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Qudricuspid Aortic Valve: A Case Report

Tamer Turan[1], **Neslihan Gülmez**[2]

[1]*Acibadem Bursa Hospital, Department of Cardiovascular Surgery, Bursa, Turkey*
[2]*Setbaşı Medical Center, Department of Cardiology, Bursa, Turkey*

ABSTRACT

Quadricuspid aortic valve is a rare congenital condition that frequently appears as an isolated congenital anomaly. This case report, with images, describes a 50 years old adult female who presented clinically with dyspnea and palpitations. Transthoracic echocardiography revealed, quadricuspid aortic valve, mild aortic regurgitation and slight mitral regurgitation. On CT angiographic examination, we showed that aortic valve had 4 equal cusps.

Key words: quadricuspid aortic valve, congenital anomaly, aortic valve, CT angiography

DÖRT YAPRAKÇIKLI AORT KAPAK: OLGU SUNUMU

ÖZET

Dört yaprakçıklı aort kapağı, genellikle tek başına ve nadir görülen doğumsal bir anomalidir. Bu görüntülü olgu sunumunda, 50 yaşında, çarpıntı ve nefes darlığı yakınması olan kadın hastanın, dört yaprakçıklı aort kapağına ait ekokardiyografi ve CT görüntüleri sunulmuş ve tartışılmıştır.

Anahtar sözcükler: aort kapak, dört yaprakçıklı aort kapak, doğumsal anomali, BT anjiyografi

Quadricuspid aortic valve is a rare congenital condition that frequently appears as an isolated congenital anomaly, but may also be associated with other malformations, especially with coronary anomalies (1). A review from an historical autopsy showed that the prevalence of isolated quadricuspid aortic valve was approximately 0.008%. But today, current technology enables non-invasive diagnosis in most cases and modern echocardiographic data bases presented the prevalence to be somewhat higher (0.013%- 0.043%) (2).

Clinically quadricuspid aortic valve is represented by aortic insufficiency which is rare before adulthood. The anatomy of the quadricuspid aortic valve is variable, according to the size of each individual cusp. The more frequent morphologic variations are as follows: 4 equal cusps, 3 equal cusps with one minor and 2 large cusps with 2 small ones (3).

Case

This study describes a 50 year old adult female who represented clinically history of dyspnea and palpitations for 5 years. She had no history of endocarditis or rheumatic disease and no coronary risk factor. On physical examination, her blood pressure was 130/60 mm-Hg and cardiac auscultation revealed a 2/6 diastolic murmur along the left sternal border. Twelve-leads electrocardiogram showed a sinus rhythm. Transthoracic echocardiography revealed, normal left ventricular ejection fraction, quadricuspid aortic valve (Figure 1), mild aortic regurgitation and slight mitral regurgitation. On CT angiographic examination, we showed that aortic valve had 4 equal cusps morphologically and there was no associated coronary anomalies (Figure 2). The patient is being followed under medical treatment.

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Correspondence: Tamer Turan • **Phone:** 0 (224) 270 43 76 • **E-mail:** tamert@superonline.com

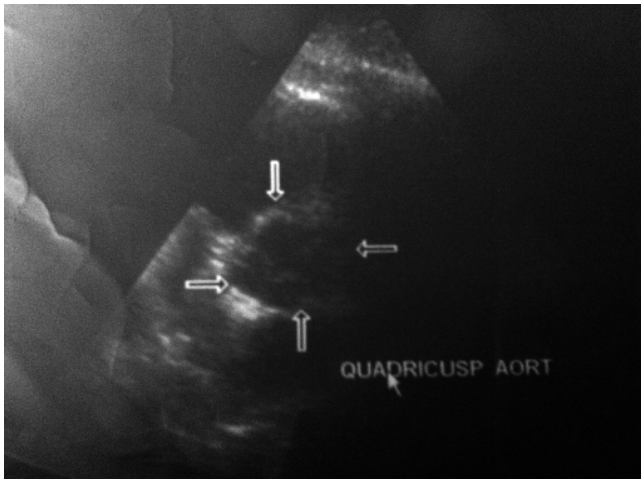

Figure 1. Echocardiographic view.

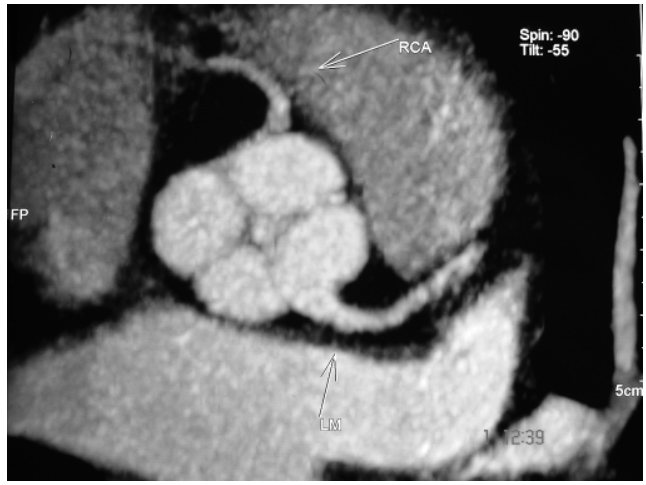

Figure 1. CT Angiographic view.

Discussion

The quadricuspid aortic valve is a rare manifestation of congenital aortic valve abnormalities. The anatomy of the quadricuspid aortic valve is variable, according to the size of each individual cusp. The more frequent morphologic variations are as follows: 4 equal cusps, 3 equal cusps with one minor and 2 large cusps with 2 small ones. Our case has 4 equal cusps. Different embryologic mechanisms have been suggested including excavation of one of the valve cushions and septation of a normal valve cushion as a result of inflammatory response (3).

The functional aspect of the quadricuspid aortic valve is mainly represented by pure insufficiency. The physiopathology of the valve dysfunction is poorly understood: anatomical abnormalities of the cusps could induce unequal shear stress leading to fibrosis and incomplete coaptation (4). However, insufficiency is also observed in cases of quadricuspid valve with four equal cusps.

Patients with quadricuspid aortic valve should be followed closely. The risk of endocarditis is probably higher in patient who have unequal cusps. Endocarditis prophylaxis is no longer recommended in the management of patients with quadricuspid aortic valve (5).

When there is an indication for surgical intervention, aortic valve replacement is the most acceptable procedure, although successful surgical repair has been reported.

Conclusion

Quadricuspid aortic valve is an unusual congenital defect, diagnosed mainly in adult life, and many cause aortic valve dysfunction, commonly aortic regurgitation.

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