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Original Research Paper

Cardiology

A RARE ANOMALY OF AORTIC VALVE

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ABSTRACT **Background:** The Quadricuspid aortic valve (QAV) is a rare; often isolated, sometimes associated with other heart diseases. We report a case of quadricuspid aortic valve. **Case Report:** 46 yr old Female who came to our OPD with complaints of breathlessness, palpitations past 2 months. On further evaluation echo showed quadricuspid aortic valve , severe Aortic regurgitation. Patient was referred for surgery. **Conclusion:** The quadricuspid aortic valve is one cause of significant aortic regurgitation that is the predominant clinical finding. It's early recognition is particularly important for surgical management.

KEYWORDS : quadricuspid aortic valve, transesophageal echocardiography, aortic regurgitation

INTRODUCTION:

Quadricuspid aortic valve (QAV) is a rare congenital cardiac defect with an estimated incidence of <0.05%.[1] Before the era of echocardiography, the diagnosis was made incidentally at autopsy or during surgery of valve replacement. The extensive use of echocardiography has allowed an early and accurate diagnosis. In many cases, the transthoracic echo approach is suitable for the diagnosis but, transesophageal echocardiography is a tool for the accurate definition of the valve anatomy.

Debates remain in the management strategies of the patients with a quadricuspid aortic valve in terms of surgical indication, and antibiotic prophylaxis against infective endocarditis.

Case Report:

A 46yr female with complaints of progressive dyspnea on exertion, chest discomfort, palpitations for past 2 months. Upon initial evaluation , patient was afebrile , Bp was 120/60mmhg ; HR 78/min. Cardiac examination - regular rhythm , auscultation revealed a grade ¾ diastolic murmur beat heard at left lower sternal border and no signs of heart failure.

Transthoracic and Transesophageal echo showed quadricuspid aortic valve (figure 1 to 4) with severe aortic regurgitation with mild LV systolic dysfunction. Patient was referred for surgery. Coronary angiogram before surgery showed normal coronaries. Patient underwent aortic valve replacement. Pathology of valve showed 4 leaflets with thickening and focal fibrosis.

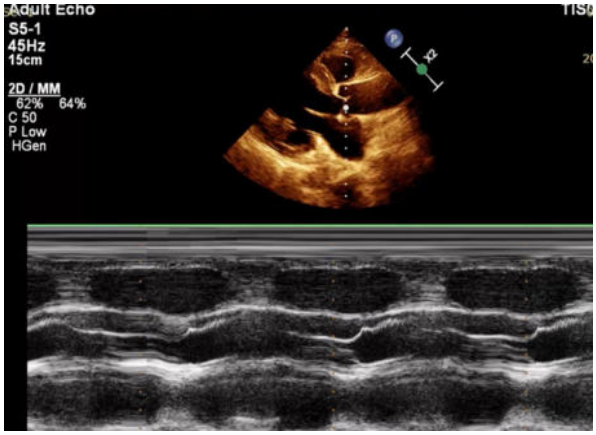

Fig 1 M Mode Of Quadricuspid Aortic Valve

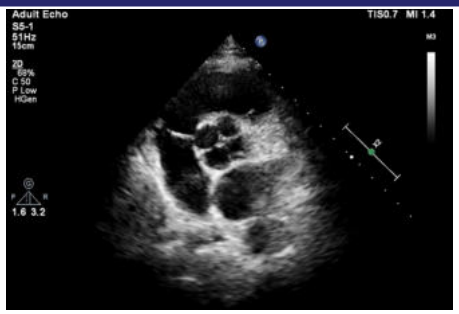

Fig 2 : Trans Thoracic Short Axis View Of Quadricuspid Aortic Valve

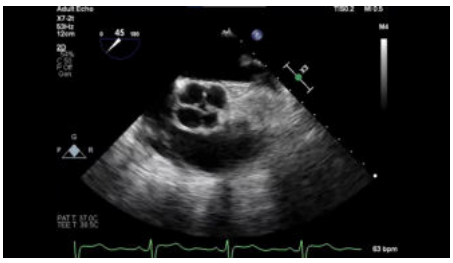

Fig 3 Trans Esophageal Short Axis

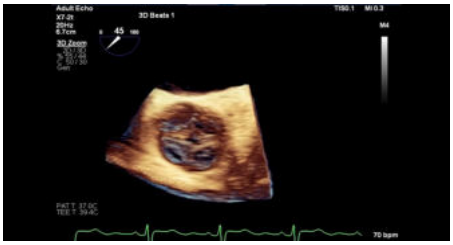

Fig 4 Trans Esophageal 3d View Of Quadricuspid Aortic Valve

DISCUSSION:

QAV is rare cardiac abnormality can be associated with other abnormalities which include pulmonary valve stenosis, Ventricular septal defect, subaortic fibromuscular stenosis, and coronary ostial malformation.[2]

The mechanisms of development are not well established. It has been suggested that the development of QAV is from abnormal division of one of the three mesenchymal ridges that later become the aortic valve cushions.[3]

There are several classifications of QAV based on the location of accessory cusp and sizes of the cusps. Hurwitz and Roberts[4]

classified QAV into subtypes A to G on the basis of the relative sizes of the valve leaflets. The most common morphologies identified are type A (four equally sized cusps) and type B (three equally sized cusps and a smaller accessory cusp).[5]

Patients with QAV present with symptoms of aortic regurgitation such as dyspnea on exertion and angina.[3]

Echocardiography is the standard imaging modality used in patients with suspicion of valvular heart disease.[6] TEE helps to delineate the valve's morphology and functionality. However, despite the use of echocardiography, in few cases, the final diagnosis is not made until the valve is directly visualized during surgery or autopsy.[3]

The most common indications for surgery are severe aortic stenosis, severe aortic regurgitation, or dysfunctional QAV with other lesions, such as an occluded left coronary ostium.[2] Most patients undergo surgery for chronic severe aortic regurgitation.[6]

CONCLUSION :

QAV is a rare cause of aortic regurgitation with an incidence of <1%. Echocardiography, especially TEE, is useful diagnostic modality. Patients with this QAV often undergo aortic valve replacement for left ventricular systolic dysfunction, symptomatic valvular disease, or associated structural abnormalities found associated with the valve.

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