Quadricuspid Aortic Valve Associated with Aortic Insufficiency Contributors

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Abstract

A 51-year-old male presented with a wound in his right hand that was suspicious for possible septic emboli of cardiac origin. With transesophageal echocardiography, the patient was found to have a rare quadricuspid aortic valve. This quadricuspid valve can present with variable symptoms and physical exam findings. Due to embryological defects, this pathology is associated with several other anatomical defects that are important to recognize prior to surgical intervention. Transesophaegeal echocardiography remains the gold standard in detection of quadricuspid aortic valve and identification of other possible cardiac lesions.

Keywords: *Aortic valve, quadricuspid valve, transesophageal echocardiography*

History

A 51-year-old male presented with a nonhealing wound of several weeks on his fourth and fifth digits in his right hand. On physical examination, the patient was found to have a diastolic murmur in the aortic region, while the hand wound was concerning for possible septic emboli from the cardiac origin. The patient denied any complaints of chest pain, dyspnea, and palpitations. The patient underwent transesophageal echocardiography to examine for possible cardiac lesions. Incidentally, it was found to have a quadricuspid aortic valve associated with severe aortic insufficiency seen in Figures 1 and 2.

Differential Diagnosis

Aortic regurgitation, endocarditis, ankylosing spondylitis.

Discussion

Quadricuspid aortic valve is one of the rarest forms of valvulopathy documented with an incidence around 0.01%–0.04%.[1] It has been hypothesized that during the 5th week of embryogenesis, disruption of the mesenchymal ridges from fusing leads to this presentation.[2] This valvulopathy has been documented to have a predominance for men, with a mean presenting age between 45 and 60 years of age.[3] Although the patient presented without any revealing symptoms, the quadricuspid aortic valve has been reported with chest pain, dyspnea, palpitations, and syncope.[4] Importantly, this anomaly is associated with other anatomical abnormalities such as aberrant coronary arteries, ventricular septal defects, patent ductus arteriosus, and pulmonary stenosis. Although this patient

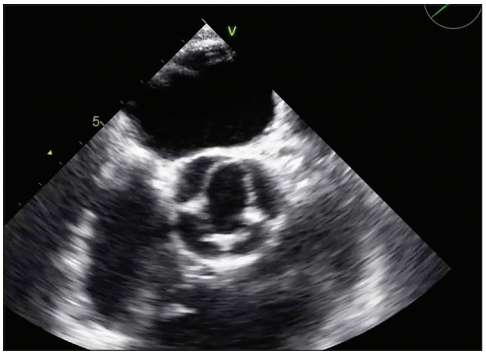

Figure 1: Transesophageal echocardiogram at mid-esophageal aortic valve short axis view in systole

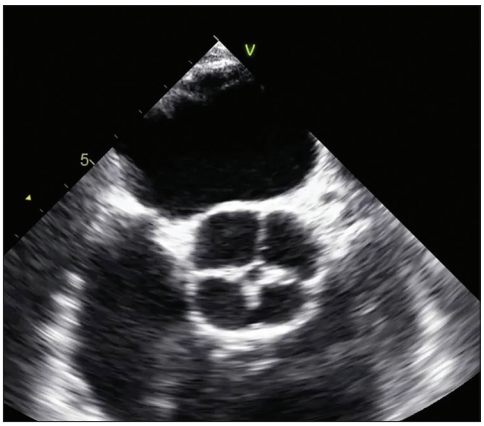

Figure 2: Transesophageal echocardiogram at mid-esophageal aortic valve short axis view in diastole

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had severe aortic insufficiency, it is imperative to evaluate for these possible abnormalities before valve surgery.[3] Transesophageal echocardiography is considered to be the gold standard for the detection of this rare valve anomaly, as well as its associated anatomical cardiac defects.[5] Of those with quadricuspid aortic valve associated with aortic insufficiency, 50% of that population will require surgical intervention in their lifetime.[1] However, this patient was lost to follow-up.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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