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The "X-Valve". Current considerations on quadricuspid aortic valve disease

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Abstract

Quadricuspid aortic valve (QAV) disease is a rare congenital aortic valve abnormality that is commonly found incidentally [1,2]. It mainly leads to aortic regurgitation (AR) and might be correlated with other major or minor structural cardiac defects [3]. Apart from the severity of the AR, management of QAV is related to symptoms as well as left ventricle (LV) dimensions. Hence, the correct time for surgical aortic valve replacement remains crucial. We present a case of QAV detected by echocardiogram in patient with moderate AR.

Keywords

aortic valve; quadricuspid aortic valve; aortic regurgitation; echocardiogram

Background

Quadricuspid aortic valve (QAV) disease is an infrequent aortic valve irregularity leading to AR and perhaps other structural cardiac defects. Diagnostic and therapeutic management of this condition and most importantly proper timing for aortic valve intervention is a matter of concern. Current considerations of this infrequent aortic valve variation are presented [1,3].

Case Presentation

The patient is a 43-year-old man that was presented at the emergency department complaining of fatigue and dizziness which started a few weeks ago. He was not receiving any medication and had no prior medical history. On clinical examination, a diastolic heart murmur was audible at the second left parasternal border. The rest of physical examination as well his vital signs were unremarkable. Electrocardiogram (ECG) was normal without ischemic changes and the laboratory results were also unremarkable (Figure 1). The patient was admitted to the Cardiology department for further investigation.

Transthoracic echocardiogram (TTE) revealed normal left ventricular ejection fraction (LVEF), moderate AR and an atypical aortic valve without the "Y-shaped" trileaflet morphology. The aortic root, the mitral, tricuspid and pulmonary valves were normal. There were no other cardiac defects or signs of rheumatic valve disease. The quantification of the aortic valve defect showed a Vena Contracta at 3.6mm estimated at parasternal long axis view. The regurgitant volume was calculated by the continuity

equation method and was 45ml and the regurgitant orifice was calculated to be 0.2cm^2 [2]. Reverse flow during early diastolic period was revealed in the descending thoracic aorta. Transesophageal echocardiogram (TEE) was performed consequently for a more detailed evaluation. During diastolic phase, the commissural lines formed by the valve leaflets resulted in an ''X" modulation. The presence of a QAV with three equally sized cusps along with a smaller one and moderate regurgitation was verified. According to the Hurwitz and Roberts classification, our patient had the type B QAV (Figure 2).

During hospitalization, his general condition remained stable. The heart team decided that the patient should receive follow up care with echocardiogram evaluation every six months due to lack of symptoms and LV dysfunction.

Discussion

QAV is a rare congenital valve malformation with an estimated incidence between 0.003% and 0.043% at autopsy and to 1% among patients posted for aortic valve replacement [3,4]. The first case was described by Balington in 1862 and approximately 200 cases have been reported since then [5]. In the past, QAV was randomly diagnosed during surgery or autopsy. Nowadays, both TTE and TEE play a major role to detect this entity. Moreover, the diagnostic use of computed tomography (CT) and cardiac magnetic resonance imaging (MRI) have contributed to the increased prevalence of QAV [6].

The mean age of diagnosis is around 50 years with a slight male predominance. The most usual valvular abnormality in QAV is AR whereas isolated aortic stenosis is extremely rare (0.7%). Combination of QAV regurgitation and stenosis has been described in approximately 8% of all reported cases. Asymmetric distribution of stress around the four cusps along with abnormal leaflet coaptation is the main mechanism of regurgitation [7,8].

QAV might be correlated with several congenital cardiac defects including coronary artery and coronary ostium anomalies, atrial or ventricular septal defects, mitral valve prolapse and regurgitation and/or subaortic fibromuscular stenosis. Additionally, the risk of infective endocarditis is higher due to the progressive deterioration of the leaflets, especially in patients with unequally sized cusps [8].

To date there are two classification schemes (Table 1). The Hurwitz and Roberts classification divides QAV into seven types from A to G according to the size of each leaflet. Over 85% of the reported cases are of type A (four equal leaflets), B (three equally sized cusps with a smaller one) or C (two equal larger and two equal smaller cusps) [6,9]. Nakamura and colleagues designed a most simplified classification focused on the position of the accessory leaflet. Type II QAV in which supernumerary cusp is located between right and non-coronary cusps is considered the most common valve malformation that account for 30% [10,11]. In our case, the QAV was classified as type B with three equal sized aortic cusps along with a smaller one.

Management of QAV mainly depends on patient's symptoms, LV geometry and enlargement and the extent of AR. According to current guidelines the indications for surgery are similar to any regurgitant aortic valve [8,12,13]. Surgical aortic valve replacement is the gold standard for treatment of severe AR and is recommended irrespective of the LVEF value in symptomatic patients. Furthermore, surgical treatment is also indicated in asymptomatic patients with severe AR and LVEF ≤50% or LV enlargement

with LV end-diastolic diameter (LVEDD) >70mm or LV end-systolic diameter (LVESD) >50mm. The performance of aortic valve replacement in patients with severe AR associated with LV dysfunction results in a significant survival benefit and should not be avoided even inpatients with EF<20% despite the higher surgical risk [14]. In younger patients like in our case with mild symptoms, moderate AR and normal LV a conservative strategy with regular clinical and TTE follow up is the appropriate choice of treatment. Diagnostic angiogram is recommended only in patients with predisposing risk factors for coronary artery disease or for pre – surgery evaluation.

Transcatheter aortic valve replacement (TAVR) is emerging as novel treatment option among patients with pure AR. It might be technically feasible in selected high-risk patients with native pure AR and acceptable early morbidity and mortality. According to a recent meta-analysis the estimated procedural mortality was lower than 30% and significant post-procedural AR ranged from 0% to 88%. However, the entirely different anatomical pattern of both diseases challenges the proper positioning and stability of current transcatheter valve devices in patients with pure native AR compared to those with aortic valve stenosis [15].

Figures

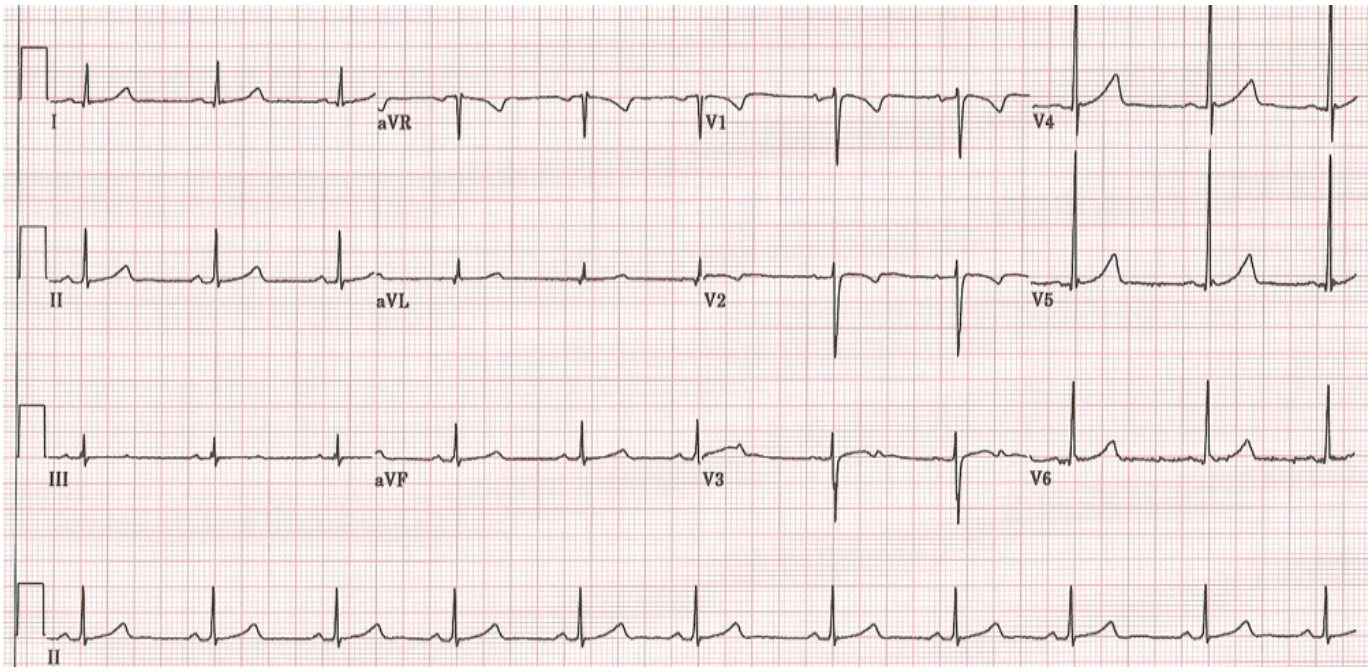

Figure 1: Patient's electrocardiogram

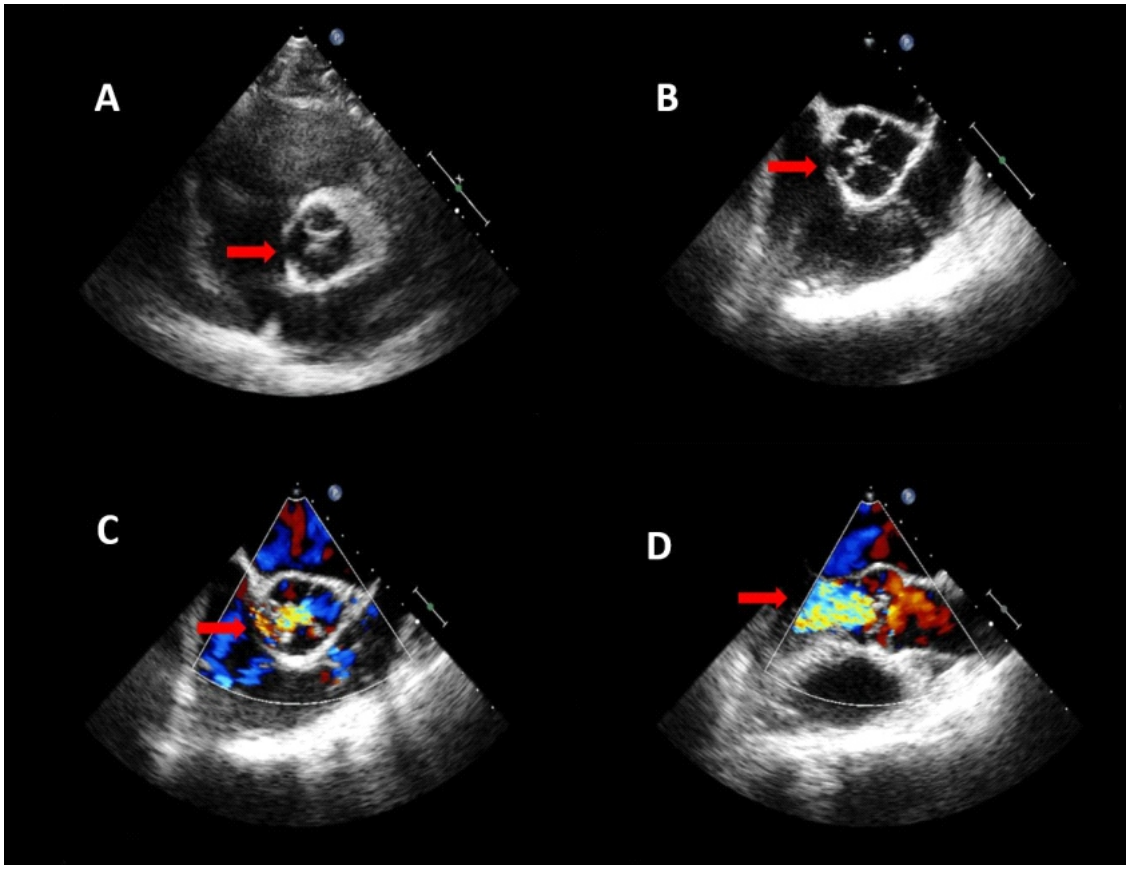

Figure 2: Transthoracic and transesophageal echocardiographic view of the aortic valve.

A. TTE PSAX –aortic valve during diastole
B. TEE PSAX –aortic valve during diastole
C. TEE PSAX – Color Doppler visualization of the regurgitant jet
D. TEE PSAX – Color Doppler visualization of the regurgitant jet

Tables

Table 1: Classification schemes of QAV (Quadricuspid aortic valve).

Hurwitz and Roberts classification	
Type A	Four equally sized leaflets
Type B	Three equally sized leaflets and one smaller leaflet
Type C	Two equally sized large leaflets and two equally sized smaller leaflets
Type D	One large leaflet, two mid-sized leaflets and one smaller leaflet
Type E	Three equally sized leaflets and one larger leaflet
Type F	Four unequally sized leaflets
Type G	One large leaflet, one mid-sized leaflet and two equally sized smaller leaflets
Nakamura and colleagues classification	
Type I	Supernumerary leaflet between the left and right coronary leaflets
Type II	Supernumerary leaflet between the right and non-coronary leaflets
Type III	Supernumerary cusp between the left and non-coronary leaflets
Type IV	Unidentified supernumerary cusp - two equally sized smaller leaflets

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