WCC 2017-C-109: CONGENITAL QUADRICUSPID AORTIC VALVE ASSOCIATED WITH SEVERE AORTIC REGURGITATION: A RARE CASE REPORT

Pavaneel Bhandary
Palanki Surya Satyagopal, Muppiri Vijay Kumar, RavinuthalaVenkata Kumar

ABSTRACT:

Quadricuspid aortic valve is a very rare congenital valvular anomaly. Most of these cases present with aortic insufficiency. We present a 38 year old male patient with aquadricuspid aortic valve with severe aortic regurgitation.

INTRODUCTION:

Congenital Quadricuspid aortic valve is very rare among all the congenital valvular abnormalities. Aortic regurgitation is common with this anomaly due to unknown reasons. We present a 38 year old male patient with aquadricuspid aortic valve with severe aortic regurgitation and underwent aortic valve replacement.

CASE REPORT:

A 39 year old male presented with shortness of breath NYHA class III since 4 months. He is known diabetic and hypertensive since 5 years. He is on antidepressants for treatment of depression since 5 years. He is using Allopurinol for the treatment of hyperuricemia since 2 years. On clinical examination, he has hyperdynamic apex and grades 4 diastolic murmurs along the left sternal border. Chest x-ray revealed cardiomegaly with left ventricular apex. Electrocardiogram indicates left ventricular volume overload. Transthoracic echocardiograph in short axis view showed aortic valve with four cusps (Fig1a).

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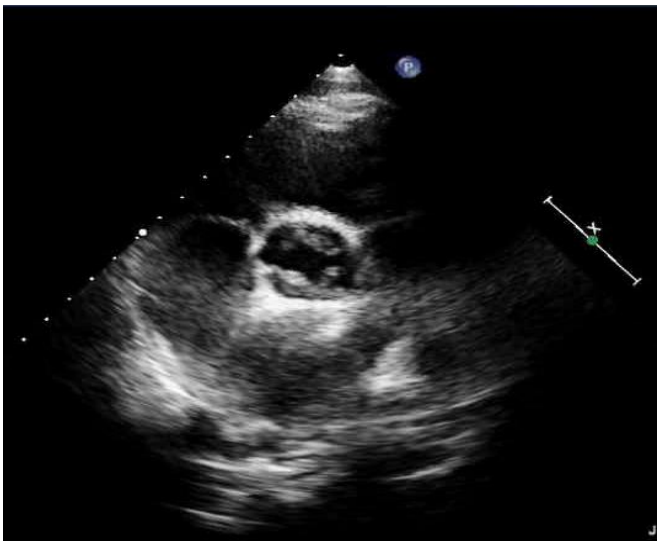
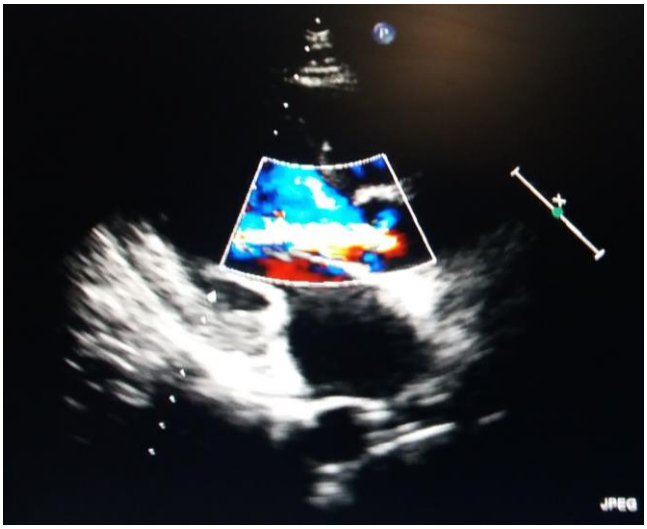
Pavaneel Bhandary [1] , Palanki Surya Satyagopal [2], Muppiri Vijay Kumar [3] , Ravinuthala Venkata Kumar [4]
[1] Senior Resident, Department of Cardiothoracic surgery, NIMS, India
[2]Assisstant Professor, Department of Cardiothoracic surgery, NIMS.
[3]Associate. Professor, Department of Cardiothoracic surgery, NIMS.
[4] Professor &HOD, Department of Cardiothoracic surgery, NIMS.
Corresponding author: Pavaneel Bhandary
Email: pavaneel@gmail.com

Fig 1a: Preoperative echocardiography showing quadricuspid aortic valve with noncoapting leaflets.



There is holodiastolic reversal of flow and severe aortic regurgitation (figure 1b).
Fig 1b: Colour Doppler showing severe Aortic Regurgitation.



The left ventricle is dilated with moderate left ventricular dysfunction. Ejection fraction is 40%.Mitral, Tricuspid and pulmonary valves are normal. Aortic annulus measures 2.3 cm and Ascending aorta is 2.9 cm. Contrast enhanced CT aortogram and coronary angiogram were done which also revealed four equal size cusps with normal sized ascending aorta and normal coronaries (Figure 2a,b).

Fig 2a: CT scan cross sectional view showing Quadricuspid aortic valve with four equal sized noncoapting leaflets.

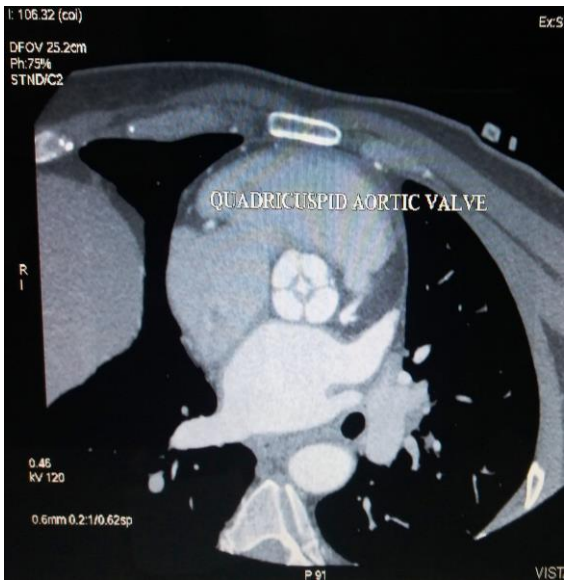
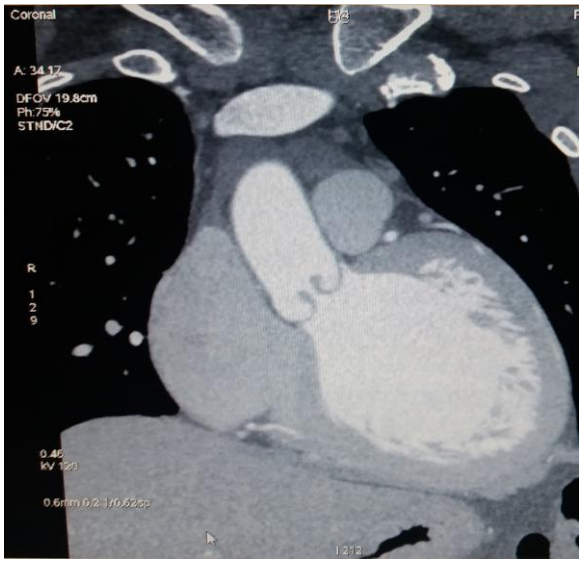


Fig 2b: CT scan Coronal view showing normal size ascending aorta with noncoapting leaflets.



After initial medical stabilization, patient underwent surgery. Intraoperative findings are normal sized ascending aorta, four equal size cusps (2 anterior,2 posterior) of aortic valve , left main coronary artery arising from left posterior cusp and right coronary artery arising from right anterior cusp. Aortic valve leaflets are pliable, noncoapting and having four well defined commissures (Figure3a, b).

Fig 3 a) Excised aortic valve showing 4 equal sized leaflets.

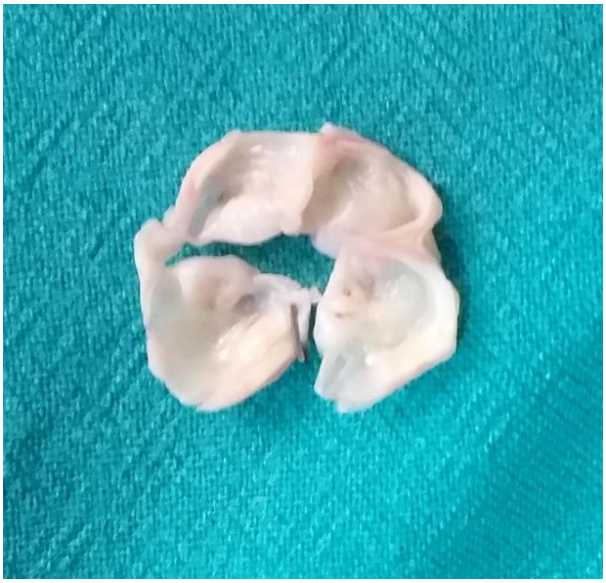
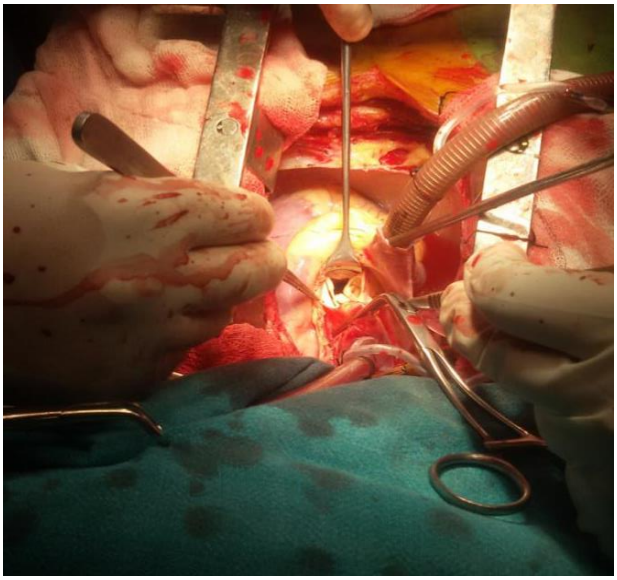


Fig 3b: Intraoperative picture showing quadricuspid aortic valve.



Coronary Ostia are normal. All the four leaflets were excised and aortic valve replaced with 21 mm St Jude rotatable mechanical valve. He had raised renal parameters during initial post operative period but recovered with conservative medical management. The patient is discharged in stable condition. Post operative echo revealed good functioning valve with acceptable gradients and improved ejection fraction (58%).

DISCUSSION:

Quadricuspid aortic valve is one of the rarely seen congenital valve abnormalities. Normal aortic valve has three equal size leaflets.

Quadricuspid aortic valve has four leaflets and may be of varying sizes, but in our case, all are equal in size. Abnormal cusps are due t o a developmental anomaly during embryological truncal septation. Most common aortic valve anomaly is bicuspid followed by unicuspid valve. Quadricuspid aortic valve is very rare and is even less common than quadricuspid pulmonary valve [1]. Quadricuspid aortic valve has incidence of 0.013~0.043% according to autopsy results [2]. QAV also is seen in only0.55~1.46% of aortic valve surgical patients [3, 4].In our center, this is the first case reported.

According to the anatomy of the four cusps, Hurwitz and Roberts categorized QAV into seven subtypes (A to G) [5]. The two most frequent types are type a (four equal cusps) and type B (three normal cusps with one smaller cusp). It is thought that type B has a greater probability of developing aortic regurgitation because of the asymmetric shear stress of the cusps. Our case belongs to Type A with all equal size cusps but presented with severe aortic regurgitation. Jagannathet al. Have done detailed literature review and simplified the classification of QAV (type I to type IV) [6]. Type I and type II are the same as the types A and B of Hurwitz and Roberts classification. Unlike the relatively stable quadricuspid pulmonary valve, more than half of QAV patients develop aortic regurgitation progressively and aortic stenosis is rarely seen. Most of them need surgery in their fifties to sixties.

Some QAVs are often associated with other abnormalities, such as displacement of the coronary sinus and ostium, ventricular septal defect, patent duct arteriosus, subaortic stenosis , cardiomyopathy, Val s a l v a aneurysm, and mitral valve regurgitation [13-15]. Aortic root dilatation was rare in QAV patients as given in the review by Jagannathetal and in our patient also the aortic root was normal. Left ventricular dysfunction was also occasionally seen in some cases. The indication for surgery in QAV a patient depends on the extent of aortic insufficiency and its associated lesions. For aortic insufficiency, the indication for surgery is almost the same as regurgitation caused by other causes like a degenerative or rheumatic disease. Care should be taken not to injure the origin of the coronary arteries. Aortic valve repair by tricuspidisation or bicuspidation give only short term benefits and so not advisable. Valve replacement should be considered in these patients as it provides long term benefit.

CONCLUSION:

Advancements in computer tomography (CT) made the diagnosis of quadricuspid aortic valve more accurate. Treatment of such patients should be individualized based on the degree of regurgitation and associated lesions. As of now, valve replacement is considered as the procedure of choice.

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