Case Reports

Bicuspid aortic valve with severe stenosis and ruptured sinus of Valsalva aneurysm to left ventricle

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ABSTRACT

Bicuspid aortic valve (BAV) is a common congenital cardiac anomaly, and rarely, it is associated with sinus of Valsalva aneurysms (SOVAs). And very infrequently, these SOVAs rupture into left side of heart. We hereby report a case of 12-year-old male with BAV with severe aortic stenosis with large SOVA that ruptured into the left side of the heart. The anatomy was delineated with multimodality imaging; initially with two-dimensional transthoracic echocardiography (TTE), and later on with three-dimensional TTE and with multi detector computed tomography. Operative interventions were planned for the patient.

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1. Introduction

Bicuspid aortic valve (BAV) is the most common congenital cardiac anomaly. It is associated with various other congenital anomalies, and sometimes associated with sinus of Valsalva aneurysms (SOVAs). These SOVAs may rupture into the right side of heart, producing a left to right shunt and volume overload of different chambers depending upon the site of rupture; and very rarely, they rupture into left side of heart, producing left sided volume overload. Prompt recognition of this fatal complication is necessary for further management. We are hereby, describing a case of young male with BAV with severe aortic stenosis with large SOVA from right coronary sinus, going up to the left coronary sinus and rupturing into the left ventricle.

2. Case

A 12-year-old male child presented in our department with dyspnea on exertion for the last 2 years. At presentation, he was in NYHA functional class III. General physical examination revealed a low volume pulse at a rate of 100 per minute and blood pressure of 100/76 mmHg. Cardiovascular examination was suggestive of sustained apex beat in 5th intercostal

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space, single S2 and grade IV/VI mid systolic murmur at aortic area and a faint early diastolic murmur at third left parasternal area. Chest X-ray showed normal cardiac and aortic root size, but electrocardiogram showed left ventricular hypertrophy (LVH). Evaluation with two-dimensional trans-thoracic echocardiography (2D-TTE) in parasternal long axis (PLAX) view showed LVH and thickened aortic valve with eccentric opening (Fig. 1A and C, Supplementary Movie 1). A SOVA was seen, facing the right coronary sinus (RCS), which was also communicating with left ventricular outflow tract (LVOT) through a small rent. Color Doppler in PLAX view during systole showed two forward jets through LVOT, one through

Fig. 1 – Two-dimensional trans-thoracic echocardiography (2D-TTE) with color Doppler and color M mode. Parasternal long axis (PLAX) views in systole (A and B) showing two forward turbulent jets from left ventricular outflow tract (LVOT), one through the bicuspid aortic valve (BAV) into aorta and another through the rent into the sinus of Valsalva aneurysm (SOVA). Slightly dilated coronary sinus was also seen. PLAX views in diastole (C and D) showing two regurgitant jets into LV, one as trivial aortic regurgitation and another from aneurysm to LV through the rent (arrow in A and C). Color M mode through aorta and aneurysm (E) showing two different turbulent jets through them. 2D-TTE in parasternal short axis view (F) showing the BAV with SOVA and its communication with the aortic root (arrowhead in A, C and F showing the opening of SOVA into aorta). AO, aorta; AN, sinus of Valsalva aneurysm (SOVA); LA, left atrium; LV, left ventricle; RA, right atrium.
stenotic aortic valve and another through the rent into the aneurysm (Fig. 1B and E, Supplementary Movie 2). During diastole, two regurgitant jets were seen, one from aneurysm to LVOT through the rent and another of valvular aortic regurgitation (Fig. 1D, Supplementary Movie 2). 2D-TTE in parasternal short axis view showed concentric LVH with normal LV systolic functions with BAV, and the SOVA overlying RCS (Fig. 1F, Supplementary Movies 3 and 4). Continuous Doppler through aortic valve demonstrated severe aortic stenosis. Three-dimensional (3D)-TTE with color Doppler showed the thick, stenotic BAV and the ruptured SOVA and two systolic turbulent jets through LVOT (Fig. 2A–D, Supplementary Movies 5 and 6). 3D-TTE clearly demonstrated the proximity of SOVA opening to the aortic cusps, as this was partially closed by the aortic cusp during ventricular systole (Fig. 2A, Supplementary Movie 7). The 64-slice computed tomography (CT) angiography showed the large intra-pericardial SOVA, abutting RCS and its anterior extension up to the origin of left coronary artery (LCA) and its communication with LVOT through the small rent (Fig. 2A–C). Its size was approximately 25 mm × 40 mm and neck was 20 mm wide. 3D-volume rendered CT image showed the extension of aneurysm across the two coronary sinuses and superior displacement of LCA by the aneurysm, without causing any flow impediment (Fig. 3D). After this complete evaluation, surgical correction was planned; but patient had sudden demise while waiting for the same.

Supplementary material related to this article can be found, in the online version, at doi:10.1016/j.ihj.2015.07.045.

3. Discussion

BAV is the most common congenital cardiac anomaly, comprising 1–2% of adult population, whereas SOVAs comprise 1% of congenital anomalies of the heart. BAVs are rarely associated with SOVAs.1,2 Although, frequently congenital absence of the tissue in the aortic wall is the cause of SOVAs, they are also caused by infection or degenerative disease of aortic wall.3 They usually remain asymptomatic until ruptured.4 Unruptured, SOVAs may compress coronary arteries, or can obstruct left or right ventricular outflow tracts. Rupture of
the SOVAs usually occur in right sided cardiac chambers and produce a state of left to right shunt and volume overload of different chambers of heart, which depend on the site of rupture. Rarely, they rupture into left sided cardiac chambers. In our case, the SOVA along with the BAV is most probably congenital in origin, and later on the SOVA ruptured into the LV. Both BAV and ruptured SOVA were diagnosed on 2D-TTE, and color Doppler demonstrated its rupture into the LV. These findings were confirmed with 3D-TTE and CT scan. Although the diagnosis was made on 2D-TTE, 3D-TTE demonstrated the spatial orientation of aneurysm around LVOT and aortic root and its wide neck just above the aortic valve and its proximity with the same. These provide the 3D anatomy of the structures, which can help surgeons in planning the operative steps. To the best of our knowledge, this is one of the very rare presentation of BAV with severe aortic stenosis with SOVA involving the area of two sinuses and rupturing into the left side of heart.

Conflicts of interest

The authors have none to declare.

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