A Case of Successful Valve Repair for Aortic Insufficiency Associated with Discrete Subaortic Stenosis

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Discrete subaortic stenosis (DSS) is an uncommon form of left ventricular outflow tract obstruction especially in adulthood. Moreover, aortic regurgitation (AR), which is a common sequence of DSS, requiring surgical correction is extremely rare. We report the case of a 33-year-old man who had severe DSS accompanied with moderate aortic insufficiency. He underwent successful surgery including relief of DSS and aortic valve repair. Although careful follow-up is mandatory for recurrent AR and DSS, our approach was thought to be feasible for a young adult patient with DSS complicated with AR.

Keywords: discrete subaortic stenosis, aortic insufficiency, aortic valve repair

Case Report

A 33-year-old man was referred for evaluation of his heart murmur which had been detected at the age of 15. On physical examination, a Levine 3/6 systolic and diastolic murmur was noted at the right sternal border in the 3rd intercostal space. Transthoracic echocardiography showed subaortic stenosis with a peak pressure gradient of 66 mmHg through the left ventricular outflow tract (LVOT) and moderate aortic regurgitation with turbulent jet caused by an abnormal subvalvular structure (Fig. 1). Left ventricular dimensions in end-diastole and systole were 47 and 22 mm with an ejection fraction of 85%. No other cardiac anomalies were detected. Three-Dimensional transesophageal echocardiography demonstrated circumferential membranous tissue just beneath the aortic valve (Fig. 2), and the diagnosis of discrete subaortic stenosis (DSS) was made. Cardiac catheterization revealed a pressure gradient of 76 mmHg across the LVOT. Surgical intervention was planned for the significant LVOT obstruction. Cardiopulmonary bypass was established with ascending aortic and bicaval cannulation. After cardiopleic cardiac arrest, transverse aortotomy was made, and the LVOT was inspected through the aortic valve. White fibrous tissue was circumferentially attached to the subvalvular portion. The tissue was carefully excised (Fig. 3). Then, the aortic valve was inspected. All three cusps were slightly thickened, and the left coronary cusp seemed to be redundant. We performed aortic annular plication by placing 4–0 prolene mattress sutures at each commissure and the plication of the redundant left coronary tissue with a 6–0 prolene suture. The postoperative course was uneventful. On postoperative echocardiography, the peak gradient through LVOT decreased to 13 mmHg, and only trivial aortic regurgitation was noted. Histological examination of resected specimen showed irregular endocardial fibrous tissue proliferation. During the follow-up of 6 months, the patient has been well without deterioration of aortic valve competence.
Discussion

DSS is an uncommon form of LVOT obstruction in adulthood. There are only a few published reports on the occurrence of DSS in adults. Oliver et al. reported that DSS occurs in 6.5% of adult patients with congenital heart disease.\(^1\) The present case had no other congenital anomaly. Although the definitive pathogenesis for isolated DSS has not been established, it is thought to be an acquired fibrous ridge in the LVOT portion.\(^2\) His 18-year history of heart murmur implied that isolated DSS might be a very slow progressive disease; therefore, clinically non relevant in most cases.

The operative indication for DSS is proposed in
patients with peak pressure gradient over 50 mmHg at LVOTO. Surgical management consists of removal of the circumferential fibrous obstructive ridge with or without myectomy. In our case, LVOT obstruction was successfully relieved only by resecting the fibrous ridge. The peak pressure gradient dramatically decreased from 92 mmHg to 13 mmHg, postoperatively.

DSS frequently causes aortic valve incompetence especially in older patients. Etiology of AR has been reported, and these include (i) trauma to the aortic valve from the turbulent jet during left ventricular systole; (ii) tethering of the cusps to fibrous membrane; and (iii) development of infective endocarditis. Early surgical repair of DSS has been advocated to prevent development of significant AR. However, there is little information about the timing of surgery for existing AR. In our case, moderate degree of AR was noted without any deterioration of left ventricular function. Oliver et al. reported that the benefits of prophylactic early repair for AR should be questioned because observed AR was non-progressive disease. However, another group reported that AR was sometimes increased after relief of DSS, if the valve were damaged, or if residual LVOT obstruction were present. We believe that valve surgery is feasible in our patient because of his young age and relatively preserved leaflet structures enough to avoid valve replacement.

Durability of aortic valve repair for AR is still controversial, and recurrent DSS after operation is also documented with a reported incidence of 6% to 30%. Although careful follow-up is mandatory for the recurrence of AR and DSS, our case demonstrated that concomitant valve repair with relief of DSS might provide a better quality of life, avoiding the future deterioration of AR and implantation of a prosthetic heart valve.

References