International Journal of Medical Science and Clinical Research Studies

ISSN(print): 2767-8326, ISSN(online): 2767-8342

Volume 04 Issue 03 March 2024

Page No: 556-558

DOI: https://doi.org/10.47191/ijmscrs/v4-i03-33, Impact Factor: 7.949

Diagnostic Challenges of Quadricuspid Aortic Valve

Roberto Cintra A. Aragao*[1], Amanda Raquel Costa Cruz[2], Iana Carine Machado Bispo[3], Vitor Vahle[4], Luiz Flavio Galvao Goncalves[5], José Augusto Soares Barreto Filho[6]

ABSTRACT

The quadricuspid aortic valve (QAV) is a rare anatomical variation, and the estimated incidence is lower than 0,05% of all congenital heart diseases (1). It is usually found as an isolated alteration, however, it may be associated with other concomitant cardiac anomalies (2).

In this case report, an atypical presentation of the QAV was observed, with a lot of clinical dissociation versus complementary exams of excellent accuracy for the diagnosis.

KEYWORDS: Quadricuspid aortic valve, Dyspnea, Echocardiogram, Cardiac catheterization, Aortic regurgitation,PD2.

ARTICLE DETAILS

Published On:
30 March 2024

Available on:
https://ijmscr.org/

CASE REPORT

Patient sought medical attention reporting dyspnea on exertion for 2 years, during pregnancy, with improvement after delivery and initiation of losartan to treat the recent diagnosis of arterial hypertension. In the last 2 months, he noticed a worsening of his dyspnea, now with medium/small efforts such as simple household chores, in addition to paroxysmal nocturnal dyspnea and orthopnea. Physical examination revealed: blood pressure (BP) 120/60mmHg, heart rate (HR) 70bpm, afebrile, SpO2 98% on room air, regular heart rhythm in 2 strokes, with aortic aspiration diastolic murmur, grade 2, clean lungs, with good peripheral perfusion and no edema.

She had preserved systolic function on transthoracic echocardiogram (TTE), left ventricular (LV) ejection fraction of 60%, moderate aortic regurgitation due to the quadricuspid valve and brain natriuretic peptide (BNP) 24.3pg/ml (reference value: 0 to 100pg/ml). Transesophageal echocardiogram (TEE) showed moderate aortic insufficiency, vena contracta 0.4 cm, and left ventricular dysfunction by Strain -17% (RV: > -18%). The symptoms of dyspnea presented by the patient during exertion, associated with reduced values of peak VO2 (41% of predicted), anaerobic threshold VO2 (39% of predicted VO2 max), peak oxygen pulse 8ml/beat (predicted value: 10.6ml/beat) and increased VE/VO2 slope ratio 40.44 (RV: < 34) suggest cardiovascular limitation according to the cardiopulmonary exercise test.

Dyspnea of pulmonary origin was ruled out through further investigation, and cardiac catheterization was indicated for aortography and left ventricular end-diastolic pressure (PD2) measurement due to clinical-echocardiographic disagreement, demonstrating significant aortic insufficiency and PD2 of 16 mmHg (Figure 1). There were no other anatomical alterations in coronary CT angiography and the morphological diagnosis of type B VAQ (Three Equal Cusp, One Smaller Cusp) was confirmed (Figure 2).

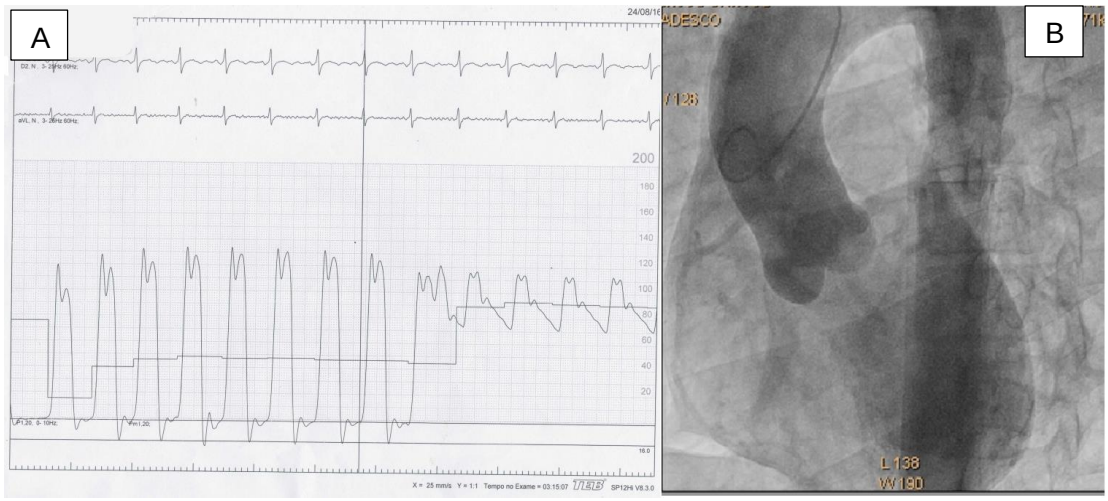

Figure 1. Left ventricular (LV) and aortic (AO) pressure curves with LV-AO peak gradient 6mmHg and PD2 16mmHg (A). Severe Aortic Regurgitation: contrast filling of the entire left ventricle in diastole equal in density to the contrast opacification of the ascending aorta and clearing only after several cardiac cycles (B).

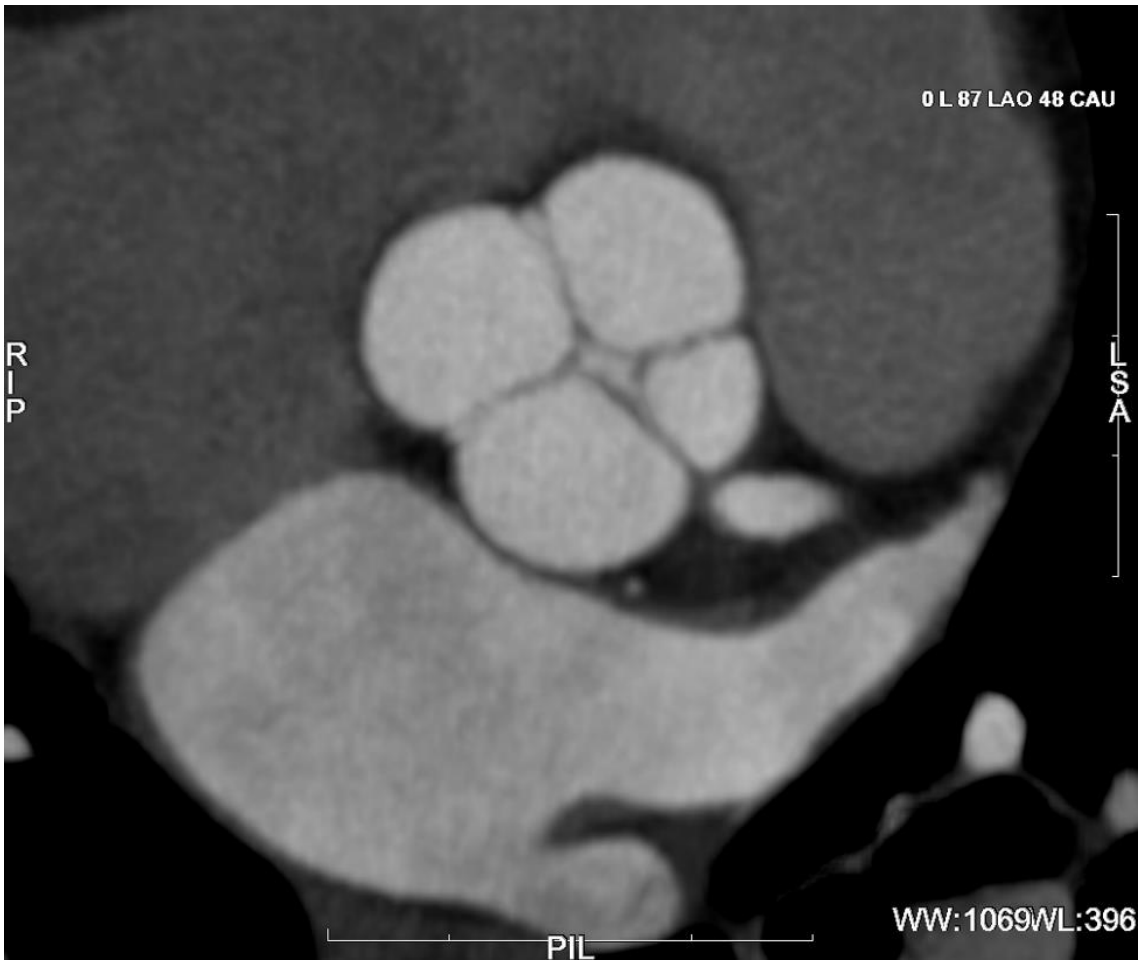

Figure 2. Type B quadricuspid aortic valve, in the Hurwitz & Roberts classification, represents three equal-sized cusps and one smaller cusp.

Modified therapy for perindopril and furosemide, evolving with partial improvement of symptoms.

Decided in the Heart Team and opted for a surgical approach of the aortic valve with complete resolution of symptoms.

DISCUSSION

A quadricuspid semilunar valve is an uncommon condition. According to the morphology of the leaflet, 7 types of QAV were described by Hurwitz and Roberts, named from A to G (1).

Initially found incidentally in autopsies, highlighting, currently, transthoracic, and transesophageal echocardiography as the main diagnostic tool (2)

Aortic regurgitation is the predominant valvular dysfunction and is seen in up to 75% of documented cases, on the other hand, quadricuspid aortic valve is rarely associated with aortic stenosis (3).

The quadricuspid aortic valve is usually kept normal when the patient is at age of <18 years, and it is worsening at >40 years. A significant valvular disorder is often present with subsequent operation performed in the fifth to sixth decade of life. Palpitations, chest pain, shortness of breath, fatigue, pedal edema, syncope, and congestive heart failure can be the presenting symptom (3-4).

There is no established association between QAV morphology and the severity of aortic regurgitation, which suggests that the dysfunction is associated with evolutionary degeneration (4).

The preferred diagnostic tool seems to be transesophageal echocardiography because it shows the morphology of the QAV and reveals an anomaly of the coronary ostium and ascending aorta (4), however, there is no consensus on the best diagnostic method for detecting the QAV, given the low prevalence of this pathology (5). M. Dencker and M. Stagmo reported a case of a symptomatic patient with severe QAV diagnosed through TEE, with two previous normal TTEs performed in 6 years of follow-up and elective valve replacement was indicated (5).

It is speculated that the eccentricity of the reflux jet makes it difficult to quantify aortic regurgitation by traditional parameters and that in quadricuspid morphology this alteration is more frequent. There are no data reported in the literature to date, comparing the degree of aortic regurgitation diagnosed using the two methods which may justify the use of the invasive diagnostic method in selected cases.

The surgical intervention follows current guidelines, however, decision-making becomes difficult given the wide spectrum of structural clinical presentations of VAQ.

CONCLUSION

In the reported case, there was a dissociation between symptoms and echocardiography in quadricuspid aortic valve, with the diagnosis of significant aortic regurgitation being made by aortography and LV end-diastolic pressure measurements, suggesting a limitation of the echocardiographic method in selected cases. Therefore, this case demonstrates a limitation of the echocardiographic method in QAVs and the importance of invasive analysis for diagnosing important valvopathies in dubious cases.

REFERENCES

I. Hurwitz LE, Robert WC. Quadricuspid semilunar valves. Am J Cardiol 1973;31:623-626. doi: 10.1016/0002-9149(73)90332-9.

II. Tsang MY, Abudiab MM, Ammash NM, et al. Quadricuspid aortic valve: characteristics, associated structural cardiovascular abnormalities, and clinical outcomes. Circulation 2016; 133: 312-319.
doi: 10.1161/CIRCULATIONAHA.115.017743.

III. Tutarel O. The quadricuspid aortic valve: a comprehensive review. J Heart Valve Dis 2004; 13: 534-537.

IV. Yuan S-M. Quadricuspid aortic valve: a comprehensive review. Braz J Cardiovasc Surg. 2016;31:454-60.

V. Dencker M, Stagmo M. Quadricuspid aortic valve not discovered by transthoracic echocardiography. Cardiovasc Ultrasound. 2006;4:41.