

Rare Type C Quadricuspid Aortic Valve Presenting with Aortic Stenosis and Aortic Insufficiency

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Citation

A Mahal, P Gupta, W Hunter, J Sugimoto. *Rare Type C Quadricuspid Aortic Valve Presenting with Aortic Stenosis and Aortic Insufficiency*. The Internet Journal of Cardiology. 2009 Volume 8 Number 1.

Abstract

Quadricuspid aortic valve (QAV) is a rare congenital anomaly that often comes to clinical attention due to aortic regurgitation. The incidence of QAV has been reported between .008 to .043 percent making it the rarest malformation of the aortic valve.¹⁻² We report a case on a patient who presented with heart failure secondary to severe aortic stenosis and moderate aortic regurgitation and underwent aortic valve replacement. This is the second report of a Type C QAV i.e., two equal larger cusps and two equal smaller cusps, presenting with both aortic regurgitation and aortic stenosis.

CASE REPORT

A 73 year old white woman with known aortic stenosis, congestive heart failure, hypertension and paroxysmal atrial fibrillation presented to the Emergency Room with increasing shortness of breath, orthopnea, and fatigue. Transthoracic echocardiogram revealed an ejection fraction of 25 - 30% with an aortic valve area of 0.86 cm², mean gradient of 27mmHg and moderate aortic regurgitation.

The patient underwent aortic valve replacement. Findings at operation were a quadricuspid valve with two "larger" and two "smaller" cusps (Figure 1). There were mild calcifications and slightly fused commissures. The patient recovered well from the procedure and was released for home rehabilitation.

Figure 1

Figure 1



DISCUSSION

Quadricuspid aortic valves (QAV) were first described in 1862 by Balington.³ A recent survey of articles, in 2004, cited 186 reported cases.⁴ We performed a PubMed search including articles up to December of 2008 and found that 254 cases had been reported.

Aortic regurgitation (AR) is the most common hemodynamic abnormality associated with QAV.^{5,6} Approximately 74.7 % of patients will present with AR. However, only 8.4% of patients present with combined regurgitation and stenosis.² Very few, less than 1%, present with aortic stenosis alone.⁴ Hurwitz and Robert classified quadricuspid aortic valves into seven groups.⁷ The valve

identified in this case was type C, i.e., two equal larger cusps and two equal smaller cusps. Up to December of 2008, only fourteen cases of type C had been reported. This is the second case in which the patient presented with documented combined aortic stenosis and regurgitation in a type C QAV.³

QAV was first diagnosed by two-dimensional echocardiography in 1984 by Herman et al.⁸ His description has led to the X sign being pathognomonic for QAV in short axis view of the aorta. Since 1980, 60% of QAV have been identified by echocardiography.³ QAV has also been diagnosed by multi-slice computed tomography.^{9,10} Our patient had three echocardiographic examinations prior to her surgery, none of which identified the valve as quadricuspid. On retrospective review of the final echocardiogram, the anomaly was visualized.

Some attempts at surgical repair of QAV have been found in the literature.^{11,12} Long term outcomes, however, have not been described. Aortic valve replacement is the standard of care.²

CONCLUSION

This case and review of the literature suggest that QAV remains a rare cause of hemodynamic disturbance, usually presenting with aortic regurgitation but occasionally presenting with a combined AR and AS. Quadricuspid aortic valve is diagnosed more frequently now, possibly due to increased use of imaging studies. This is a unique case of mixed aortic stenosis and regurgitation with a type C valve, undiagnosed by echocardiography. This is only the second time this has been reported in the literature. We offer this

report to augment the growing literature regarding QAV and for use for future investigation of QAV risks, incidence and prevalence.

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