Quadricuspid Aortic Valve Replaced via Minithoracotomy: A Short Case Report

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Introduction

The advancement of echocardiographic techniques enabled the early diagnosis of quadricuspid aortic valve (QAV), a rare congenital heart disease that typically leads to a significant valvular disease around the fourth, fifth or sixth decade of life. Two-dimensional transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) have been increasingly used to detect such abnormality. Real-time three-dimensional transesophageal echocardiography does online evaluation of cardiac structures, providing new points of view of cardiac conditions, even the most complex ones, although it does not add any information for the diagnosis of QAV.[1,2]

Case report

A 51-year-old hypertensive female, former smoker, diagnosed with aortic regurgitation 5 years ago, was referred to Instituto do Coração de São Paulo (InCor-HCFMUSP) for evaluation. In the previous, she complained of progressive worsening of symptoms of fatigue and dyspnea, with significant limitation for routine activities, currently in NYHA functional class III. The patient was taking enalapril 40 mg/day, amlodipine 10 mg/day, furosemide 40 mg/day and spironolactone 25 mg/day. Physical examination revealed aspiration murmur in early diastole and along the left sternal border. Her blood pressure was 142/50 mmHg. Transthoracic echocardiogram showed slight increase in the left ventricular cavity with preserved systolic function. Color flow Doppler imaging showed severe aortic regurgitation (Figure 1). Signs of coaptation failure were noticed between the valves, although an accurate anatomical characterization was not possible due to limitation of the acoustic window in the transverse plane. End left ventricular ejection diastolic diameter was 56 mm, systolic diameter was 33 mm and ejection fraction was 0.71. Due to the recent worsening of symptoms, coupled with the findings of physical examination and echocardiography scans, elective valve replacement surgery was indicated. Mini-thoracotomy was the chosen technique.

Keywords

Keywords: Heart Valve Diseases; Aortica Valve/surgery; Heart Defects, Congenital; Thoracotomy; Echocardiography.

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Intraoperative transesophageal echocardiography was requested to assist the surgery and it revealed quadricuspid aortic valve as a cause of aortic regurgitation (Figure 2), which was not shown in the previous transthoracic scans. The surgery (Figure 3) did not have any major complications. Bioprosthesis Braile 21 mm was implanted. The patient had an excellent recovery and was discharged on the sixth day after surgery. She is currently asymptomatic.

Discussion

Quadricuspid aortic valve (QAV) is a rare cardiac malformation, with an incidence of 0.003 to 0.043% of all congenital heart defects.[1,2] Many cases are incidentally diagnosed in aortic surgeries or autopsies. The mechanism responsible for this abnormality is not known, but the strongest hypothesis is abnormal septation of embryological artery trunk.[3]

Two classifications are used to describe the QAV. The most widely used is the one by Hurwitz and Roberts,[1] which divides 7 alphabetical subtypes based on the cusps size: type A - 4 identical leaflets, type B - 3 identical leaflets and 1 smaller leaflet, type C - 2 larger identical leaflets and 2 smaller identical leaflets, type D - 1 larger leaflet, 2 intermediate leaflets and 1 smaller leaflet, type E - 3 identical leaflets and 1 larger leaflet, type F - 2 larger identical leaflets and 2 smaller non-identical leaflets, type G - 4 non-identical leaflets. The other classification is the one by Nakamura et al.,[4] very useful as it is based on the non-dominant extranumerary cusp position: I - accessory valve between the left coronary cusp and right coronary cusp, II - accessory valve between the right coronary valve and the non-coronary valve, III - accessory valve between the left coronary valve and the non-coronary valve, IV - ancillary aortic valve indistinguishable because the valve non-coronary valve is divided into 2 equal parts.

On echocardiography, QAV is identified by its characteristic "X" shape during diastole (different from the "Y" of the standard trivalvular aortic valve) and its rectangular shape during systole.[5] According to the Hurwitz and Roberts classification,[1] only 12% of the quadrivalvular aortic valves have same-size valves.[1]

Although TTE allows, compared to TEE, a better aortic valve imaging resolution, there is no consensus on the best diagnostic method for detecting QAV given the low prevalence of this pathology.[6]

Functionally speaking, the quadricuspid aortic valve tends to evolve to failure over decades (rarely before adulthood),[7] due to the asymmetry in the distribution of transvalvar flow and inequality in coaptation of the leaflets. Some cases of mixed valve dysfunction have been described, but the finding of pure valvular stenosis is very rare.[6,8,9]

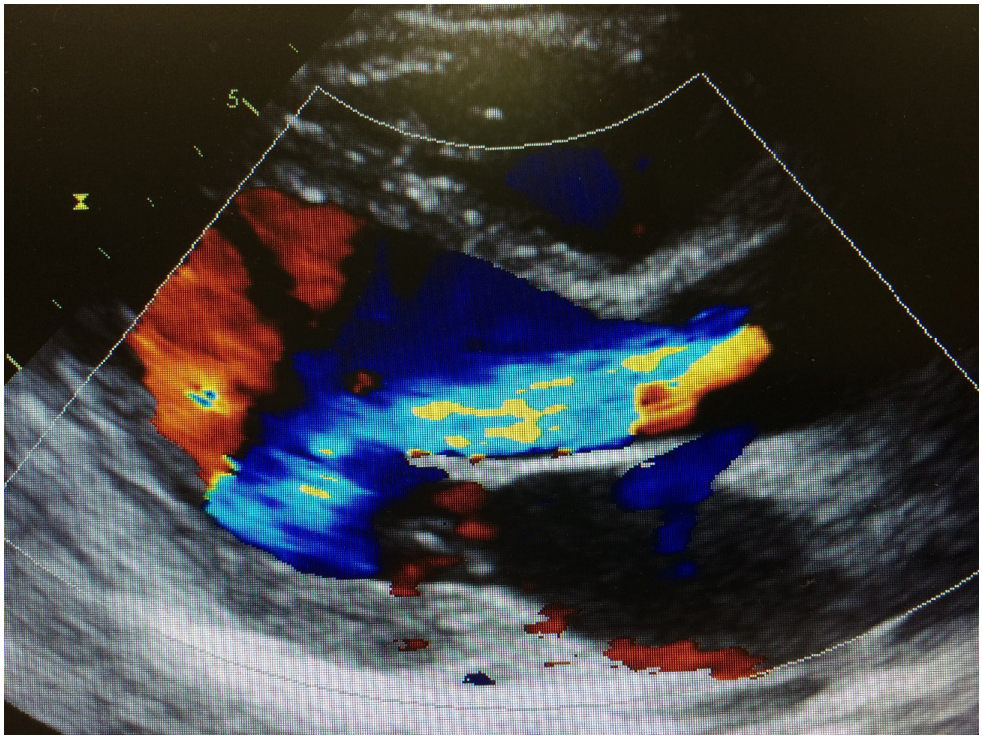

Figure 1 – *Transthoracic echocardiography in longitudinal parasternal view showing severe aortic regurgitation.*

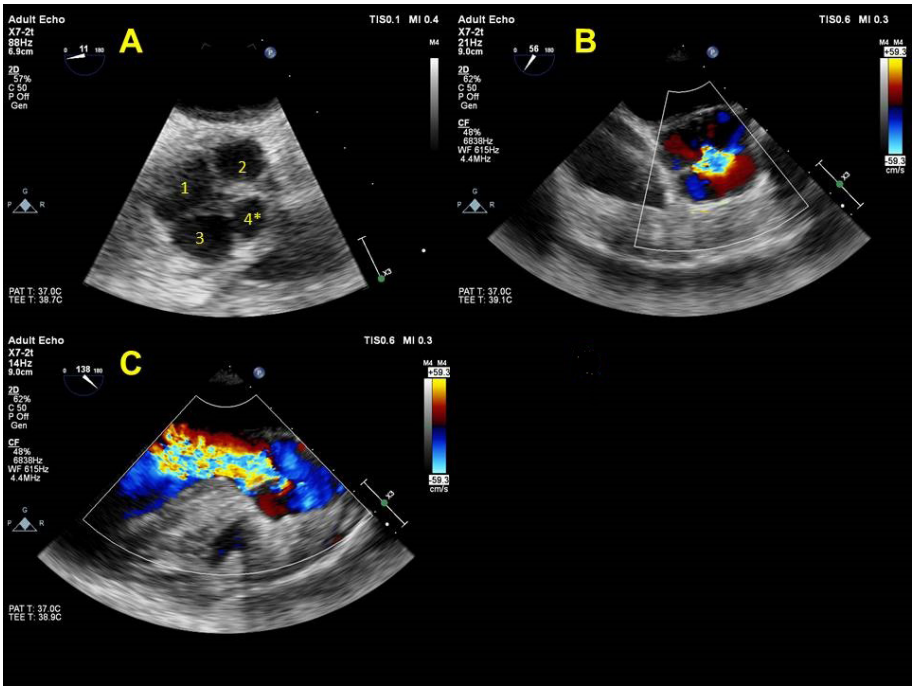

Figure 2 – *Intraoperative transesophageal echocardiography showing: A: quadricuspid aortic valve (cross-sectional view of medium-high esophagus at the aortic valve level); B: central jet of severe aortic regurgitation due to cusps coaptation failure (cross-sectional view of medium-high esophagus at the aortic valve level); C: severe aortic regurgitation (longitudinal view of medium-high esophagus).*

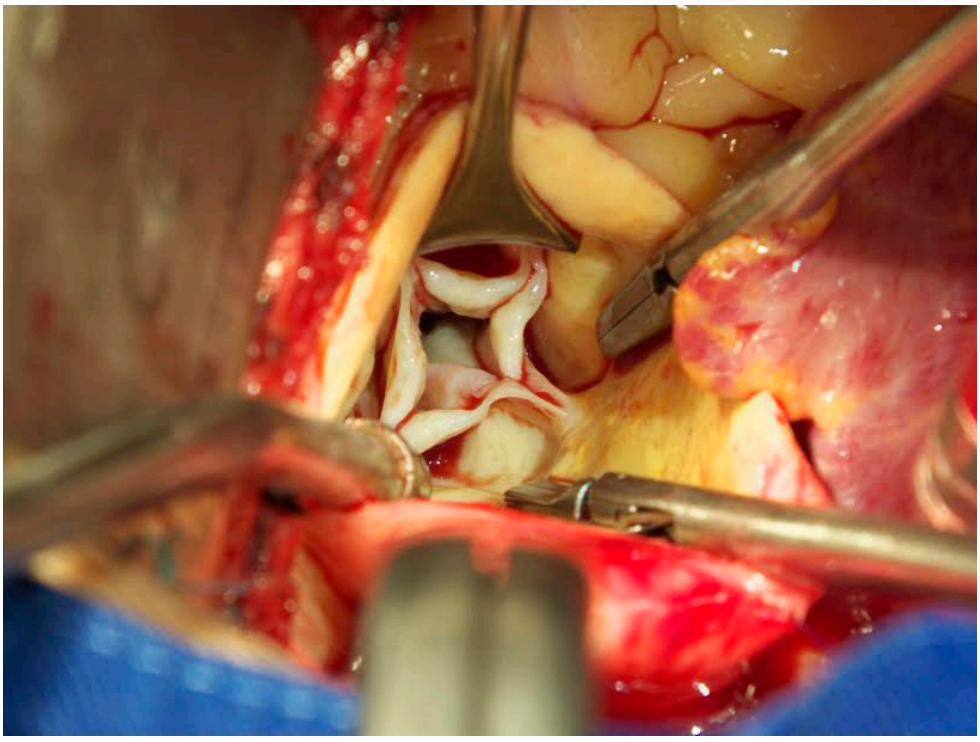

Figure 3 – *Photo of anatomical specimen, taken during surgery, showing the quadricuspid aortic valve.*

The patient in question had been diagnosed with severe aortic regurgitation, but in previous transthoracic scans, the presence of quadricuspid aortic valve had not been detected. The patient was then referred for elective aortic valve replacement. Intraoperative TEE in turn, revealed QAV (Figure 2) with 3 larger identical cusps higher and a smaller cusp, corresponding to type B in the Hurwitz and Roberts classification.[1] The accessory cusp was located between the left coronary and right coronary cusps (group I in the Nakamura classification).[4]

Authors' contributions

Research creation and design: Machado CCS; Data acquisition: Machado CCS, Lima MSM; Data analysis and interpretation: Machado CCS, Lima MSM, Brandão CMA, Veronese ET, Tsutsui JM, Mathias Jr W; Manuscript drafting: Machado CCS, Lima MSM; Critical revision of the manuscript as for important intellectual content: Machado CCS, Lima MSM, Brandão CMA, Veronese ET, Tsutsui JM, Mathias Jr W

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Potential Conflicts of Interest

There are no relevant conflicts of interest.

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Academic Association

This study is not associated with any graduate program.

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