. The apex of the heart was elevated into the wound, and through an apical ventriculotomy, we performed an extensive midventricular septal myectomy. The apical ventriculotomy was closed with felt strip reinforcement. Hemodynamics was satisfactory following cardiopulmonary bypass, and intraoperative transesophageal echocardiography demonstrated a good result from myectomy with no residual systolic anterior motion of the mitral valve and a maximum intraventricular gradient of 14 mmHg. Postoperative pathology confirmed a non-calcified fibrous thickening (up to 1 cm) and non-granulomatous lymphoplasmacytic infiltration of the pericardium. On postoperative TTE, left ventricular ejection fraction was 67% and there was no systolic anterior motion of the mitral valve or residual subaortic obstruction. She was dismissed home from the hospital 7 days after operation. At the time of last follow-up three years postoperatively the patient was well and free of cardiac symptoms.

Comment

Residual subaortic obstruction is a common cause of persistent symptoms following septal reduction therapy (SRT) and may be more common following ASA compared to surgical myectomy. As illustrated in our patient, however, other cardiac problems may contribute to poor functional outcome post procedure. Patients with HCM and midventricular obstruction have effort-related symptoms that are often indistinguishable from those caused by subaortic obstruction, and it is important to identify this before SRT. Combined subaortic and midventricular obstruction is difficult to eliminate with ASA and may require injection in multiple septal perforating arteries with excess volume of alcohol. For surgical myectomy in patients with both subaortic and midventricular obstruction, we prefer transapical approach for mid ventricular septal resection combined with transaortic exposure for the subaortic myectomy.[3]

Constrictive pericarditis (CP) causes impaired diastolic ventricular filling, [4] and it may be difficult to distinguish the symptoms of dyspnea and fatigue between CP and HCM, There were no antecedent events in our patient that might have caused constriction and raised suspicion regarding the diagnosis. Detailed hemodynamic assessment did suggest constrictive pericarditis and this important finding supported proceeding with operation. [5]

Extensive calcification of the ascending aorta, "porcelain aorta," is prevalent in the elderly and may reflect increased arterial stiffness.[6]. Of more importance to the surgeon is the potential difficulty in aortic clamping and securing hemostasis following simple aortotomy when extensive plaque is present in the ascending and sinus portions of the aorta. In the present case, excision of the ascending aorta and graft replacement appeared to be the safest method for exposing the subaortic area and for ensuring hemostasis.

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Figures:

Figure 1 . Preoperative CT angiogram (3D view) demonstrating multiple calcified plaques in the ascending and descending aorta.

Figure 2 . Preoperative hemodynamic study with tracings of the electrocardiogram. **Panel A** shows fixed midventricular obstruction. **Panel B** shows dynamic subaortic obstruction with a marked increase in gradient following a premature ventricular contraction. **Panel C** the arrows show ventricular discordance (interdependence) with an increase in right ventricular pressure and a simultaneous decrease in left ventricular pressure during inspiration suggesting constrictive pericarditis.

ECG: electrocardiogram (white line); LVP: left ventricular pressure (blue line); AP: aortic pressure (green line); LAP: left atrial pressure (red line); RVP: right ventricular pressure (gray line).

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