

Bicuspid Aortic Valve: An Unusual Cause of Aneurysm of Left Coronary Sinus of Valsalva

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Abstract

Bicuspid aortic valve is traditionally considered an innocuous congenital anomaly. Due to a better and widespread availability of non-invasive imaging techniques, it has come to the fore that 30% of these cases develop complications, viz., valve abnormality (aortic regurgitation and stenosis), and aneurysm of aortic root and ascending aorta. Sinus of Valsalva aneurysm is an uncommon complication of bicuspid aortic valve and more so those arising from the left coronary sinus are the rarest. These complications generally occur in the third or fourth decade of life. We present a case of the left sinus of Valsalva aneurysm in conjunction with bicuspid aortic valve and ascending aorta aneurysm at a very young age in a girl in her early adolescence. This case is to remind the paediatricians about the not so “innocuous image”, but the serious implications of the bicuspid aortic valve and to regularly follow these cases for early diagnosis of potential complications so as to prevent catastrophic outcomes.

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Keywords • Bicuspid aortic valve • Sinus of valsalva • Aortic aneurysm

What's Known

- Bicuspid aortic valve (BAV) is the most common congenital cardiac malformation affecting 1% to 2% of the general population. It is associated with complications including dilatation of aortic root. Sinus of Valsalva aneurysm is an uncommon complication of bicuspid aortic valve and more so those arising from left coronary sinus are the rarest (2%). These complications generally occur in the 3rd or 4th decade of life.

What's New

- Here we present a case of bicuspid aortic valve with such rare complications viz, left sinus of Valsalva aneurysm and ascending aorta aneurysm in a young adolescent girl.

Introduction

Aneurysms of the sinuses of Valsalva (ASV) are thin-walled outpouchings, most commonly involving the right or non-coronary sinuses. An unruptured aneurysm of the left sinus of Valsalva is extremely rare (1%). It is usually associated with other congenital malformations of the heart. We describe a 14-year-old girl having a combination of a bicuspid aortic valve and an aneurysm of the left sinus of Valsalva with severe aortic stenosis, moderate regurgitation, and an aneurysm of the ascending aorta.

Case Presentation

A 14-year-old girl presented with complaints of progressive exertional dyspnoea since 2 months, along with swelling of both legs and face for 5 days that was relieved on taking medications from a local doctor. There was no history of fever, sore throat, joint pain, orthopnoea, palpitations, chest pain, haemoptysis, syncope, oliguria, rashes, and weight loss. Past medical history and family history were insignificant.

She was of normal height and build, with normal features, and normal vision with a pulse rate 90/min, respiratory rate 30/min, and blood pressure 110/70. There was no pallor, icterus, easy bruisability, pedal edema, lymphadenopathy,

subcutaneous nodule, hyperextension, or laxity of joints. Cardiac examination revealed systolic murmur in the aortic area radiating to carotids and early diastolic murmur in the third left intercostal area. Per abdomen examination revealed hepatomegaly (liver 3 cm below costal margins, soft and non-tender). There were fine basal creps. Nervous system examination was unremarkable. Investigations revealed normal blood counts and ESR. Serum electrolytes, kidney, and liver functional tests were normal. ASO, anti-DNase, ANA, and RA factor were normal. Chest roentgenogram was normal. Electrocardiography revealed left ventricular hypertrophy. Echocardiography showed bicuspid aortic valve with fusion of the right and non-coronary cusp, unruptured aneurysm of left coronary sinus of Valsalva bulging into left atrium, gross dilation of aortic root (aortic annulus: 2.03 cm, aortic sinus: 3.2 cm, Z-score: +3.3) and ascending aorta (4.7cm) (figure 1A and 1B), severe aortic stenosis (aortic valve area: 0.6 cm²), moderate aortic regurgitation, with no other valvular lesion, and a normal ejection fraction. There was no coarctation and pulmonary flow was normal. MR angiography (figure 2A and B) confirmed the presence of aneurysm, which was

not compressing the left coronary artery. It also revealed right coronary to be the dominant coronary artery, common origin of left common carotid and right innominate artery and ascending aortic aneurysm extending until the left subclavian artery. The child is planned for Bentall procedure. Consent from the child's guardians was taken to publish this article.

Discussion

Bicuspid aortic valve (BAV) is the most common congenital cardiac malformation affecting 1% to 2% of the general population.¹ In one-third of patients, BAV is associated with significant valvular disease (aortic stenosis and regurgitation), aortic root, ascending aorta, and arch complications (dilatation and/or aneurysm of the aortic root, ascending aorta). In addition, it is associated with several other congenital abnormalities of the heart, including aortic coarctation, hypoplastic left ventricle, atrial and ventricular septal defect (VSD), and reversal of dominance of coronary arteries.²

Congenital or acquired ASVs are rare anomalies and account for 0.14% of all open-heart surgical procedures. Right coronary sinus aneurysm is most common (65-85%) followed by non-coronary sinus (10-30%) and left coronary sinus (1-5%).³ With a male predominance, these aneurysms are usually detected in the third or fourth decade of life.⁴ However, our case was a young 14-year-old female patient with the involvement of the left coronary sinus.

Congenital ASVs (Marfan syndrome, Ehler-Danlos syndrome) account for 0.1-3.5% of all congenital heart defects and are most often caused by the absence of muscular and elastic tissue in the aortic wall behind the sinus of Valsalva. It might be associated with other heart defects like VSD (30-60%), aortic regurgitation (20-30%), BAV (10%), and aortic stenosis (6.5%). Acquired ASV is caused by infections (bacterial endocarditis, tuberculosis), connective tissue disease, or trauma.⁴

Unruptured ASV is less frequently reported because in most instances they are asymptomatic. Rarely can they cause complications such as conduction system abnormalities and malignant arrhythmias (complete heart block, ventricular tachycardia) attributable to extension into the interventricular septum, thromboembolism originating in the aneurysm sac, right ventricular outflow tract obstruction, and myocardial ischemia consequent to coronary compression. Infective endocarditis is a dreaded complication.⁴

Kieffer and Winchell in their review of 78 cases of ASVs reported 24% to be unruptured.⁵ Reports

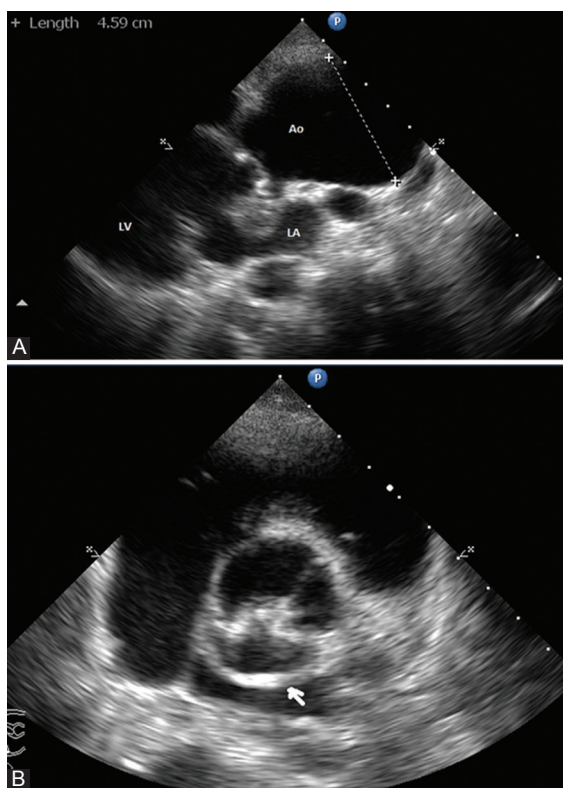


Figure 1: (A) Echocardiography-parasternal long axis view showing dilated aortic sinus and ascending aorta (LV: Left ventricle, LA: Left atrium, Ao: Aorta). (B) Echocardiography-short axis view showing aneurysm of left coronary cusp of sinus of Valsalva (arrow) impinging into the left atrium.

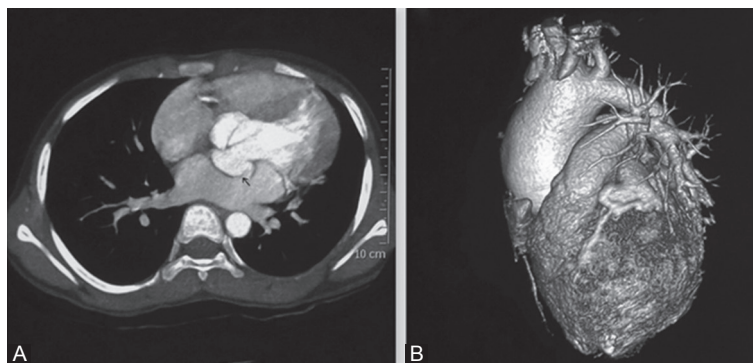


Figure 2: (A) Cardiac MRI depicting aneurysm of sinus of Valsalva (arrow) impinging into left atrium. (B) Cardiac MRI showing dilated ascending aorta till the left subclavian artery.

concerning unruptured, isolated ASV associated with BAV are rare. Moustafa et al.⁶ found that ASV was associated with BAV in only 9% of cases. In his landmark study, most of the ASV were unruptured (66%) and originated from the left coronary sinus in 7% of the cases. In contrast, Vural et al.⁷ found BAV in only 2% of ASV and unruptured SVA constituted 36% of the cases of which left coronary sinus aneurysm constituted only 15%. Our patient had unruptured, yet symptomatic left ASV and presented on account of left ventricular failure due to severe AS and moderate AR.

The most dramatic complication of ASV is abrupt rupture, either intracardiac or extracardiac presenting as chest pain, heart failure, or a loud continuous murmur. Rupture into the pericardium causes cardiac tamponade and death.⁴

Many theories have been postulated about the cause of aortopathy in BAV. Altered hemodynamics due to aortic stenosis and regurgitation and also defects in aortic media like elastin fragmentation, decreased fibrillin, matrix disruption, loss of smooth muscle and increase in collagen have been thought to be the cause. In addition, certain gene mutations (GATA5, NOTCH1, ACTA2) are associated with BAV defect, leaflet calcification and aortic dilation.

Keane et al. found that aortic root dilatation occurs irrespective of the presence and severity of aortic stenosis in BAV.⁸ Wall shear stress overload in the ascending aorta generated due to flow patterns through BAV even in the absence of stenosis or regurgitation initiates cellular signalling cascades resulting in increased expression of MMPs and growth factors leading to matrix disruption and vascular smooth muscle cell loss and decreased fibrillin; thus leading to aortopathy.⁸

Crawford and Roldan observed that aortic root dilatation is common in AS irrespective of severity. Yasuda et al. reported the presence of aortic dilatation in both tricuspid and bicuspid aortic

valve, however, the prevalence and severity of aortic dilatation were more with BAV.⁸ Our case had bicuspid aortic valve with severe stenosis. The presence of both bicuspid valve per se and aortic stenosis could have contributed to the development of aortic dilatation in the index case.

Regurgitant BAVs have more aortic root dilatation due to higher stroke volumes. Both paediatric and adult studies have demonstrated early and significant faster aortic dilatation with BAV versus TAV (mean: 49 vs. 61-64 years old).⁷

Fusion of the right-coronary and left-coronary leaflets (antero-posteriorly oriented aortic valves) is associated with moderate to severe aortic stenosis while fusion of the right-coronary and non-coronary leaflets (right-left orientation) were more likely to have moderate aortic regurgitation.⁹ Stenotic lesions are more common than regurgitant.

Mid ascending aortic dilatation is common with older age, whereas aortic root dilatation is correlated with younger age and male sex; 60% do not have any degree of aortic dilatation.⁸

Our case was a very young female and had aneurysm of both the aortic root and ascending aorta. She had fusion of the right and the non-coronary cusp but had a combination of severe aortic stenosis and moderate aortic regurgitation.

The rate of growth of ascending aortas with BAV ranges from 0.2 to 1.9 mm per year with larger aortas having faster expansion rates. Prevalence increases with age, beginning in childhood and reaching 90% by 80 years of age.⁸ BAV has a 6.14% lifetime risk of aortic dissection, which is 9-fold higher than the general population with aortic diameter being its significant predictor. Once the ascending aorta reaches 6 cm, the annual rates of rupture, dissection, and aorta-related death are 3.6%, 3.7%, and 10.8%, respectively and the cumulative rate of any of those events is 14.1%.

Annual monitoring by transthoracic echocardiography (TTE) should be done for

valve function as well as the diameter of the aortic root and ascending aorta. If the aortic root or ascending aorta is 4.0 cm on TTE, a CT-scan or MRI should be performed to ascertain the findings as well as screen for coarctation.

The American College of Cardiology and American Heart Association guidelines¹⁰ recommend that in patients with BAV and ascending aortic aneurysms exceeding 4.5 cm, simultaneous aortic replacement should be done at the time of aortic valve replacement. Surgery to repair the aortic root or replace the ascending aorta is indicated in patients with BAVs if the diameter of the aortic root or ascending aorta is >5.0 cm, or if the rate of increase in diameter is 0.5 cm/year or more. A lower threshold value for intervention should be kept for patients of small stature of either gender.

Beta-blocker treatment is suggested for a patient with BAV and ascending aortic enlargement >4.0 cm (in the absence of severe aortic regurgitation). There is no evidence to suggest that ACE inhibitors are better.

Conclusion

BAV is very common, but not as innocuous as it has been traditionally thought of. It is the biggest cause of aortic valve as well as aortic root surgery. Paediatricians should take due cognition of this fact and diligently follow these lesions for early diagnosis and prevention of its complications.

Conflict of Interest: None declared.

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