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Case Report

Quadricuspid Aortic Valve by Transesophageal Echocardiography

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Abstract

Quadricuspid aortic valves (QAVs) are a rare congenital anomaly associated with increased risk of aortic insufficiency. This case presents the incidental finding of a quadricuspid aortic valve on intraoperative transesophageal echocardiography after going undetected on transthoracic echocardiography multiple times, suggesting that transesophageal echocardiography 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 AVR.

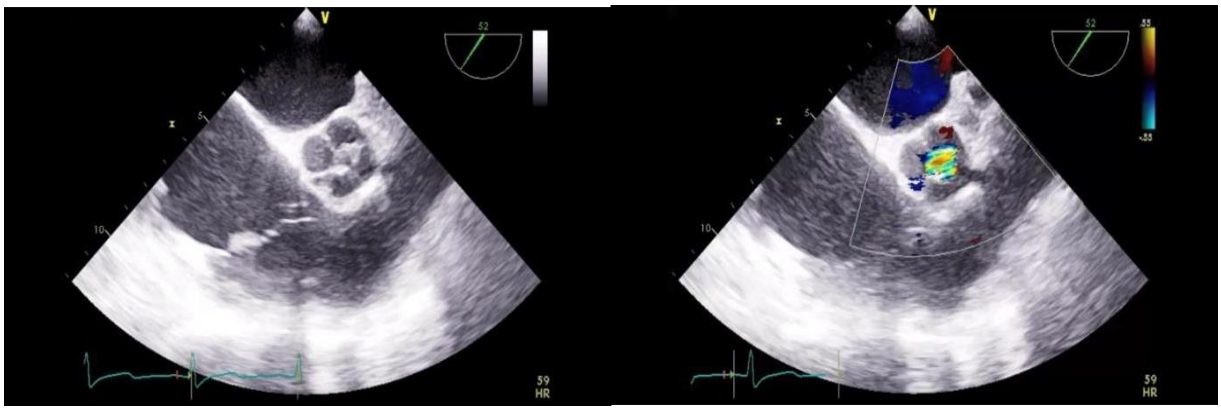
Keywords: quadricuspid; aortic; valve; echocardiography; transesophageal

Introduction

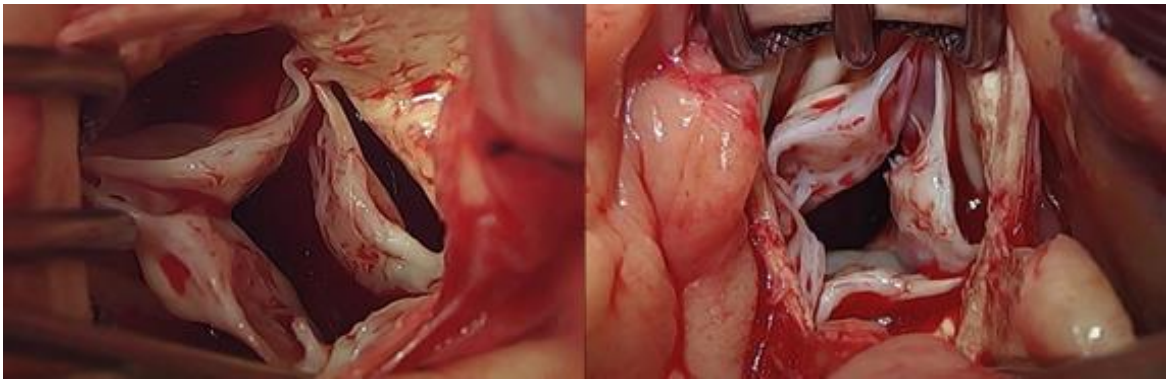
A 65-year-old female was found to have hypertensive urgency and diagnosed with pulmonary embolism shortly after undergoing a mandibulectomy for squamous cell carcinoma. She was found to have an acute coronary syndrome and subsequent transthoracic echocardiogram revealed an LVEF of 35%, though the quadricuspid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

Case Presentation

During this procedure, transesophageal echocardiography revealed the patient to have a four-leaflet aortic valve as shown in Figures 1a and 1b. The valve had four equal-sized cusps, as shown in Figures 2a and 2b, consistent with type A on Hurwitz and Roberts classification system.1 Aortic valve replacement was performed with a bovine pericardial tissue valve. The patient was discharged home without associated complication on postoperative day 20.



Figures 1a and 1b: Short-axis view of quadricuspid aortic valve on intraoperative TEE revealing severe aortic regurgitation.



Figures 2a and 2b: Intraoperative view of quadricuspid aortic valve with 4 equal-sized cusps consistent with type A on Hurwitz and Roberts classification system.

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).

Quadricuspid aortic valves are a very rare congenital cardiac anomaly that usually present in isolation, but occasionally present 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 Figure 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. Transthoracic echocardiography is used as a primary screening tool, however the advent of transesophageal echocardiography 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 which 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 diagnosis of QAVs, ultimately improving patient outcomes without requiring magnetic resonance imaging or computed tomography angiograms [5].

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