Quadricuspid Aortic Valve by Transesophageal Echocardiography

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Quadricuspid aortic valves (QAVs) are a rare congenital anomaly associated with increased risk of aortic insufficiency. This case presents the incidental finding of a QAV on intraoperative transesophageal echocardiography (TEE) after going undetected on transthoracic echocardiography multiple times, suggesting that TEE may be a superior imaging modality for the identification of this defect. This patient with a history of coronary artery disease presented with sudden onset moderate to severe aortic insufficiency and required subsequent aortic valve replacement (AVR).

Keywords: *quadricuspid aortic valve; aortic valve; aortic insufficiency; aortic regurgitation; transesophageal echocardiography*

INTRODUCTION

A 65-year-old woman with a known history of moderate aortic insufficiency was found to have hypertensive urgency and diagnosed with pulmonary embolism shortly after undergoing a mandibulectomy for squamous cell carcinoma. She was found to have an acute coronary syndrome and subsequent transthoracic echocardiogram (TTE) revealed a left ventricular ejection fraction of 35%, though the quadricuspid aortic valve (QAV) was not readily visualized at this time. She then underwent left cardiac catheterization, which demonstrated severe aortic insufficiency and 80% stenosis of the right coronary artery. Her aortic root measured 33 mm on preoperative CT angiography with the ascending aorta measuring 29 mm. Patient was taken to the operating room and underwent single coronary artery bypass grafting and aortic valve replacement (AVR).

During this procedure, transesophageal echocardiography (TEE) revealed the patient to have a four-leaflet aortic valve as shown in Fig. 1. The valve

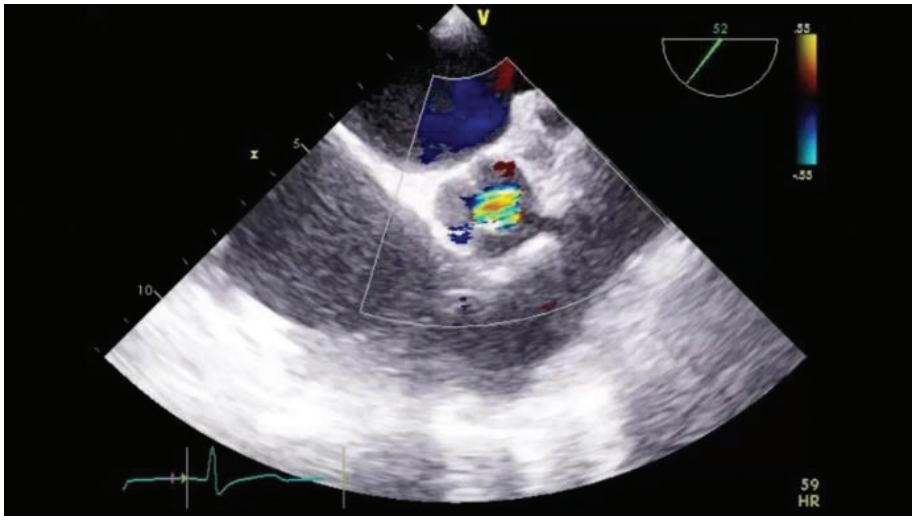

Figure 1. Short-axis view of quadricuspid aortic valve on intraoperative TEE revealing severe aortic regurgitation.

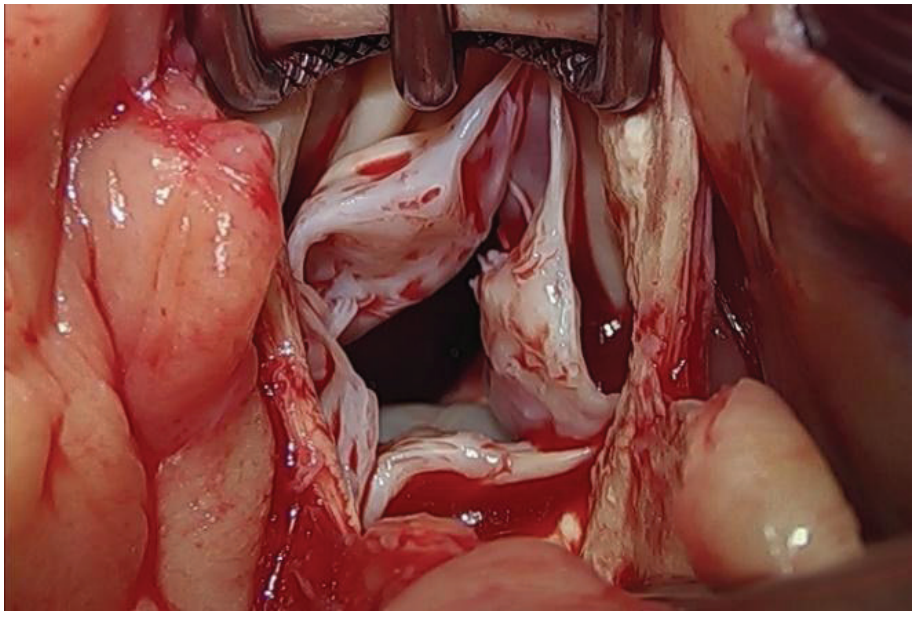

Figure 2. Intraoperative view of quadricuspid aortic valve with four equal-sized cusps consistent with type A on Hurwitz and Roberts classification system.

had four equal-sized cusps, as shown in Fig. 2, consistent with type A on the Hurwitz and Roberts classification system.[1] AVR was performed with a bovine pericardial tissue valve. The patient was discharged home without associated complications on postoperative day 20.

DISCUSSION

QAVs are a very rare congenital cardiac anomaly that usually presents in isolation but occasionally presents with other defects. The bicuspid aortic valve is the most common aortic anomaly, present in 2% of the population, followed by the unicuspid aortic valve.[2,3] QAVs have an estimated prevalence of 0.008–0.033% by autopsy and 0.013–0.043% by echocardiography.[1,4] The first QAV was incidentally discovered in 1862 and less than 300 cases have been described since this time.[5]

Studies on Syrian hamsters have suggested that QAVs arise from an extra partition in one of the three mesenchymal swellings that form cusps during early development.[6,7] Other studies propose that the anatomy of malformed semilunar valves tends to suggest an alteration later in development, such as further septation of a normal valve cushion, as opposed to a malformation.[8] Embryological formation of QAVs remains largely unknown.

QAVs are classified into seven different types based on a schematic devised by Hurwitz and Roberts. Of these, type A (four equal cusps), type B (three equal cusps and one smaller cusp), and type C (two equal larger cusps and two equal smaller cusps) are the most common presentations of QAV as shown in Fig. 3.[5] Our patient demonstrated a type A QAV based on this classification system, composed of four equal-sized cusps.

QAVs were historically detected primarily during surgery or at autopsy. They are now most often detected via echocardiography, followed by surgery, at autopsy, and by other diagnostic means such as computed tomography angiogram or magnetic resonance imaging. TTE is used as a primary screening tool; however, the advent of TEE has enabled increased diagnostic accuracy.[9,10] TEE gives the transducer better access to image the aortic valve with less structures between it and the valve, as opposed to TTE that must penetrate lung and chest wall tissue.[10] This study provides one example of TTE failing to detect a QAV, which was then detected on TEE. Screening with TEE rather than TTE may increase the diagnosis of QAVs, allowing for better preparation and planning prior to surgical management. TEE may also provide a better assessment of valve morphology and measurement of other parameters useful for surgery, ultimately improving patient

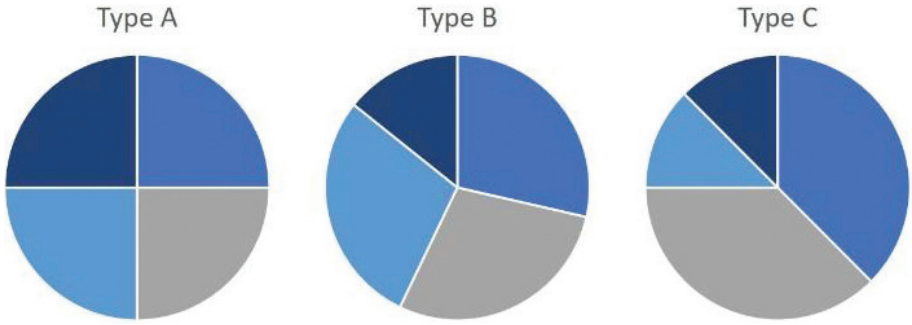

Figure 3. Hurwitz and Roberts classification of the most common types of quadricuspid semilunar valves: Type A (four equal cusps), type B (three equal cusps and one smaller cusp), and type C (two equal larger cusps and two equal smaller cusps).

outcomes without requiring magnetic resonance imaging or computed tomography angiograms.[5,11]

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