

Case Report

Supracristal ventricular septal defect with severe right coronary cusp prolapse

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Abstract

The case of a 20 years old male, diagnosed as supracristal ventricular septal defect (VSD) for last 6 years is being presented. He came in emergency department with decompensated congestive cardiac failure. After initial stabilization, he underwent transthoracic echocardiogram which showed large supracristal VSD, severely prolapsing right coronary cusp, severe aortic regurgitation and severe pulmonary hypertension. Right heart catheterization was performed which documented reversible pulmonary vascular resistance after high flow oxygen inhalation. He underwent VSD repair, right coronary cusp was excised and aortic valve was replaced by mechanical prosthesis. Post operative recovery was uneventful. He was discharged home in one week.

Keywords: Ventricular septal defect, Aortic valve prosthesis, Cardiopulmonary bypass.

Introduction

Supracristal VSD, synonymous with subarterial, conal, infundibular and type I VSD is relatively common among Asians.¹ These lesions result from abnormalities in fusion of the embryonal conal septum. The association of prolapse of the right and noncoronary aortic cusps and aortic regurgitation (AR) has been well documented. Surgical closure of the defect is indicated in patient with associated progressive AR. Aortic valvoplasty can be considered in selected patients with aortic cusp prolapse and regurgitation in an attempt to avoid prosthetic aortic valve.²

Case Report

A 20 years old male presented to us with worsening exertional dyspnoea, orthopnoea and paroxysmal nocturnal dyspnoea for last one month. He was having exertional dyspnoea New York Heart Association (NYHA) class II equivalent for last 8 years. He was diagnosed to have Supracristal VSD with left to right shunt for last 6 years. He was advised surgical closure in view of his symptoms and Qp/Qs of >1.5 but he was reluctant for surgery despite having prior multiple admissions with congestive cardiac failure.

On arrival he was in decompensated cardiac failure with raised juglar venous pressure, pedal oedema, hepatomegaly, ascites and bilateral pulmonary crackles upto mid-zone. A grade 4/6 pansystolic murmur was audible at left

lower sternal edge and an early diastolic murmur of grade 3/6 intensity was appreciated at aortic area. His chest radiograph was consistent with pulmonary oedema. He made significant clinical improvement with initial management with diuretics and nitrates.

After initial stabilization he underwent trans-thoracic echocardiogram that showed dilated left ventricle with preserved systolic function, dilated and hypertrophied right ventricle with moderate systolic dysfunction, larger supracristal VSD with severe prolapse of right coronary cusp into right ventricle, severe aortic regurgitation, severe tricuspid regurgitation, dilated non-collapsing inferior vena cava with estimated pulmonary artery systolic pressure of 120 mm of Hg consistent with severe pulmonary hypertension (Figure-1).

In order to look for reversibility of pulmonary vascular resistance, right heart catheterization was performed. Right heart catheterization showed a pulmonary vascular resistance of 11 wood units which reduced to 6 wood units after high flow oxygen inhalation.

He was planned for surgery in view of, reversible pulmonary hypertension, large VSD, severe AR and congestive cardiac failure. High risk surgical consent was taken and he underwent surgery.

At operation right ventricle was severely enlarged, left ventricle and pulmonary artery were also enlarged. Aorto-



Figure-1: Transthoracic echocardiogram, parasternal long axis view showing severe prolapse of right coronary cusp into right ventricular outflow tract via ventricular septal defect.



Figure-2: Gross appearance of prolapsing right coronary cusp at surgery.

bicaval cannulation was secured and aortic valve was exposed through an oblique aortotomy. Right coronary cusp was large, redundant and was prolapsing through large supracristal VSD into right ventricular outflow tract (Figure-2). Aortic cusps were excised, aortic valve was replaced with mechanical prosthesis (St Jude 27mm) and VSD was repaired with D shape Dacron patch through transaortic approach.

Intraoperative transesophageal echocardiogram showed successful repair of VSD without any residual shunting and trivial aortic regurgitation post aortic valve replacement.

He made uneventful post operative recovery and was discharged on 7th post-operative day. He is doing well in follow up.

Discussion

The association of ventricular septal defect and aortic regurgitation was first described by Laubry and Pezzi in 1921.³ AR, balloon like dilatation of the right coronary sinus of Valsalva, prolapse of the right coronary cusp and rarely the left coronary cusp into the right ventricular infundibulum, all have been reported as complications of the supracristal ventricular septal defect.⁴

AR has been reported as an associated lesion in approximately 3-8% of patients with a VSD. However in 40% of patients with a VSD and AR, setpal defect is supracristal type.¹

A supracristal defect results in decreased support especially of the right aortic cusp which may then herniated through the defect into the outflow tract of the right ventricle.⁵ Associated AR was usually progressive in severity as prolonged insult to the valves causes permanent structural abnormalities. In a case series by Hallidie-Smith and colleagues, out of 29 patients with VSD and AR, 19 had

prolapsed cusp as a cause for aortic regurgitation and in majority of patients prolapsing cusp was right coronary cusp.⁶

This combination of VSD and AR has a poor medical prognosis and so with improving surgical techniques, total correction is being more widely advocated. However surgical correction is not always easy and success depends largely on a comprehensive knowledge of the anatomy and function of the aortic valve.⁷

Echocardiography is an invaluable tool in diagnosis and identifying type of VSD, presence and severity of associated AR, and presence and extent of aortic cusp prolapse. Cusp prolapse can be graded on a three-point scale by echocardiographic imaging: Mild (buckling of aortic cusp with minimal herniation, aortic cusp protrudes slightly into defect only during early systole), Moderate (prolapse of cusp with obvious herniation into the septal defect) and Severe (prolapse of cusp and its sinus through the defect into the right ventricular outflow tract during both systole and diastole). The severity of prolapse needs to be further confirmed during operation.⁸

The type of surgical management is largely dictated by the severity of the aortic regurgitation. If the latter is minimal it is unlikely that any special measures will be required to deal with the aortic valve. It is also important to take in account the degree of prolapse, shown by echocardiography.⁹ Cheung and colleagues had shown that in patients with mild to moderate degree of aortic cusp prolapse before operation, simple closure of the defect without aortic valvoplasty effectively reduces or halts the progression of AR. Closure of the defect alone is believed to prevent progression of AR by reducing the Venturi effect.¹⁰

In patients with moderate or severe aortic regurgitation the valve should be very carefully inspected in order to determine the exact cause of regurgitation. Some valves are capable of reconstruction or repair whereas others are so grossly deformed that replacement with a homograft or a mechanical prosthesis is required.

Our case is particularly interesting because our patient had large supracristal VSD with severely prolapsing right coronary cusp which is merely reaching distal part of right ventricular cavity and associated with severe AR. He had severe pulmonary artery hypertension which was documented to be reversible after high flow oxygen inhalation. He underwent successful repair of VSD along with resection of redundant, prolapsing right coronary cusp and aortic valve replacement with mechanical prosthesis. The case also highlights the importance of looking for reversibility of pulmonary hypertension which may be found in considerable number of patients making them amenable for surgical correction.

Acknowledgement

We are thankful to all the staff members of Tabba Heart Institute especially Miss Tadeeb Anwar for her co-operation, assistance and efforts.

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