CASE REPORT

Isolated supravalvular aortic stenosis with left ventricular diverticulum and cleft mitral valve: Surgical repair in adulthood

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Supravalvular aortic stenosis is an uncommon but well characterized congenital narrowing of the ascending aorta above the level of the coronary arteries. It can be a familial disorder, can occur sporadically, or can be associated with Williams syndrome. We are reporting a very rare presentation of supravalvular aortic stenosis with associated left ventricular diverticulum and cleft mitral valve. Repair consisted of resection of the ascending aorta, patch augmentation of the aortic root, and mitral valve repair. Follow-up echocardiography demonstrated normal mitral and aortic valve function and a postoperative three-dimensional computed tomographic scan showed a normal shape of the reconstructed ascending aorta.

Keywords: Brom's aortoplasty, Cleft mitral valve, Supravalvular aortic stenosis, Ventricular diverticulum

Introduction

Congenital supravalvular aortic stenosis (SVAS), either in the form of a discrete or a diffuse narrowing of the ascending aorta, is the least common type of left ventricular outflow obstruction. Peripheral pulmonary artery stenosis, coronary lesions, or abnormalities of the aortic valve leaflets may also be present in patients with SVAS. Calcification of the aortic annulus is uncommon. Moreover, survival beyond the fourth decade is rare because of the secondary effects of the disease on the left ventricle, the coronary circulation, and the aortic valve. Our case had a rare combination of SVAS with associated left ventricular diverticulum and cleft mitral valve.

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Case report

A 20-year-old female presented with progressive exertional dyspnea for one year. No facial dysmorphism, physical deformities, growth retardation or mental retardation were noted in this patient. Further evaluation for Williams syndrome was negative. Auscultation revealed a Grade 4/6 systolic ejection murmur. Transthoracic echocardiography demonstrated severe concentric left ventricular hypertrophy, and hourglass constriction above the coronary artery origin with peak gradients of 140 mmHg across the ascending aorta. The aortic valve appeared bicuspid with mild valvular incompetency, due to the presence of a very small and dysplastic left coronary cusp. Angiography (Fig. 1) showed a severe hourglass constriction of the proximal segment of the ascending aorta and a 1/4 aortic incompetence. With a pressure gradient of about 140 mmHg, the coronary ostia were enlarged and the right coronary and the left main coronary were dilated and tortuous, but the coronary vessels were free of any obstructive disease. Pulmonary vasculature was normal. Left ventricular angiogram showed large apical diverticulum with good contraction (Fig. 2). The patient had moderate mitral incompetence due to cleft mitral valve (Fig. 2).

Operative findings

In addition to significant SVAS (Fig. 3), the patient had rudimentary noncoronary cusps (Fig. 4). Cleft mitral valve was noted and it was responsible for moderate mitral regurgitation.

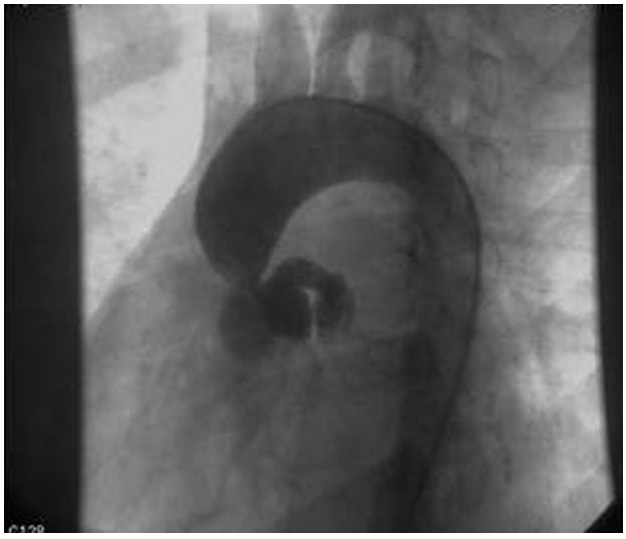

Figure 1. Preoperative aortic angiography showing hourglass constriction of ascending aorta, above the aortic sinus and origin of coronaries.

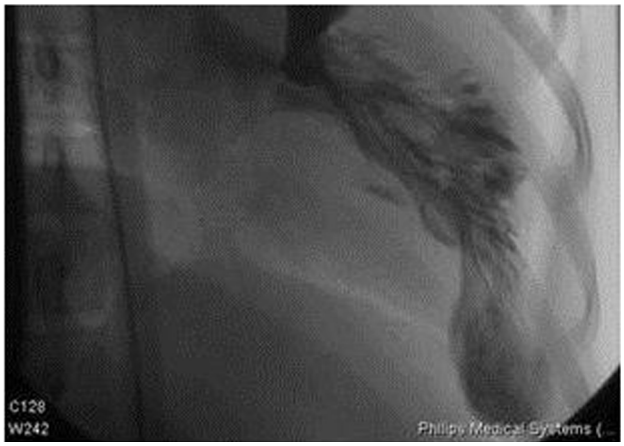

Figure 2. Preoperative left ventricular angiogram showing left ventricular diverticulum and mitral incompetence.

Operative technique

Brom's aortoplasty with mitral valve repair (cleft Anterior Mitral Leaflet (AML) repair) [1,2].

After median sternotomy, anatomy was confirmed and cardiopulmonary bypass established with high ascending aortic and bicaval cannulation. On moderate hypothermia, the aorta was cross clamped and the first dose of antegrade cold blood cardioplegia was given, followed by retrograde cardioplegia. The aorta was transected several millimeters distal to the area of maximal stenosis. First longitudinal incision was made in the center of the noncoronary sinus to facilitate exposure for the incisions into the right and left coronary sinuses and to identify coronary ostia. Direct ostial cardioplegia was given. Anatomy was confirmed. A second incision was made in the right coronary sinus to the left of the right coronary orifice. A third incision was made to the right of the left coronary orifice. Three separate appropriate sized shield shaped patches were sutured with 6-0 polypropylene accordingly into three sinuses. Aortic anastomosis was done with 6-0 running polypropylene sutures between the proximal and distal ascending aorta. The Left Atrium (LA) was opened and anatomy of the Left Ventricle (LV) diverticulum and mitral valve was inspected. A small muscular extension of the LV cavity was found (Fig. 5). Since it was only a muscular extension and was contracting with LV, it was decided not to repair the LV diverticulum. The mitral valve was inspected and a cleft was found in AML, which was closed with interrupted 6-0 polypropylene sutures. Competency of the mitral valve was tested with a saline probe and was found to be satisfactory. The aortic cross clamp was released after closing the LA incision

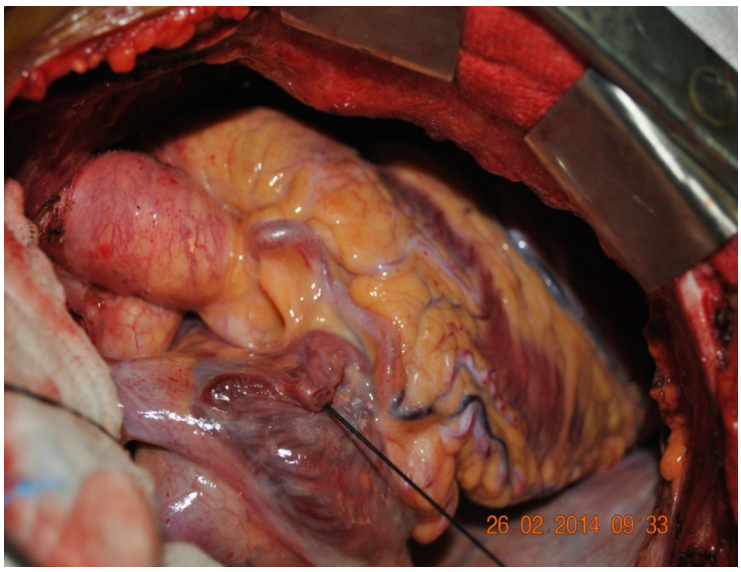

Figure 3. Intraoperative image showing constriction of the aorta above the coronary sinus.

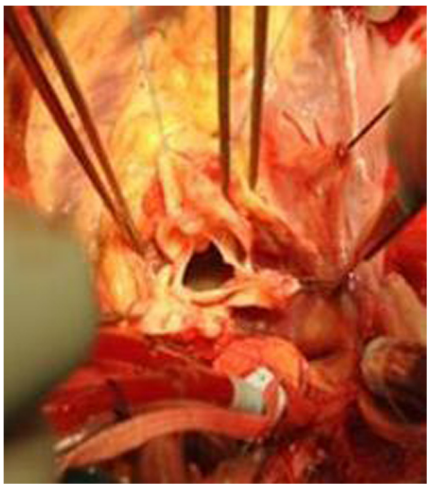

Figure 4. Intraoperative image showing rudimentary noncoronary aortic valve.

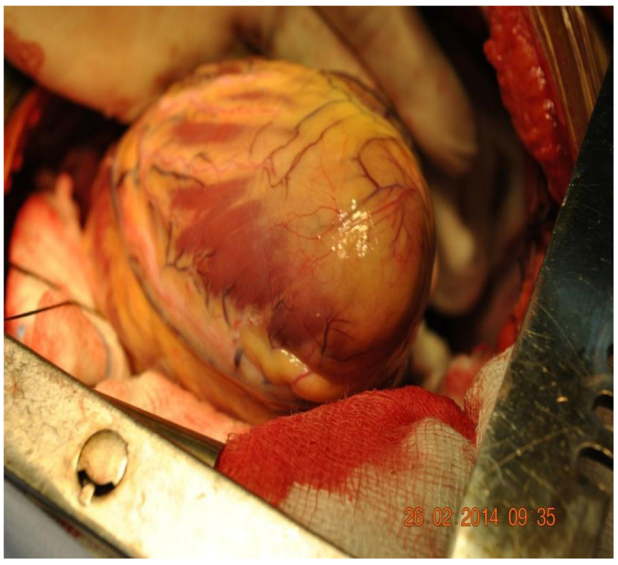

Figure 5. Left ventricle showing bulged out pouching in the apex, which was confirmed in a left ventricular angiogram and magnetic resonance imaging (Magnetic Resonance Imaging (MRI)) as diverticulum.

and thorough deairing. Intraoperative Trans Esophageal Echocardiography (TEE) showed competent aortic and mitral valves with mild aortic and mitral regurgitation. The total duration of cross clamping was 2 hours 18 minutes and cardiopulmonary bypass time was 2 hours 48 minutes. The patient was extubated the following day and was moved to a ward on Day 3. She had an uneventful in-hospital stay and was discharged with stable vitals on Day 10. Five months later, the patient underwent Computed Tomography (CT) imaging to assess the postsurgical anatomy, which showed a well reconstructed segment of aorta (Fig. 6A and B).

Discussion

Isolated SVAS associated with left ventricular diverticulum and cleft mitral valve is a very rare combination. Two types of diverticulum have been reported; the first one is a muscular type which is identified by the presence of contraction and the second one is a fibrous type where the diverticulum does not contract with the left ventricle. A higher incidence of thromboembolism was reported with the fibrous type. Muscular

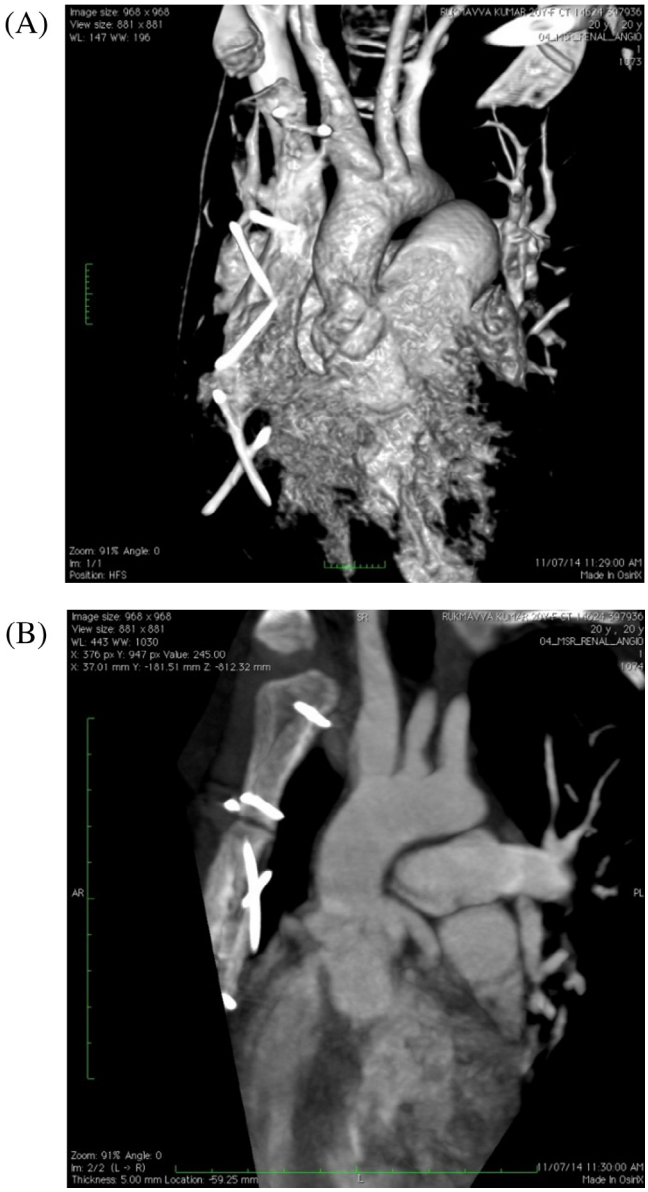

Figure 6. (A) Three-dimensional CT reconstruction image postoperatively showing normal supravalvular segment of aorta; (B) CT image postoperatively showing normal supravalvular segment of aorta.

diverticulum is frequently associated with other congenital anomalies of the abdominal wall (involving the diaphragm), sternum, pericardium (Centrell syndrome), and heart itself, usually ventricular septal defect. Left ventricular diverticulum with congenital pulmonic stenosis has been reported [3]. Sudden cardiac death due to ventricular fibrillation is a rare complication of left ventricular diverticulum [4].

Structural changes of the coronary vessels are either related to a well-known elastin gene defect [5] or to the effects of chronic elevated prestenotic systolic pressure. These promote dilatation and tortuosity of coronary vessels or premature arteriosclerosis and may cause impairment of coronary circulation [6]. SVAS with other associated anomalies reduces the overall survival, especially when there is a left ventricular diverticulum

Mitral cleft with incompetence of valve is rarely reported in these patients and may represent a surgical challenge. To enlarge the aortic root, two techniques have been described, which both require an extension of the conventional aortotomy incision through the commissure between the left and noncoronary sinuses [7], or through the midpoint of the noncoronary sinus across the

mitral valve [8], and the insertion of a teardrop shaped pericardial patch cut to the required dimensions. In our patient, we chose the first technique, which moreover seemed a safer choice over composite root replacement because it avoided manipulation of the enlarged and fragile coronary ostia [9]. Left ventricular diverticulum has not been reported in association with SVAS. Presence of left ventricular diverticulum might further reduce the long term survival.

Conclusion

Rare manifestations of congenital heart disease in adult patients impose great clinical and diagnostic challenge. Our case of SVAS with cleft mitral valve and LV diverticulum posed a big challenge of surgical management. A thorough understanding of the anatomy is recommended to plan optimal surgical strategy. In our case, we decided to correct the stenosis and cleft valve using Brom's aortoplasty with mitral valve repair. We did not repair the ventricular diverticulum which in our case showed reduction in its size.

Disclosures and freedom of investigation

Our study has no financial and non-financial relationships to disclose. We had obtained prior consent for complete evaluation of the study participant and acquired necessary consents for publication work.

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