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CASE REPORT

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Multimodal imaging in the assessment of quadricuspid aortic valve

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Abstract

Quadricuspid aortic valve (QAV) is a rare congenital anomaly of the aortic valve, with an incidence of 0.05-0.1%, often associated with aortic regurgitation. The condition typically presents between the ages of 46 and 50, with a slight male predominance. While diagnosis is generally made via transthoracic echocardiography (TTE), this method can occasionally fail to identify QAV, necessitating the use of transoesophageal echocardiography and cardiac computed tomography for more accurate assessment of valve morphology. We present the case of a 57-year-old male who experienced chest pain for three months. Although TTE revealed severe aortic regurgitation, it did not detect the QAV. The anomaly was ultimately identified through advanced imaging techniques prior to surgery, which confirmed the presence of this rare aortic valve morphology.

Keywords Quadricuspid aortic valve, Aortic regurgitation, Transthoracic echocardiography, Transoesophageal echocardiography, Cardiac CT

Case report

A 57-year-old male with a history of acute rheumatic fever in childhood presented with chest pain that had persisted for approximately three months, predominantly at night. Initial evaluation with transthoracic echocardiography (TTE) revealed severe aortic regurgitation and fibrotic thickening of the aortic leaflets. However, the TTE did not identify the presence of a quadricuspid aortic valve (QAV). The anomaly was subsequently diagnosed through cardiac computer tomography (CT) which provided a detailed assessment of the aortic anatomy, revealing an aortic annulus size ranging from 27 mm, an aortic root diameter of 35 mm, and a sino-tubular junction diameter of 37 mm. The patient was scheduled for aortic valve replacement. Intraoperative transoesophageal echocardiography (TEE) was performed and confirmed the findings of the cardiac CT scan. The definitive confirmation of the QAV morphology was made by the surgeon after aortotomy. The patient had an uneventful postoperative course and was discharged without complications (Figs. 1 and 2).

Discussion

Quadricuspid aortic valve (QAV) is a rare congenital anomaly, with a reported incidence of 0.05-0.1% [1] and a prevalence in the general population ranging from 0.013–0.043% [2]. It is frequently associated with aortic regurgitation (AR), and patients typically present between the ages of 45 and 50, with a slight male predominance [3]. While transthoracic echocardiography (TTE) is the most common diagnostic tool, it can occasionally miss the diagnosis, necessitating the use of transoesophageal

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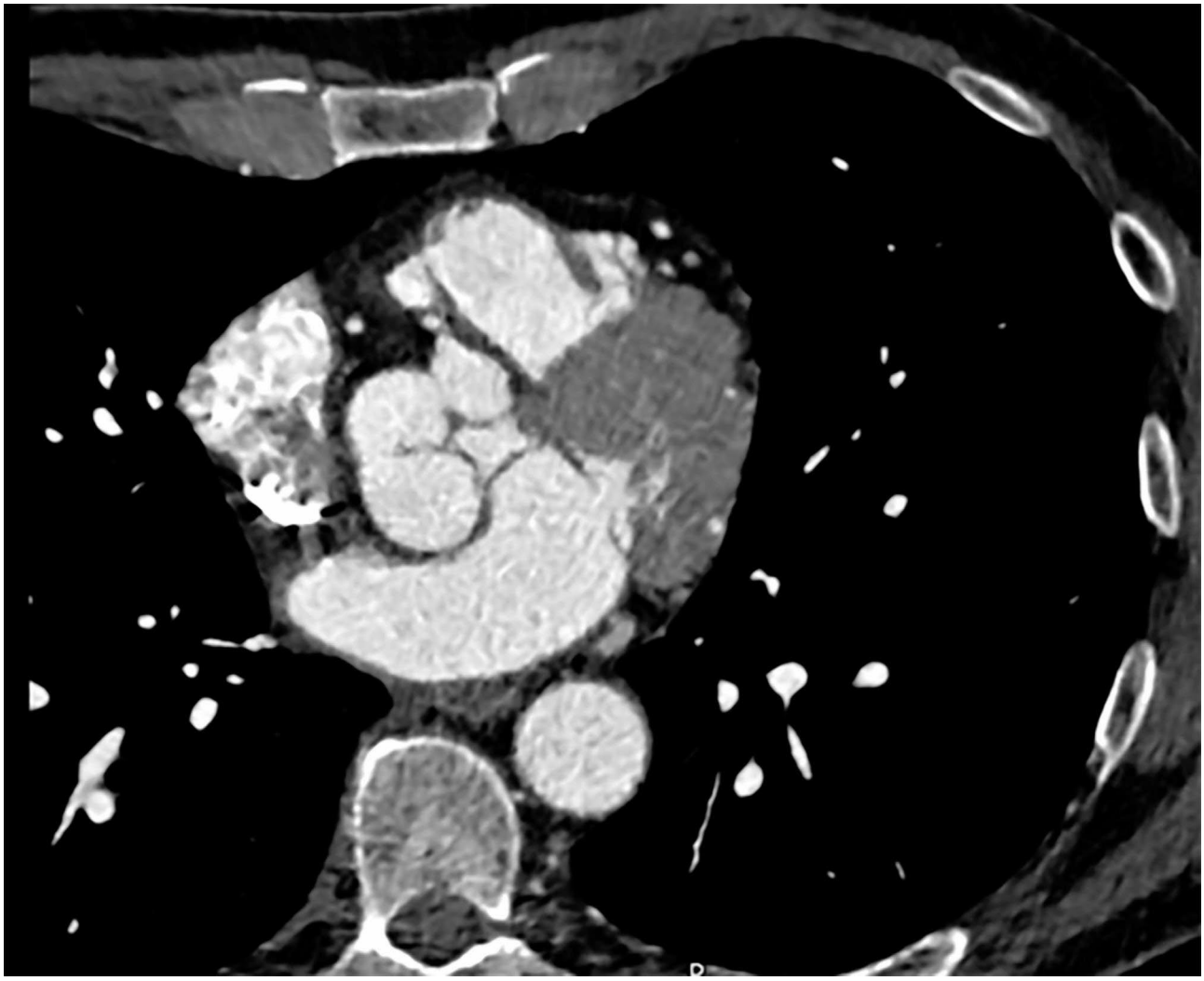

Fig. 1 Cardiac TC, axial projection showing the quadricuspid aortic valve with asymmetric leaflets

echocardiography (TEE) or cardiac CT scan for more accurate assessment of the valve morphology [4].

The embryogenesis of QAV remains unclear, but it is believed to result from abnormal septation of the conotruncus or division of one of the three mesenchymal ridges that normally form the aortic valve cusps [5, 6]. True QAVs should be differentiated from pseudo-QAVs, which can result from conditions like bacterial endocarditis or rheumatic fever [6].

QAV was first identified at autopsy in 1862, and the first in vivo description was provided in 1968. Although QAV often presents as an isolated anomaly, it can be associated with other congenital heart defects in 18-32% of cases, including coronary artery anomalies, atrial septal defects, and ventricular septal defects [1, 7]. The most common complication of QAV is aortic regurgitation, but aortic valve stenosis and infective endocarditis have also been reported, albeit less frequently [7, 8].

Preoperative identification of QAV is crucial for recognizing associated anomalies that may impact surgical planning [9]. While TTE allows for early detection, its sensitivity may be insufficient, as demonstrated in this case report where the QAV was identified only after further imaging with cardiac CT and confirmed during surgery by TEE. Cardiac CT is particularly useful for visualizing valve anatomy, including leaflet coaptation and coronary ostia, which are essential for planning surgical intervention.

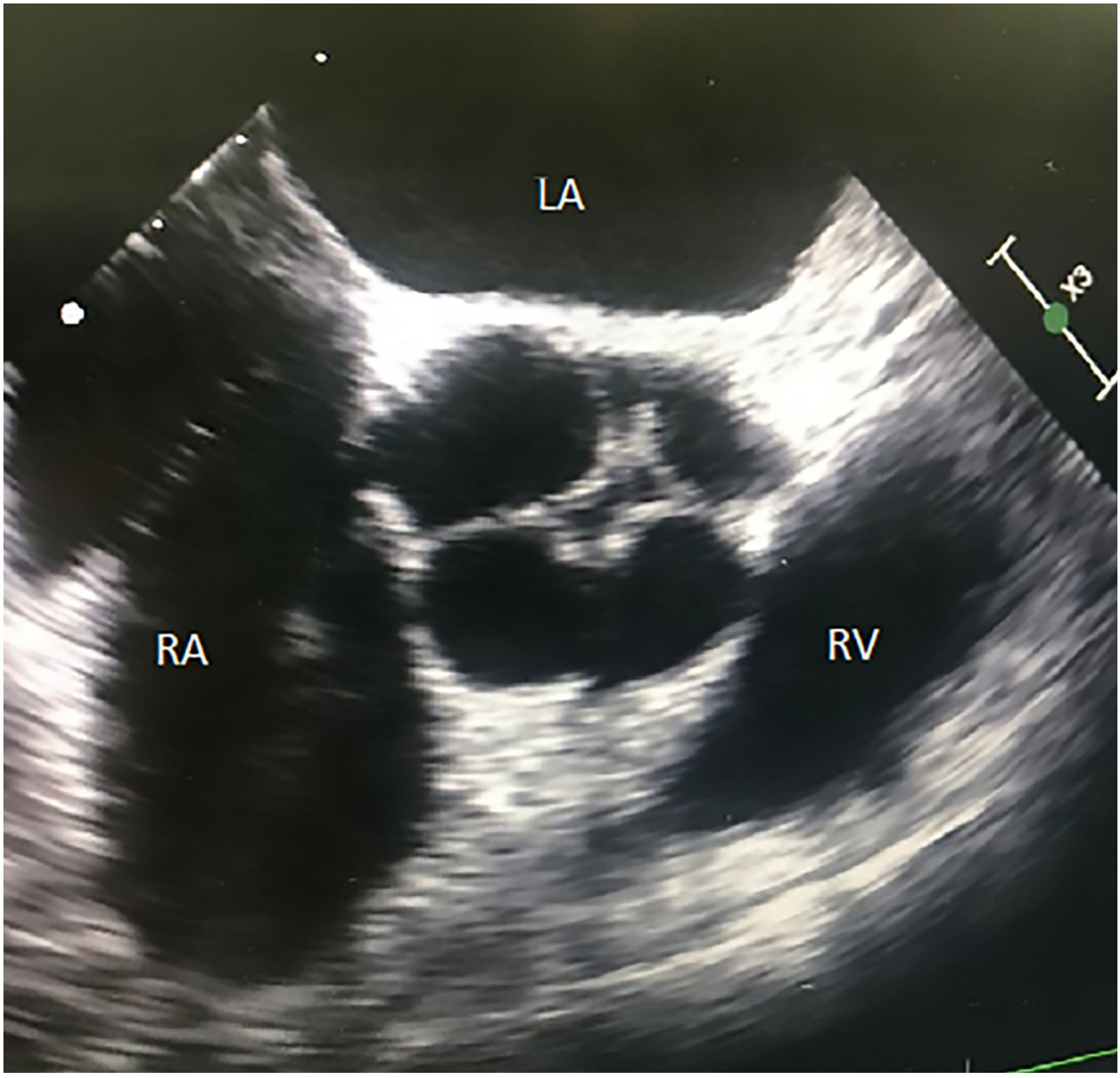

Fig. 2 Transesophageal echocardiography showing quadricuspid aortic valve. LA: left atrium, RA: right atrium, RV: right ventricle

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Competing interests
The authors declare no competing interests.

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References

1. Yuan SM. Quadricuspid aortic valve: A comprehensive review. brazilian journal of cardiovascular surgery. Published online 2016. https://doi.org/10.5935/1678-9741.20160090
2. Feldman BJ, Khandheria BK, Warnes CA, Seward JB, Taylor CL, Tajik AJ. Incidence, description and functional assessment of isolated quadricuspid aortic valves. Am J Cardiol. 1990;65(13):937–8. https://doi.org/10.1016/0002-9149(90)91446-d.
3. Savino K, Quintavalle E, Ambrosio G. Quadricuspid aortic valve: A case report and review of the literature. J Cardiovasc Echography. 2015;25(3):72. https://doi.org/10.4103/2211-4122.166077.

4. Godefroid O, Colles P, Vercauteren S, Louagie Y, Marchandise B. Quadricuspid aortic valve: a rare etiology of aortic regurgitation. Eur J Echocardiogr. 2006;7(2):168–70. https://doi.org/10.1016/j.euje.2005.03.011.
5. Fernández B, Durán AC, Martire A, López D, Sans-Coma V. New embryological evidence for the formation of quadricuspid aortic valves in the Syrian hamster (Mesocricetus auratus). J Comp Pathol. 1999;121(1):89–94. https://doi.org/10.1053/jcpa.1998.0299.
6. Peretz DI, Changfoot GH, Gourlay RH. Four-cusped aortic valve with significant hemodynamic abnormality. Am J Cardiol. 1969;23(2):291–3. https://doi.org/10.1016/0002-9149(69)90081-2.
7. Tsang MYC, Abudiab MM, Ammash NM, et al. Quadricuspid Aortic Valve: Characteristics, Associated Structural Cardiovascular Abnormalities, and Clinical Outcomes. Circulation. 2016;133(3):312–9. https://doi.org/10.1161/CIRCULATIONAHA.115.017743.
8. Takeda N, Ohtaki E, Kasegawa H, Tobaru T, Sumiyoshi T. Infective endocarditis associated with quadricuspid aortic valve. Jpn Heart J. 2003;44(3):441–5. https://doi.org/10.1536/jhj.44.441.
9. Chapman CB, Kohmoto T, Kelly AF, Thornton F, Keevil JG. Cardiac computed tomography and quadricuspid aortic valve: a case report. WMJ. 2010;109(4):219–21.

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