

## A rare cause of aortic insufficiency: quadricuspid aortic valve

Aort yetersizliğinin nadir bir nedeni: Dört yaprakçıklı aort kapağı

Burcu Demirhan, M.D., Yesim Guray, M.D., Aysel Turkvatan,<sup>1</sup> M.D., Omac Tufekcioglu, M.D.

Türkiye Yüksek İhtisas Training and Research Hospital, Department of Cardiology, <sup>1</sup>Department of Radiology, Ankara

Quadricuspid aortic valve can be documented incidentally as a rare cause of aortic insufficiency. An asymptomatic 24-year-old male patient underwent cardiac evaluation after detection of a cardiac murmur on routine examination. There was a grade 1/6 early diastolic murmur on the right upper side of the sternum. The electrocardiogram was in sinus rhythm and normal. Transthoracic echocardiography showed normal left ventricular dimensions and functions, but it did not provide an adequate visualization of valve structure. Color Doppler examination showed mild aortic insufficiency. Transesophageal echocardiography revealed a quadricuspid aortic valve composed of two equal large cusps and two equal small cusps. Color Doppler scans demonstrated aortic regurgitant flow at the coaptation sites of the small and large cusps. Further studies by aortic computed tomographic (CT), angiography and cardiac CT did not show aortic coarctation or any other coronary abnormality, but the latter confirmed quadricuspid aortic valve.

**Key words:** Aortic valve/abnormalities; aortic valve insufficiency/etiology; echocardiography, transesophageal.

Dört yaprakçıklı aort kapağı, aort yetersizliğinin nadir bir nedeni olarak genellikle tesadüfi bir bulgu şeklinde karşımıza çıkmaktadır. Yirmi dört yaşında asemptomatik bir erkek hastada, rutin muayenede kardiyak üfürüm saptanması üzerine kardiyak değerlendirme yapıldı. Sternumun sağ üst kenarında, 1/6 derecede erken diyastolik üfürüm duyuldu. Elektrokardiyografisi sinüs ritminde ve normaldi. Transtorasik ekokardiyografide sol ventrikül çapları ve sistolik fonksiyonları normal bulunurken, kapak yapısı net değerlendirilemedi. Renkli Doppler ekokardiyografide hafif aort yetersizliği saptandı. Transözofageal ekokardiyografide aort kapağının birbirine eşit büyüklükte iki büyük ve iki küçük yaprakçık içerdiği görüldü. Renkli Doppler çalışmada, küçük ve büyük yaprakçıkların birleştiği her iki bölgeden aort yetersizlik akımları izlendi. Aort bilgisayarlı tomografik (BT) anjiyografi ve kardiyak BT incelemelerinde aort koarktasyonu ya da herhangi bir koroner anomali saptanmazken, dört yaprakçıklı kapak yapısı kardiyak BT ile de doğrulandı.

**Anahtar sözcükler:** Aort kapağı/anormallik; aort kapağı yetersizliği/etyoloji; ekokardiyografi, transözofageal.

Although bicuspid aortic valve is the most common aortic valve-related congenital anomaly, quadricuspid aortic valve, a rare cause of aortic insufficiency, is often incidentally encountered on echocardiographic, surgical or postmortem examination.<sup>[1]</sup> We presented a case diagnosed with quadricuspid aortic valve during investigation of an asymptomatic cardiac murmur.

### CASE REPORT

A 24-year-old asymptomatic male patient visited our clinic for cardiac assessment following the diagnosis of a cardiac murmur during routine physical examination. Physical examination revealed regular heart beats (74 beats/min). The patient who did not have any abnormal finding in his medical and family history had a blood pressure of 110/80 mmHg. His carotid and perip-

heral pulses were found to be normal. A grade 1/6 early diastolic murmur at the right upper sternum which did not radiate to any other focus was heard. Physical examination of the other systems revealed normal functions. The 12-lead electrocardiography was in sinus rhythm and normal. In addition, transthoracic echocardiography showed normal left ventricular dimensions and normal systolic functions, while M-mode and two-dimensional echocardiography demonstrated normal aortic valve opening. However, the valvular structure in the parasternal short-axis view was not accurately assessed. Color Doppler echocardiography revealed double mild aortic insufficiency jet. No gradient was found in the aortic valve; however, suprasternal evaluation showed a gradient of 20 mmHg in the descending aorta. Transesophageal echocardiography was perfor-

Received: 13.08.2008 Accepted: 18.09.2008

Corresponding address: Dr. Burcu Demirhan, Türkiye Yüksek İhtisas Hastanesi, B-Poliklinik Binası, 1. Kat, 06100, Samanpazarı, Ankara  
Tel: +90 - 312 -306 18 28 e-mail: burcume@gmail.com



**Figure 1.** (A) Long axis image on the transesophageal echocardiography. (B) Image of quadricuspid valvular structure on transesophageal echocardiography. (C) Image of mild aortic insufficiency on color Doppler echocardiography.

med on the patient since the aortic valve could not accurately be assessed. The short-axis views revealed an aortic valve composed of two large cusps and two small cusps of equal sizes. Color Doppler scans also demonstrated aortic regurgitant flow at the coaptation sites of the small and large cusps (Figure 1).

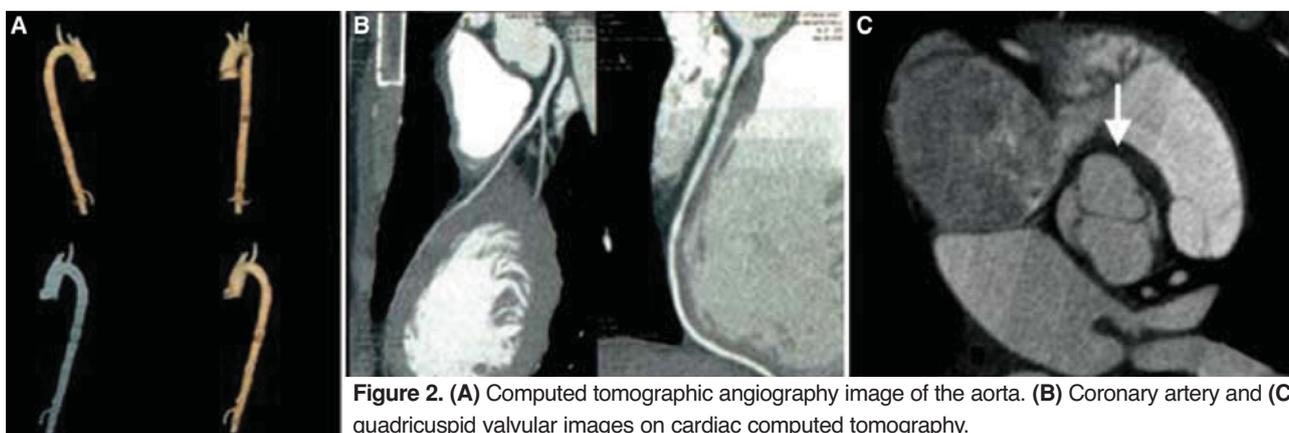
The patient underwent assessment by computed tomographic (CT) angiography for possible aortic coarctation and by cardiac CT for possible coronary abnormality. No aortic coarctation or any other coronary abnormality was revealed; however, cardiac CT confirmed quadricuspid aortic valve (Figure 2).

## DISCUSSION

Semilunar valve malformations are the most common congenital anomalies observed in the heart and the major vessels. Bicuspid aortic valve is the most common among these, with an incidence of %2.<sup>[1]</sup> When compared to bicuspid aortic valve, quadricuspid aortic valve is a very rare condition. The incidence of quadricuspid aortic valve in postmortem examination (0.003%) has been on an increase with the increasing use of echocardiography (0.013%-0.043%).<sup>[2]</sup> Mean age at diagnosis is 50.7, and the incidence has been reported

to be more in men than in women (the man/woman ratio is 1.6/1).<sup>[3]</sup> Quadricuspid aortic valve has been reported to present with other congenital anomalies at a rate of 18.3%. These congenital anomalies include coronary artery abnormalities, ventricular septal defect, patent ductus arteriosus, pulmonary stenosis, ruptured sinus of Valsalva, complete heart block, and hypertrophic cardiomyopathy.<sup>[3-5]</sup> Infective endocarditis involving quadricuspid aortic valve has also been reported.<sup>[6,7]</sup>

Paralleled with embryological division of truncus, three mesenchymal proliferations develop towards the lumen of each major vessel to form semilunar valves in aortic and pulmonary trunk. These proliferations demonstrate the transformation of three semilunar valves united at the center of valve orifice. The formation of a number of various valves may be associated with early stage truncal developmental changes, or with a numerical disorder in the primordial valves, or with abnormal development of mesenchymal proliferation or fusion.<sup>[8]</sup> The incidence of the quadricuspid pulmonary valve is nine times higher than quadricuspid aortic valve and it does not lead any significant pulmonary dysfunction. On the other hand, the aortic valve is generally adversely affected by this valvular structure.<sup>[8]</sup> 75% of the pa-



**Figure 2.** (A) Computed tomographic angiography image of the aorta. (B) Coronary artery and (C) quadricuspid valvular images on cardiac computed tomography.

tients present with aortic insufficiency under this condition.<sup>[3]</sup> Aortic insufficiency is thought to result from deformation or thickening of the valvular structure due to abnormal valve formation or asymmetric mechanical stress.<sup>[9]</sup> On the contrary, aortic stenosis is very rarely encountered.

Hurwitz and Roberts<sup>[10]</sup> described seven anatomical types (type A-G) of quadricuspid aortic valve according to the size of cusps. Type A includes cusps of equal sizes, whereas type G includes cusps of different sizes. Distribution of the classes in between is as follows: Type B includes 3 cusps of equal sizes and 1 small cusp; type C includes 2 large cusps of equal sizes and 2 small cusps of equal sizes; type D includes 1 large, 2 medium-sized, and 1 small-sized cusps; type E: 3 cusps of equal sizes and 1 large cusp; type F: 2 large cusps of equal sizes and 2 different-sized small cusps. Type A, B and C are the most commonly reported types.

Our patient had two large and two small cusps of equal sizes which were described as having a type C quadricuspid valvular structure, presenting with asymptomatic mild aortic insufficiency. Follow-up with annual echocardiography was recommended and the patient was discharged since no other cardiac anomaly was found during our assessment.

In conclusion, the incidence of quadricuspid aortic valve, a rare cause of aortic insufficiency, is reported to be on an increase with the increasing use of echocardiography. On the other hand, transthoracic echocardiography may not accurately assess valvular structure. In this case, the diagnosis may be confirmed by transesophageal echocardiography. In addition, cardiac CT is another technique by which both valvular structure and anomalies of the coronary ostium can be assessed. In particular, this technique is important in identifying lo-

cation of the coronary ostium during surgery, in patients who are scheduled for valve replacement due to symptomatic and severe aortic insufficiency.

## REFERENCES

1. Roberts WC. The congenitally bicuspid aortic valve. A study of 85 autopsy cases. *Am J Cardiol* 1970;26:72-83.
2. Feldman BJ, Khandheria BK, Warnes CA, Seward JB, Taylor CL, Tajik AJ. Incidence, description and functional assessment of isolated quadricuspid aortic valves. *Am J Cardiol* 1990;65:937-8.
3. Tutarel O. The quadricuspid aortic valve: a comprehensive review. *J Heart Valve Dis* 2004;13:534-7.
4. Timperley J, Milner R, Marshall AJ, Gilbert TJ. Quadricuspid aortic valves. *Clin Cardiol* 2002;25:548-52.
5. Bilge AK, Buğra Z, Tayyareci Y, Rüzgar O, Umman S, Meriç M. An uncommon coincidence of congenital quadricuspid aortic valve accompanied by hypertrophic obstructive cardiomyopathy. *Anadolu Kardiyol Derg* 2007;7:E7-8.
6. Takeda N, Ohtaki E, Kasegawa H, Tobaru T, Sumiyoshi T. Infective endocarditis associated with quadricuspid aortic valve. *Jpn Heart J* 2003;44:441-5.
7. Watanabe Y, Taketani Y, Takei Y, Tanaka K, Watanabe Y. Complete heart block resulting from quadricuspid aortic valve penicillin-resistant pneumococcal endocarditis: a case report. *Circ J* 2003;67:275-6.
8. Davia JE, Fenoglio JJ, DeCastro CM, McAllister HA Jr, Cheitlin MD. Quadricuspid semilunar valves. *Chest* 1977;72:186-9.
9. Jacobs JE, Srichai M, Kim D, Hecht E, Kronzon I. Quadricuspid aortic valve: Imaging findings on multi-detector helical CT with echocardiographic correlation. *J Comput Assist Tomogr* 2006;30:569-71.
10. Hurwitz LE, Roberts WC. Quadricuspid semilunar valve. *Am J Cardiol* 1973;31:623-6.