A tetrad of bicuspid aortic valve association: A single-stage repair

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ABSTRACT

We report a 27 years old male who presented with a combination of both congenital and acquired cardiac defects. This syndrome complex includes congenital bicuspid aortic valve, Seller's grade II aortic regurgitation, juxta-subclavian coarctation, stenosis of ostium of left subclavian artery and ruptured sinus of Valsalva aneurysm without any evidence of infective endocarditis. This type of constellation is extremely rare. Neither coarctation of aorta with left subclavian artery stenosis nor the rupture of sinus Valsalva had a favorable pathology for percutaneus intervention. Taking account into morbidity associated with repeated surgery and anesthesia patient underwent a single stage surgical repair of both the defects by two surgical incisions. The approaches include median sternotomy for rupture of sinus of Valsalva and lateral thoracotomy for coarctation with left subclavian artery stenosis. The surgery was uneventful. After three months follow up echocardiography showed mild residual gradient across the repaired coarctation segment, mild aortic regurgitation and no residual left to right shunt. This patient is under follow up. This is an extremely rare case of single stage successful repair of coarctation and rupture of sinus of Valsalva associated with congenital bicuspid aortic valve.

Key words: Bicuspid aortic valve, juxta subclavian coarctation, rupture of sinus of valsalva, single stage repair

INTRODUCTION

When coarctation of aorta is associated with one or more congenital or acquired cardiac defects, surgery is the first option[1,2] though there are no standard management guidelines for these complex and challenging pathological conditions. Still controversy exists not only on which lesion should be corrected first, but also upon the type and timing of the procedure. Surgery can be a one- or two-staged procedure. The one-stage approach can be accomplished through a single or two incisions and the two-stage approach implicates two operations performed through median sternotomy and posterior- lateral thoracotomy respectively with a gap of weeks to months. Both of these strategies have their merits and demerits. The option of choosing one of these procedures depends upon pathology and experience of concerned managing team.

Congenital bicuspid aortic valve associated with aortic regurgitation, juxta subclavian coarctation, left subclavian artery ostial stenosis and ruptured sinus of Valsalva aneurysm is extremely rare.[3-5] The timing and sequence of surgical and/or interventional repair of this type case is controversial. We present a case of a 27-year-old male who was admitted to our department because of severe acute congestive heart failure and signs of ruptured aneurysm of the right sinus of Valsalva into the right ventricular out flow tract. Because the juxta-subclavian coarctation was not suitable for percutaneus balloon angiplasty and stenting, surgical repair of the ruptured sinus Valsalva aneurysm, coarctation repair and repair of left subclavian artery were performed in a single stage.

Access this article online	
Quick Response Code:	Website: www.jcdronline.com
	DOI: 10.4103/0975-3583.95371

CASE REPORT

A 27 -year-old man presented with congestive heart failure of NYHA class II for a period of 10 days. He had blood pressure of 150/40-0mmHg in both the upper limbs, very weak femoral pulses, grade IV superficial continuous murmur in the left parasternal border. Twelve lead surface electrocardiograms showed volume overload pattern of left ventricular hypertrophy. Chest X-ray showed cardiothoracic ratio of 65% with pulmonary congestion [Figure 1]. We used IE-33 PHILLIPS machine for transthoracic and transesophagial echocardiography. There was congenital bicuspid aortic valve [Figure 2], posterior cusp prolapse and moderate aortic valve regurgitation. The aortic valve regurgitation color doppler jet width was 40% of left ventricular outflow tract width and doppler vena contracta width was 4mm. There was rupture of sinus Valsalva aneurysm of post cusp at 9´-10'o clock position into right ventricular out flow tract of size 1.3cm at aortic end and 4mm at ventricular end with aorta to right ventricular gradient 70/28mm Hg [Figures 3 and 5]. The sinus Valsalva aneurysm was quite close to right ventricular in flow tract. Gradient across juxta- subclavian coarctation measured 60mm Hg. Biventricular function was good. Cardiac catheterization reveled oxygen saturation step up of 17% from right atrium to pulmonary artery. QP (pulmonary blood flow)/QS (systemic blood flow) ratio was 2.3:1. Aortogram confirmed the echocardiographic findings such as Seller's grade II aortic valve regurgitation, juxta -subclavian coarctation of aorta [Figure 4] with pull back gradient across coarctation of 70 mmHg and rupture of sinus Valsalva aneurysm from posterior aortic cusp into right ventricular outflow tract. There was 40-50% narrowing of left subclavian artery ostium. Neither the coarctation with significant left subclavian artery stenosis nor the rupture of sinus of Valsalva opening very close to right atrio-ventricular valve was favorable for percutaneous intervention.

It was decided to close all the three defects in a single stage repair. Under general anesthesia and on pump the rupture of sinus Valsalva was repaired by 1.5 cm Dacron patch through median sternotomy. Then coarctation and left subclavian artery repair was done off pump through lateral thoracotomy. Total procedure duration was 4 h. Recovery from anesthesia was prompt. Patient was off ventilator after 8 h. ICCU stay was uneventful. On day three, patient was shifted to step down care unit. Patient was successfully discharged on day 10. At three-month follow up, patient was found to be asymptomatic with echocardiography showing mild aortic valve regurgitation and residual coarctation gradient of 20 mmHg.

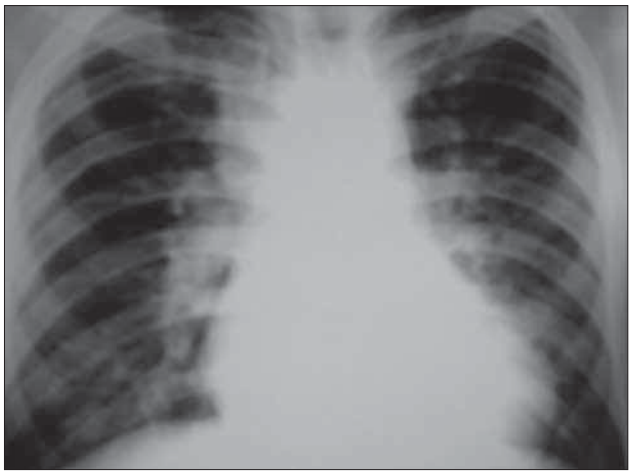

Figure 1: X-ray of chest revealing cardiomegally, pulmonary plethora and pulmonary venous hypertension

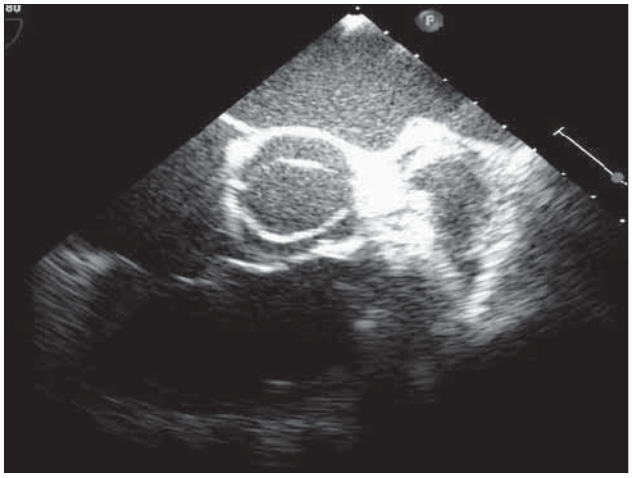

Figure 2: Transesophagial echocardiograghy at 40° showing congenital doming bicuspid aortic valve

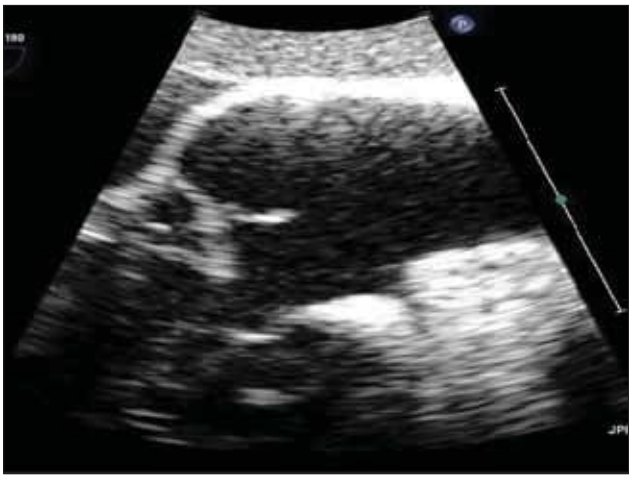

Figure 3: Transesophagial echocardiography at 95° showing rupture of sinus Valsalva with classical windsock look into right ventricular outflow tract

DISCUSSION

Ruptured sinus Valsalva aneurysm is associated with a plethora of congenital cardiac and extra cardiac defects.[3-5]

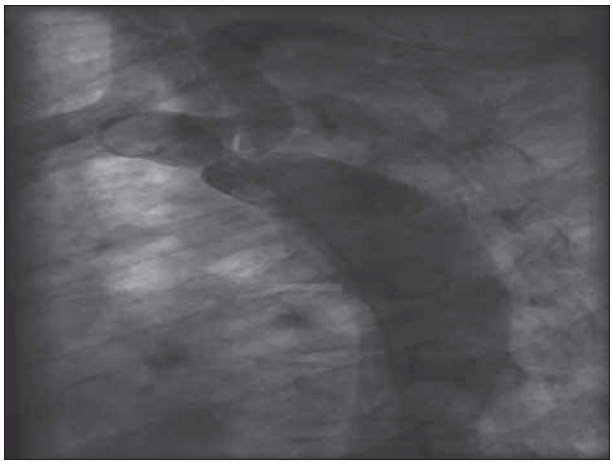

Figure 4: Aoatogram in LAO-cranial view showing ostioproximal significant narrowing of left subclavian artery with juxta-subclavian coarctation aorta with significant stenosis

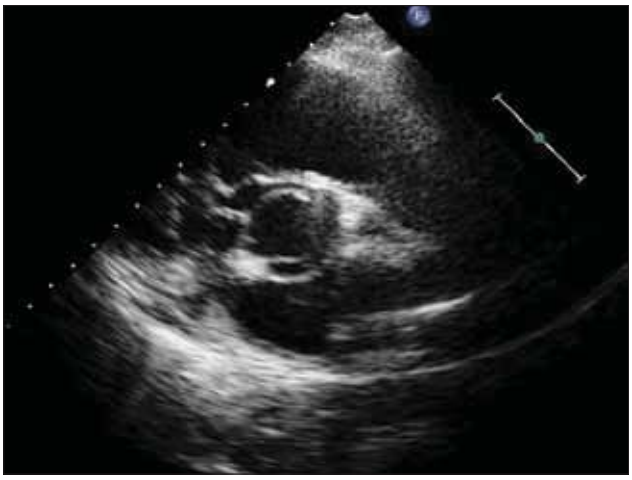

Figure 5: Transesophagial echocardiography at 40° showing congenital doming bicuspid aortic valve associated with classical windsock appearance of rupture of sinus of valve of post cusp at 9'o clock position

The incidence of aortic regurgitation is 33.6%, bicuspid aortic valve 10-20%[3,4] and coarctation of aorta 4%[3] associated with ruptured sinus Valsalva aneurysm. The coexistence of ruptured sinus Valsalva aneurysm and aortic coarctation is extremely rare.[3,6-8]

Various options are available for these complex pathological conditions. These are one stage repair, two stage surgical repair and staged hybrid procedure depending upon the clinical profile, complexity of anatomical defects and available expertise. In the one-stage repair, simultaneous correction of all the defects using two separate incisions through a median sternotomy and lateral thoracotomy are performed with success, though it is quite challenging.[9,10] As our case one-stage repair has the following advantages, such as economical, lesser morbidity and mortality of repeated surgery and anesthesia, better hemodynamic stability as there is no risk of increase or decrease in afterload because both defects are addressed at the same time, no extra pump time is required as coarctation repair is done off pump. However, a better clinical profile like good biventricular function is a favorable parameter for one-stage repair. Therefore, we opted for one-stage repair of all three pathology for the following reasons:

1. Good biventricular function.
2. The coarctation pathology was not suitable for percutaneous balloon angioplasty.
3. One time anesthesia.
4. Left subclavian artery ostial repair was essential.
5. The sinus of Valsalva aneurysm was associated with bicuspid aortic valve and was very close to tricuspid valve. Hence device closure was not a favorable option for both the defects. Review of literature revealed that left ventricular systolic dysfunction or biventricular dysfunction significantly increases mortality and morbidity if both coarctation and ruptured sinus of Valsalva are treated in the same sitting.

The followings are the explanations given for this:

1. Earlier repair rupture of sinus Valsalva repair could have led to increased after load, therefore leading to worsening of the existing heart failure.
2. First repair of coarctation could have decreased peripheral resistance significantly which would have aggravated heart failure due to myocardial ischemia due to decreased perfusion as a result peripheral run off and increase in left to right shunt.[12]
3. Prolonged procedural time, anesthesia and pump time in face of severe biventricular dysfunction.

Hence, in cases with severe left ventricular or biventricular dysfunction, two-stage surgical repair or two-stage hybrid procedure is a better option. Then two-stage repair can be performed through a combination of lateral thoracotomy and median sternotomy.[6,11] Hybrid approach is a better option for favorable pathological defects.[12] The non-surgical treatment of the aortic coarctation proved to offer a significant advantage, converting a complex and risky surgical procedure into one of the common practice with favorable pathology.[12] Surgery is the mainstay of treatment of ruptured sinus of Valsalva aneurysm, though there are several reports of transcatheter closure.[8]

CONCLUSION

Congenital bicuspid aortic valve is associated with a plethora of both congenital and acquired cardiac and extra cardiac defects. Hence management of these cases is always controversial. To avoid the morbidity associated

with repeated open heart surgery, we have managed this complex case of congenital bicuspid aortic valve, moderate aortic regurgitation, severe juxta-subclavian coarctation of aorta, left subclavian artery ostial stenosis and rupture of sinus Valsalva aneurysm using a single-stage surgical procedure with success. However, similar cases with significant left ventricular or biventricular dysfunction should be better managed by staged hybrid or staged surgical procedure.

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How to cite this article: Barik R, Patnaik AN, Mishra RC, Kumari NR, Gulati AS. A tetrad of bicuspid aortic valve association: A single-stage repair. J Cardiovasc Dis Res 2012;3:143-6.
Source of Support: Nil, **Conflict of Interest:** None declared.