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Case Report

Sub aortic fibro muscular ridge with congenital bicuspid aortic valve

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ABSTRACT

Congenital bicuspid aortic valve stenosis is estimated to occur in 1 to 2% of general population, making it the single most common congenital cardiac anomaly, but in association with fibro muscular sub aortic ridge (fibro muscular collar) is a rare combination of congenital cardiac anomalies. We present here a rare case of sub aortic fibro muscular ridge that is associated with congenital bicuspid aortic valve.

Keywords: Congenital heart diseases, Congenital bicuspid aortic value, Aortic stenosis, Sub aortic membrane, Sub aortic fibro muscular ridge

INTRODUCTION

The congenitally bicuspid aortic valve has two functional leaflets. Most have one complete line of coaptation. With this deformity the bicuspid aortic valve does not function perfectly, but it may function adequately for years without causing symptoms or obvious sign of problem**.** Bicuspid aortic valve disease caused by a connective tissue disorder that causes other circulatory systemic problem. Bicuspid aortic valve disease also associated with abnormal coronary arteries, aortic aneurysm, abnormal thoracic aorta and labile blood pressure.

CASE REPORT

A 15 years old asymptomatic male patient comes for evaluation of ejection systolic murmur over right upper sternal border. On clinical examination his growth and development were normal. General appearance was normal. There were no pallor, no clubbing, no cyanosis, all peripheral pulses are palpable, right and left brachial and carotid pulse are symmetrical. His pulse rate was 84/minute, regular, no radio femoral delay. Carotid upstroke was small volume, slow rise and delayed sustained peak (Pulsus Parvus et tardus). A systolic thrill was palpable over carotid arteries and in suprasetrnal notch. His blood pressure was 110/80 mmHg in supine position, in right upper limb. JVP and respiratory rate was normal. Cardiovascular examination revealed, apical impulse present in left 5th intercostal space which was medial to midclavicular line, diffuse, sustained and not displaced. A systolic thrill was palpable in right second intercostal space and it was radiating in to both sides of neck and supra sternal notch. On auscultation S1 was normal and aortic component of S2 was diminished. An aortic opening ejection sound precedes Grade IV ejection systolic murmur audible at base of the heart (Right second intercostal space). Murmur was loudest at base of heart that radiates along the carotid arteries in to the neck, Murmur was low-pitched, rough and rasping in character

Murmur was increase by squatting position and decreased by strain of valsalva maneuver. X ray chest PA view was normal. ECG shows normal P wave, a normal QRS axis, sinus rhythm, and LVH with strain pattern.

Transthoracic two-dimensional echocardiography was done in all views which revealed situs solitus, visceroatrial, atrioventricular, ventriculoarterial concordance and left sided aortic arch. Interatrial septum and interventricular septum both are intact. Atrioventricular valves and pulmonary valve are normal. The parasternal long axis view shows pliable leaflets and systole doming of aortic valve leaflets (Figure 4). The parasternal long axis view shows a small size discrete, broad base, thick, crescent shape fibro muscular ridge (Fibro muscular collar). The fibro muscular ridge is attached to interventricular septum just below aortic valve and slightly extending from anterior ventricular septum into the LVOT, it was not extending up to anterior leaflet of mitral valve. There was no sub aortic obstruction but trivial aortic regurgitation was present. The Basal short axis view shows mobile, stenotic, unequal size , two cusps of bicuspid aortic valve, the ventricular systolic frame shows a circle orifice opening of aortic valve (Figure 1) and the ventricular diastole frame shows a vertical commissure in between two cusps of aortic valve (Figure 2). Supra- sternal view shows post stenotic dilatation of ascending aorta. The continuous-wave Doppler recording in apical long axis and apical 5 chambers views, across aortic valve level shows, moderate aortic valve stenosis and the flow velocity was 3.5m/second, reflecting peak instantaneous gradient of 55mmHg and mean pressure gradient 32 mmHg. There is no significant gradient at sub aortic valve level.

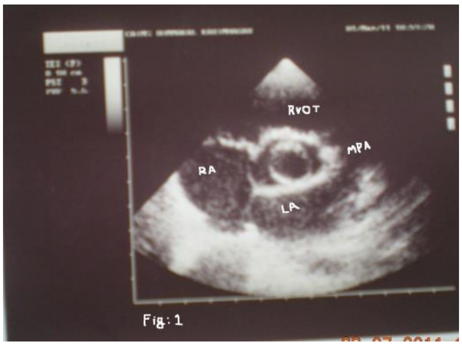

Figure 1: Basal short axis view shows, a circular opening of a bicuspid aortic valve, in ventricular systole. (RVOT- Right Ventricular Outflow Tract, MPA- Main Pulmonary Artery, LA- Left Atrium, RA- Right Atrium).

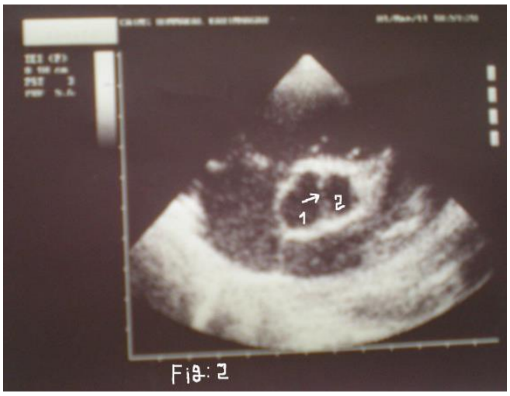

Figure 2: Ventricular diastolic frame shows, a vertical commissure (single arrow) in between two cups (1,2) of a bicuspid aortic valve.

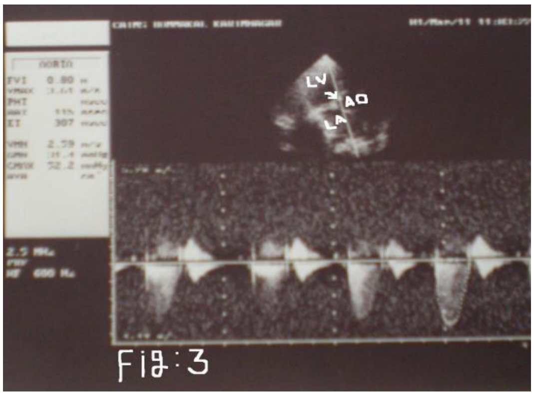

Figure 3: Continuous wave doppler recording in apical long axis view across aortic valve (arrow) shows, moderate A.S., flow velocity 3.5 m/sec, PPG 53mmHg & MPG 32mmHg.

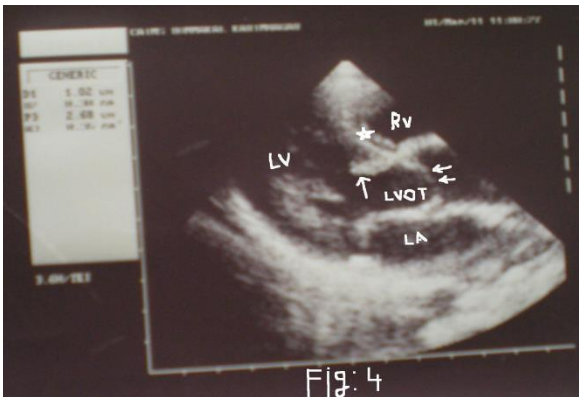

Figure 4: Parasternal long axis view shows a small, thick, fibro muscular ridge (single arrow) just below aortic valve and it is attached to inter ventricular septum (Star). Systolic doming of Aortic Valve Leaflets (paired arrows). (LV-Left Ventricle, LA- Left Atrium, LVOT- Left Ventricular Outflow Tract, RV – Right ventricle).

DISCUSSION

Aortic stenosis may be present at birth (a congenital stenotic aortic valve) or may develop over time in a congenitally abnormal but not stenotic valve. A bicuspid valve can also be stenotic at birth because of commissural fusion or dysplasia. Most often, such valves will be functionally normal at birth but gradually become stenotic over time because of progressive fibrosis and calcification. Two- dimensional echocardiography plays a major role in detection of this entity. Direct visualization of aortic cusps is possible from the parasternal short axis view through the base of heart. During diastole, the cusps of a normal tricuspid aortic valve , are closed within the plan of the scan and commissures form a "Y", sometime referred to as an inverted Merced's - Benz sign (Figure 5).

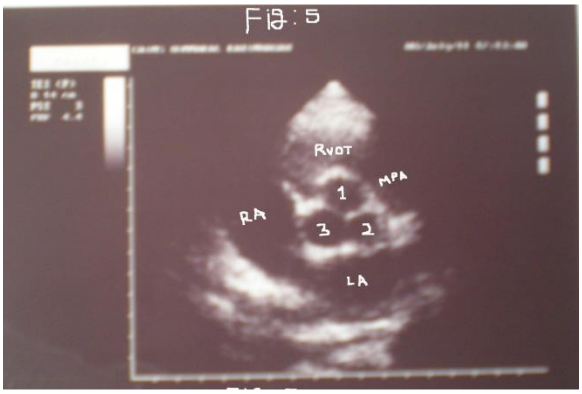

Figure 5: Parasternal short axis view demonstrates a normal tricuspid Aortic valve in diastole. Inverted Mercedes -Benz sign. (1 Right coronary cusp, 2 Left coronary cusp, 3 Non coronary cusp, RA- Right Atrium, LA- Left atrium, RVOT- Right Ventricular Outflow Tract, MPA- Main Pulmonary Artery).

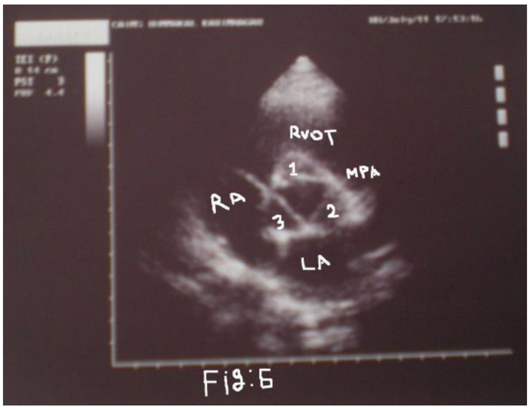

Figure 6: Parasternal short axis view shows, normal opening and office of a normal tricuspid aortic valve in systole. (RA- Right atrium, LA- Left atrium, RVOT- Right ventricular outflow tract, 1- Right coronary cusp, 2- Left coronary cusp, 3- Non coronary cusp).

A true bicuspid valve has two cusps of nearly equal size, two associated sinuses, a single linear commissure, it may be horizontal or vertical (Vertical in our case, Figure 2). A rape may be present and, if present, create the illusion of three separate cusps. By observing valve opening in systole, however the numbers of distinct cusps is apparent. Fusion of two cusps may create the appearance of a bicuspid valve, but the presence of three distinct sinuses will establish this difference. Confirming the presence of a bicuspid aortic valve with echocardiography requires high-resolution image from short-axis view for adequate visualization of valve morphology.

An accurate assessment of functional anatomy requires an analysis of the number of apparent cusps, the degree of cusp separation and recording of their mobility and excursion during systole. The short axis view is useful for determination of number of commissures, degree of fusion of commissures, movement of cusps during systole. The long axis is useful for assessment of cusps thickness and excursion of leaflet in systole, the degree of LVH and presence of post stenotic aortic root dilation. With the help of Doppler imagine, we can evaluate the severity of aortic stenosis. The apical, right parasternal and suprasternal windows should be used to obtain maximal velocity, then through the use of modified Bernoulli equation the peak pressure gradient can be calculated. Both peak instantaneous and mean pressure gradient (MPG) can be derived and in children the MPG is often used for clinical decision. (Table 1, as in our case MPG is 32mmHg, means he is having moderate AS). To calculate Aortic valve area we can use the continuity equation and we can calculate the severity of A.S. (Table 2).

Table 1: Severity of A.S. according to Mean Tran's valvular gradient (MTVG across aortic valve).

Normal MTVG	0 mmHg
Mild A.S.	0-20 mmHg
Moderate A.S.	20-40 mmHg
Severe A.S.	40-50 mmHg
Critical A.S.	>50mmHg

The sub aortic fibromuscular ridge or collar is a thin, fibrous, crescent shape, membranous or fibro muscular, located immediately below the aortic valve in LVOT. The membrane is some times relatively thick forming a fibrous or fibro muscular collar. The fibro muscular ridge is made up of collagen fibers, elastic fibers and myocytes. The membrane usually extend from anterior ventricular septum to the anterior mitral leaflet (AML) and lead to subvalvular obstruction and some time it may be very small and not reach up to AML and there is no subvalvular obstruction (as in our case). The degree of subvalvular obstruction is variable and aortic regurgitation develops in 50% of patient. In two-dimensional echocardiography, the sub aortic membrane, seen as a discrete linear echo in Left ventricular out flow tract (LVOT), perpendicular to IVS. This membrane easily detected in PLAx &AP4C view.

Table 2: Severity of A.S. according to aortic valve area. AVA (cm^2)= [(LVOT diameter^2 X 0.78540 /LVOT TVI)]/ Aortic valve TVI (Obtain by converting the diameter to area and assuming that is circular, (V) LVOT= peak outflow tract velocity and (V) AV= peak velocity across the aortic value).

Normal AVA	3-4cm^2
Mild AS	>1.5cm^2
Moderate AS	1.0 – 1.5cm^2
Severe AS	<1.0cm^2
Critical AS	<0.75cm^2

Medical Management in asymptomatic patient

1. A malformed aortic valve is a potential site of bacterial infection, so antibiotic prophylaxis, is recommended for all patients, regardless of the severity of obstruction.
2. Avoid strenuous physical activity & Participation in competitive sports.
3. Digitalis should be start, if the patients develop symptoms of diminished cardiac reserve and also considered for patient with LVH.

Onset of symptoms (Angina, Syncope and Heart failure) indicate severe A.S. and Need urgent surgical intervention

Surgical Management: Correcting surgery may be performed only after patient become symptomatic or when patient develop Left ventricular dysfunction, as evidenced by echocardiography, whichever may be earlier. Types of surgery are percutaneous balloon aortic valvuloplasty (PBAV) and aortic valve replacement (Tissue valve and prosthetic valve).

Membranectomy with concomitant Myomectomy or Myotomy is usually performed for sub aortic fibro muscular ridge.

CONCLUSION

In the present case, the patient is asymptomatic and 2-dimnsional echocardiography shows, congenital bicuspid aortic valve with non obstructive fibro muscular ridge and moderate aortic valve stenosis (according to MPG) and normal LV function so in our opinion, he should wait for surgery until unless he is asymptomatic; during asymptomatic period he should regularly come for follow-up.

REFERENCES

1. Brandenburg J, Tajik AJ, Edwards WD, et al. Accuracy of 2-dimensional echocardiography diagnosis of congenital bicuspid aortic valve: Echocardiographic- anatomic correlation in 115 patients. Am J Cardiol 1983;51:1469.
2. Blackwood RA, Bloom KR, Williams CM. Aortic stenosis in children, experience with echocardiography prediction of severity. Circulation 1978;57:263.
3. Brooks N. Rapid development of severe aortic stenosis from calcification of congenital bicuspid aortic valve. Br Med J 1980;281:424.
4. Cabrera A, Galdeano J, Zumalde J, Mondragon F, et al. Fixed sub aortic stenosis. The value of cross-sectional echocardiography in evaluating different anatomical patterns. Int J Cardiol 1989;24:151.
5. Campbell M. The natural history of congenital aortic stenosis. Br Heart J 1968;30:514.
6. Fenoglio JJ, McAllister HA, et al. Congenital bicuspid aortic valve after age 20. Am J Cardiol 1977;39:164.
7. Gale AW, Cartmill TB, Bernstein LE. Familial subaortic membrane stenosis. Austr NZ J Med 1974;4:576.
8. Huhta JC, Laston LA, Gustgesell HP, et al. Echocardiography in diagnosis and management of symptomatic aortic valve stenosis in infants. Circulation 1984;70:438.
9. Katz NM, Buckley MJ, Liberthson RR. Discrete membranous sub aortic stenosis. Circulation 1977;56:1034.
10. Kelly DT, Wulfsberg E, Rowe RD. Discrete sub aortic stenosis. Circulation 1972;46:309.

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