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Short communication

Rare congenital valve disease: Quadricuspid aortic valve

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Introduction

Quadricuspid aortic valve (QAV) is rare congenital aortic valve disease mostly occurring with aortic regurgitation. In most cases QAV may be found incidentally although it causes valve dysfunction, and associated with other cardiac abnormalities. The predominant clinical findings and management issues in QAV relate to aortic regurgitation. We report a case of QAV detected by echocardiography in patient with mild aortic regurgitation.

Case presentation

This case report represents a rare clinical finding of an isolated QAV in an otherwise healthy 68-year-old female who was referred for cardiac evaluation due to heart murmur and exertional dyspnea without chest pain. Patient had history of well-controlled arterial hypertension. On physical examination, blood pressure was 134/76 mm Hg and 2/6 early diastolic murmur was heard at the left parasternal border. The electrocardiogram revealed sinus rhythm without any abnormality. Transthoracic echocardiography (TTE) showed normal LV ejection fraction (62%), mild aortic regurgitation and an atypical aortic valve without the usual Y-aspect trileaflet closure. The aortic root (diameter: 34 mm), ascending aorta, mitral, tricuspid, and pulmonary valves were normal. There were no other cardiac abnormalities. Color Doppler examination showed mild aortic insufficiency resulting from incomplete diastolic coaptation of the cusps (Fig. 1). Transesophageal echocardiography (TEE) was performed for more accurate evaluation of aortic valve because TTE does not provide an adequate visualization. The QAV with 4 equal-sized leaflets with an "x shaped" commissural pattern in diastole and mild functional regurgitation was confirmed (Fig. 2a–b). According to the classification of Hurwitz and Roberts, our patient had the type A variant. The patient did not accept the performance of other diagnostic tests.

Discussion

QAV is a rarer congenital valve malformation than bicuspid or tricuspid valve and an estimated prevalence range is 0.013% to 0.043% with equal sex distribution [1,2]. In the past, most cases were diagnosed incidentally during surgery or autopsy [3] but now TTE or TEE plays an important role to define this malformation [4].

In 1973, Hurwitz and Roberts have classified QAV according to the size of the leaflets which described seven anatomic variants (type A–type G). Type A (4 equal cusps), type B (3 large equal cusps, 1 smaller cusp), type C (2 larger equal cusps, 2 smaller equal cusps), type D (1 large cusp, 2 intermediate cusps, 1 small cusp), E (3 equal cusps, 1 large cusp), type F (2 large equal cusps, 2 smaller unequal cusps), or type G (4 unequal cusps). The most frequent leaflet morphology types were A and B [5]. The type described in this case, with four equal cusps is type A.

Although aortic regurgitation is the most common abnormality associated with QAV, valvular stenosis is very rare (0.7%) [6]. Asymmetric distribution of stress around the four cusps and abnormal leaflet coaptation lead to aortic insufficiency [6]. Combined QAV regurgitation and stenosis have been reported in 8.4% of all documented cases [6]. QAV dysfunction is minimal or absent at an early age. The mean date of recognition is about in the 5th or 6th decade of life QAV [6].

QAV can be associated with other cardiac defects such as ventricular or atrial septal defects, patent ductus arteriosus, subaortic fibromuscular stenosis, a malformation of the mitral valve, and coronary anomalies [6–9]. Also it is related with risk of infective endocarditis because of the progressive deterioration of the leaflets [3].

Echocardiography is useful in the evaluation of valvular structure and functional status.

TTE has limitations in providing adequate visualization because of patient's morphology and may be suboptimal in recognizing this

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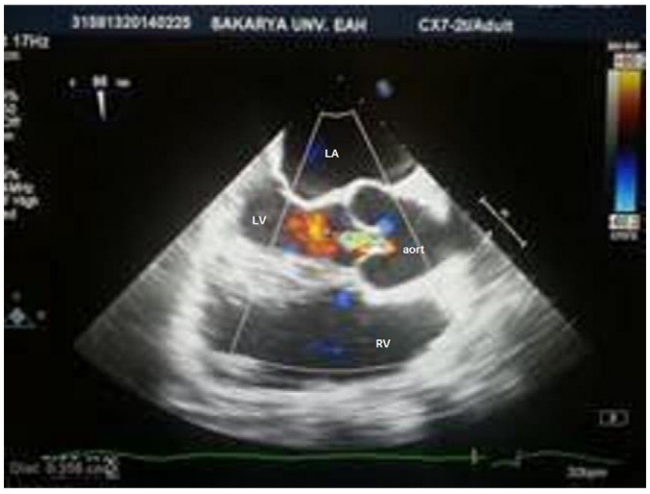

Fig. 1. Transesophageal long-axis view, color Doppler echocardiography showing mild aortic regurgitation.

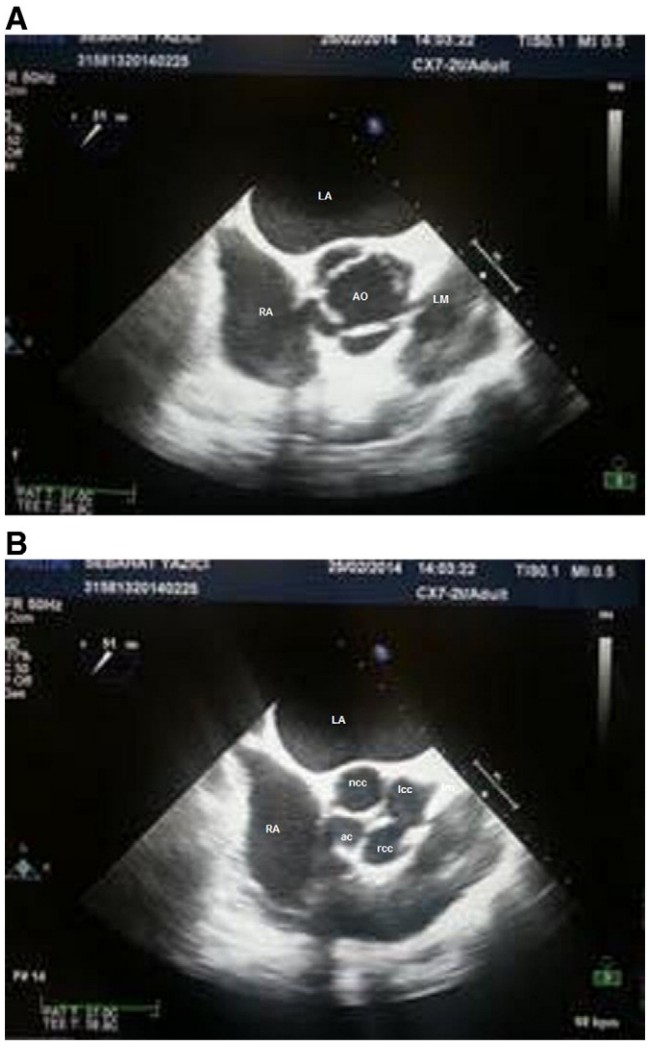

Fig. 2. A–B: Two-dimensional transesophageal short-axis view of the aortic valve; showing the quadricuspid aortic valve, which consisted of four equal cusps in both systolic (2a) and diastolic (2b) frames (type A). The "x shaped" commissural pattern in diastole was confirmed (2b). The left main coronary artery (LM in B) was detected in the normal position. ac — accessory cusp; lcc — left coronary cusp; ncc — non-coronary cusp; rcc — right coronary cusp; AO — aorta; LA — left atrium; LV — left ventricle; RA — right atrium; RV — right ventricle; LM — left main.

malformation. At these points, TEE, three-dimensional echocardiography (3-DE), aortic computed tomographic (CT), and cardiac magnetic resonance imaging (MRI) are used for detection of the aortic valve anatomy and the other cardiac defects associated with QAV.

The clinical symptoms and management of treatment in QAV relate to aortic regurgitation. Aortic valve replacement for QAV is usually the treatment of choice for severe valvular regurgitation. Surgery was applied about half of the patients with QAV (45.2%) in adulthood mainly due to progressive aortic regurgitation [10].

Conclusion

QAV is a rare innate disease, diagnosed mainly in asymptomatic adult patient incidentally. QAV may cause aortic valve dysfunction, mostly aortic regurgitation. Occasionally, it is associated with other congenital malformations. For these reasons the recognition of QAV is important. Although TTE is the method of choice in the diagnosis of QAV, TEE should be performed to investigate whether other congenital anomalies are associated with QAV. We described the case of a patient who had mild aortic regurgitation resulting from a QAV.

Conflict of interests

The authors declare that they have no conflict of interests.

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