Türk Göğüs Kalp Damar Cerrahisi Dergisi 2017;25(1):156-158
http://dx.doi.org/doi: 10.5606/tgkdc.dergisi.2017.13920

Quadricuspid aortic valve with severe aortic regurgitation

İleri aort yetmezliği ile seyreden kuadrikuspit aort kapağı

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Quadricuspid aortic valves have a reported incidence in literature of 0.01-0.04% of the general population. It may present as an isolated valve abnormality or in association with other congenital heart defects. In 1973 Hurwitz and Roberts, described and classified the quadricuspid semilunar valves into seven different groups (from A to G) according to the morphological characteristics of the valve. The precise aetiology of quadricuspid aortic valves remains unknown. We report the case of a 49-year-old female, who presented to our center with increasing dyspnea over a four-year period associated with hospital admissions due to pulmonary edema. She did not report fever or chest pain. Her past medical history included well-controlled hypertension and she was a former smoker. On examination, auscultation revealed a diastolic murmur loudest in the second right intercostal space. Transthoracic echocardiography (TTE) showed severe aortic regurgitation. The thoracic aorta was normal and the ejection fraction was estimated at 65%. There was trivial regurgitation from the mitral valve with normal tricuspid and pulmonary valves. A coronary angiogram demonstrated normal coronary arteries. Further TTE imaging demonstrated a quadricuspid aortic valve (QAV). Consequently, she was closely observed and followed up with regular echocardiography. The patient was later scheduled for an elective aortic valve replacement due to increasing dyspnea with a New York Heart Association functional classification score of three.

Our patient underwent an uncomplicated elective aortic valve replacement. Visual examination and transesophageal echocardiography (TEE) of the aortic valve intraoperatively confirmed a QAV (Figures 1 and 2). The native valve was replaced with a mechanical aortic valve of Sorin Bicarbon FitLine

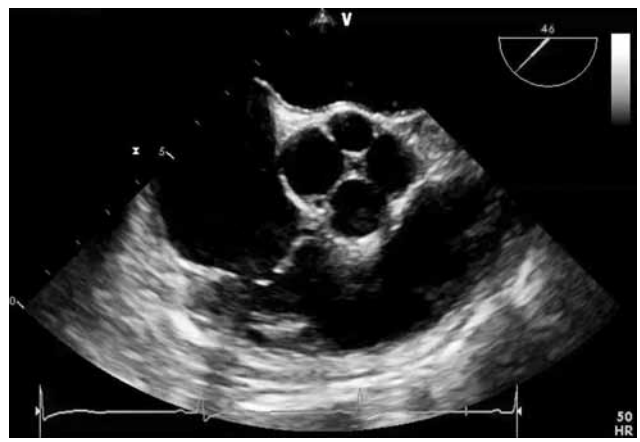

Figure 1. Intraoperative two-dimensional transesophageal echocardiography image showing X-shaped commissure pattern, characteristic of a quadricuspid aortic valve.

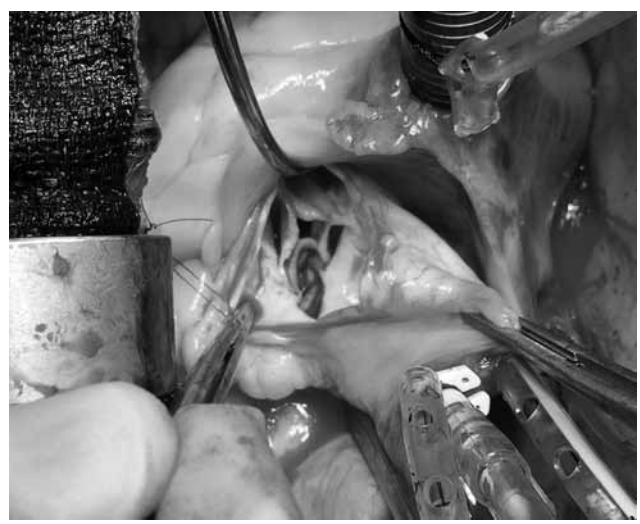

Figure 2. Intraoperative image demonstrating quadricuspid aortic valve *in situ*.

Available online at
www.tgkdc.dergisi.org
doi: 10.5606/tgkdc.dergisi.2017.13920
QR (Quick Response) Code

Received: September 22, 2016 *Accepted:* October 18, 2016

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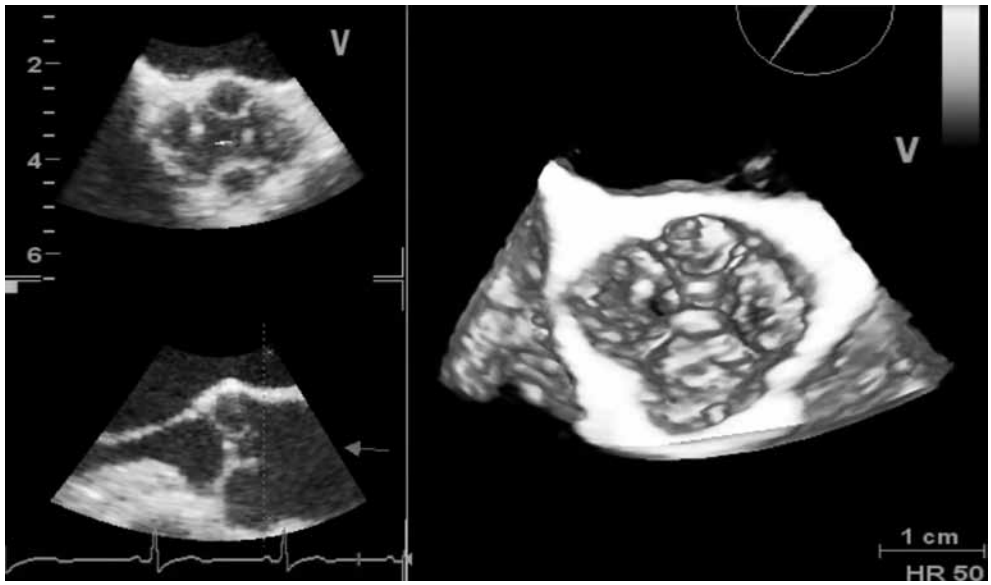

Figure 3. Intraoperative three-dimensional transesophageal echocardiography image showing quadricuspid aortic valve.

21 mm type (Sorin Biomedica Cardio SpA, Saluggia, Italy).

Postoperatively, she was transferred to the intensive care unit where her recovery was unremarkable. She was discharged on day fifth postoperative day on warfarin with marked symptomatic improvement.

DISCUSSION

Quadricuspid aortic valves remain a rare congenital cardiac abnormality with an incidence of 0.001-0.04% on echocardiography.[1] It may occur independently or in association with a number of other congenital cardiac abnormalities.

Many theories have been proposed regarding the etiology of QAVs. The semilunar valves develop from two mesenchymal ridges (known as truncoconal ridges). These ridges, then, descend to form the aorticopulmonary septum. Three mesenchymal swellings, then, develop at the junction of the conus and truncus, these swellings grow to form three triangular valve cusps.[2] This process is largely complete by week nine of gestation.[2] Quadricuspid aortic valves are thought to result from an embryological malformation, the process of which is not fully known. Previous hypotheses include a division of one of the mesenchymal ridges.[3] or an abnormal septation of the conotruncus.[4] thus creating a four-leaflet valve.

Several different anatomical variations have been described in the literature. Hurwitz and Roberts in 1973.[4] classified QAVs into categories based on the size of the leaflets, the most common types being type A (where the four cusps are of equal size) and type B (where three cusps are of equal size and one cusp is proportionally smaller).[4] Our patient had a type B QAV.

Quadricuspid aortic valves commonly remain asymptomatic until the fourth to sixth decade, where they present with aortic regurgitation. It has been hypothesized that unequal distribution of sheer stress on the valve leaflets results in fibrosis and incomplete coaptation thus resulting in valve insufficiency.[2]

The increased use and quality of TEE and cardiac magnetic resonance imaging have led to increased awareness of this rare congenital abnormality. The characteristic echocardiogram findings are a rectangular appearance in systole and an X-shaped commissure pattern in the short-axis view in diastole.[5] The main stay of treatment is aortic valve replacement with replacement of the ascending aorta if dilated.

In conclusion, in young patients with aortic regurgitation, TEE should be performed to assess the aortic valve and determine its morphology. Regular follow-up and early intervention are recommended in those diagnosed with quadricuspid aortic before left ventricular failure occurs.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

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