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Quadricuspid aortic valve with aortic regurgitation: a rare echocardiographic finding.

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CASE REPORT

Quadricuspid aortic valve with aortic regurgitation: a rare echocardiographic finding

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SUMMARY

We report on a middle-aged woman treated for chronic hepatitis C virus infection with pegylated interferon. Auscultation revealed a diastolic murmur and the peripheral signs of aortic regurgitation. She had shortness of breath on moderate exertion for the past 4 months, which she attributed to her liver disease. Echocardiogram showed a quadricuspid aortic valve with severe aortic regurgitation. She was referred to a cardiothoracic surgeon for aortic valve replacement (AVR). However, she decided against AVR despite detailed counselling, and opted for medical treatment.

BACKGROUND

A quadricuspid aortic valve (QAV) is an uncommon clinical entity.[1 2] The prevalence of QAV ranges between 0.013% and 0.043%.[3] In this patient QAV was accurately visualised on transthoracic echocardiogram (figure 1).

CASE PRESENTATION

A 46-year-old woman was being treated for chronic active hepatitis C virus infection with pegylated interferon. She had shortness of breath on exertion for 4 months, which she attributed to her liver disease. On examination, she had a pulse rate of 82 beats/min, with collapsing character, blood pressure of 140/55 mm Hg, dancing carotids, jugular venous pressure was not raised and there was no pedal oedema. The apex beat was palpable in the sixth intercostals space 1 cm lateral-to-mid clavicular line, with thrusting character. On auscultation an early diastolic murmur at the aortic area and a soft S2 was detected. She was referred for an echocardiogram.

INVESTIGATIONS

The transthoracic echocardiogram showed a QAV with all four leaflets equal in size (figure 2), thickened, calcified and noncoapting. There was severe aortic regurgitation with effective regurgitant orifice 0.35 cm^2, pressure half time 200 ms (figures 3 and 4) and vena contracta 0.6 cm in the absence of aortic stenosis. All remaining valves were normal. The left ventricle was mildly dilated with low normal systolic function (left ventricular ejection fraction was approximately 50%) and mild global hypokinesia. Left ventricular end-systolic and end-diastolic internal dimensions were 44 mm and 58 mm, respectively. Coronary artery anomalies could not be excluded, as the patient was not keen to undergo further cardiac evaluation.

TREATMENT

Given the symptoms and echocardiographic findings the patient was referred to a cardiothoracic surgeon for aortic valve replacement (AVR). She decided against the procedure despite detailed counselling by the cardiologist and cardiothoracic surgeon and opted for medical therapy.

OUTCOME AND FOLLOW-UP

She continues to have shortness of breath on moderate exertion, but is able to perform routine activities.

DISCUSSION

QAV is an uncommon congenital cardiac anomaly that is usually found incidentally at the time of open heart surgery or at autopsy.[1] First described in the literature by Balington in 1862,[2] the prevalence currently ranges between 0.13% and 0.043%.[3] In

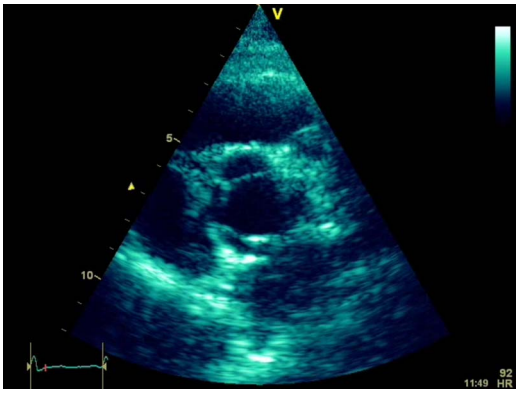

Figure 1 Echocardiographic image, left parasternal short-axis view at aortic valve level showing quadricuspid aortic valve.

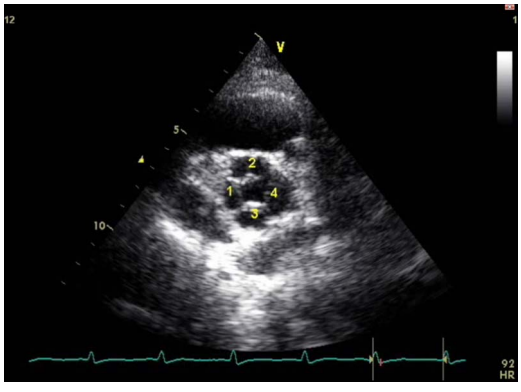

Figure 2 Echocardiographic image, left parasternal short-axis view at aortic level showing four equal size cusps of aortic valve.

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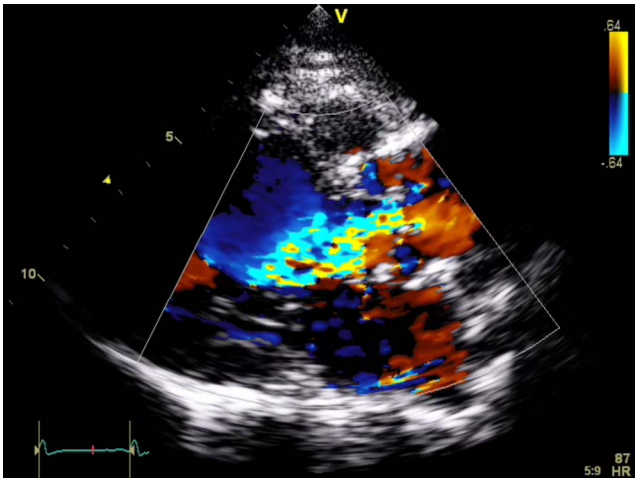

Figure 3 Echocardiographic image, left parasternal long-axis view with colour Doppler showing aortic regurgitation.

recent years, a few case reports of QAV have been published.[2–6] The most comprehensive review of the literature has been published by Tutarel.[7] Associated coronary artery anomalies have been found in 10–30%[8 9] and other abnormalities associated with QAV include subaortic stenosis, pulmonic valve stenosis and ventricular septal defect.[2 7 10 11] Few case reports of bacterial endocarditis affecting a QAV have been published as well.[12] Males and females are equally affected. Developmentally, both semilunar valves are derived from mesenchymal swellings in the pulmonary and aortic trunks after the partition of truncus. In normal embryological development, there are three swellings that bulge into the lumen of each great vessel and eventually develop into three semilunar cusps that coapt in the centre of the valve orifice. It has been hypothesised that an abnormal number of cusps results from developmental changes in the early stages of truncal partition, leading either to a dissymmetry in the number of primordial valve cusps or an abnormal proliferation of these mesenchymal tissue buds. Most patients of QAV require AVR, but aortic valve repair has also been reported.[13] In 1973, Hurwitz and Roberts[4] classified the QAV according to anatomic variation of cusp size. They have described seven

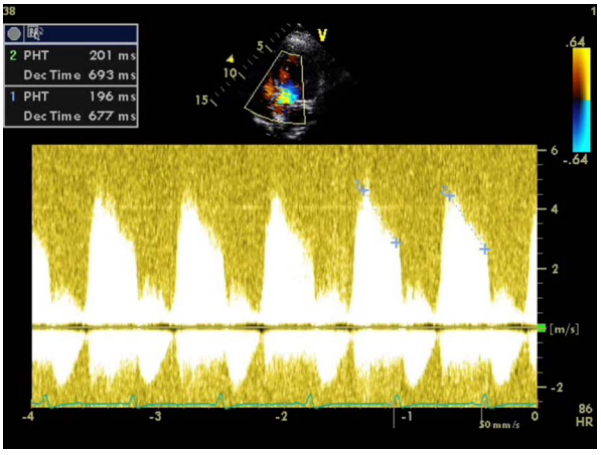

Figure 4 Echocardiographic image, continuous-wave Doppler at left ventricular outflow tract showing aortic regurgitation pressure half time of 200 ms.

varieties: (A) four equal cusps; (B) three equal cusps and one smaller cusp; (C) two equal larger and two equal smaller cusps; (D) one large, two intermediate and one small cusp; (E) three equal cusps and one larger cusp; (F) two equal larger and two unequal smaller cusps and (G) four unequal cusps.[4] The most frequent positions of the accessory cusp are located in two positions, either between the right and non-coronary cusps or between the right and left coronary cusps.[10] In accordance with these reports, our patient was diagnosed with four equal cusps (type A in Hurwitz and Roberts classification). Identification of a QAV in aortic insufficiency is very important due to progressive degeneration of the leaflets because of asymmetric mechanical stress around the four cusps and high risk of infective endocarditis. Advancement in imaging techniques especially echocardiogram has increased the capability to diagnose this congenital aortic valve disorder.[14] Newer imaging modalities like cardiac magnetic resonance also plays a pivotal role in the diagnosis of QAV. Significant aortic regurgitation due to poor coaptation of the leaflets is more frequent than aortic stenosis and valve replacement, which is commonly required in the fifth or sixth decade of life.[15 16]

Learning points

- A quadricuspid aortic valve (QAV) is an uncommon clinical entity and the first case was reported in 1862.
- QAV predominantly causes aortic regurgitation.
- There are seven varieties of QAV. In our patient all four cusps were of equal size (type A in Hurwitz and Roberts classification).
- Aortic valve replacement is the definite treatment for patients with QAV and severe aortic regurgitation.

Patient consent Obtained.

Competing interests None.

Provenance and peer review Not commissioned; externally peer reviewed.

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