INDIAN HEART JOURNAL 67 (2015) 459–461

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/ihj

Case Report

Isolated quadricuspid aortic valve referred with diagnosis of rheumatic carditis

Mustafa Kosecik [a,*], *Bahri Elmas* [b]

[a] *Division of Pediatric Cardiology, Sakarya University School of Medicine, Sakarya, Turkey*
[b] *Department of Pediatrics, Sakarya University School of Medicine, Sakarya, Turkey*

ARTICLE INFO

Article history:
Received 5 November 2014
Accepted 4 June 2015
Available online 5 August 2015

Keywords:
Quadricuspid aortic valve
Regurgitation
Echocardiography

ABSTRACT

Quadricuspid aortic valve is a rare congenital malformation of the aortic valve. It appears often isolated or sometimes associated with other truncal anomalies, and approximately 50% of patients have aortic regurgitation. In this article we reported an eight-years-old boy referred with diagnose of rheumatic carditis from the outer medical center.

1. Introduction

Quadricuspid aortic valve (QAV) is a rare congenital cardiovascular abnormality that often causes aortic regurgitation in adulthood, with also a concurrent stenosis in some patients.[1–3] The frequency is estimated in between 0.008% and 0.03% of population.[2] Most cases have been discovered incidentally at autopsy or at aortic valve replacement with fewer cases detected by echocardiography. Recent technological advances in transthorasic and transesophageal echocardiography have made it possible to detect this anomaly easily and non-invasively.[4] To the best of our knowledge, most of cases diagnosed on transthorasic echocardiography (TTE) examination are in adulthood, but there are fewer cases reported in childhood.[5]

In this article we reported an eight-years-old boy referred with diagnose of rheumatic carditis from the outer medical center.

2. Case report

An eight-years-old boy admitted to outer medical center with history of chest pain and mild fever for seven days and determined mildly aortic valve regurgitation on TTE examination was referred to our hospital with diagnosis of rheumatic carditis. No past history of rheumatic disease and endocarditis could be found. On his physical examination, body temperature was 37.8 °C, pulses were symmetrical and of normal contour, blood pressure was 100/60 mmHg, and heart rate 86 beats/min. No systolic or diastolic murmur in aortic

* *Corresponding author*. Sakarya University Research Hospital, Pediatric Cardiology Unite, Sakarya, TR-54000, Turkey. Tel.: +90 264 444 54 00; fax: +90 264 275 91 92.
E-mail address: mustafakosecik@yahoo.com (M. Kosecik).

http://dx.doi.org/10.1016/j.ihj.2015.06.009

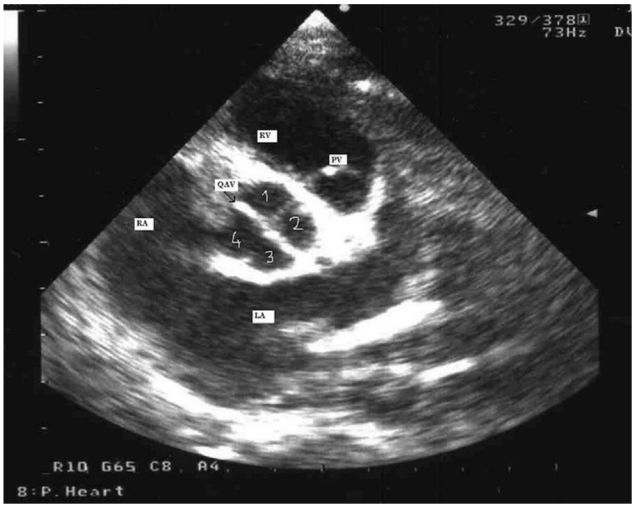

Fig. 1 – Parasternal short-axis view of the quadricuspid aortic valve in diastole showing four equal-sized cusps with the "X" configuration. LA = left atrium, RA = right atrium, RV = right ventricule, PV = pulmonary valve, QAV = quadricuspid aortic valve.

and/or mitral valve area was present. On admission, renal and liver function tests, hematologic findings, C-reactive protein, erythrocyte sedimentation rate and anti-streptolysin O titer were within normal limits, and throat culture was negative. The ECG and chest X-ray were normal. Two-dimensional TTE in parasternal short-axis of great arteries showed a QAV with a cross shape in diastole ("**X**" configuration). No thickness and calcification were found. The four cusps were of normal texture and of approximately equal size (Fig. 1). Mild aortic valve regurgitation was detected by color and continuous Doppler echocardiography. Left ventricular dimension and systolic function were normal. The patient's history, clinic and laboratory findings were insufficient for diagnosis of rheumatic fever. The patient recognized isolated QAV with mild aortic valve regurgitation was periodically invited to echocardiography examination.

3. Discussion

The first case with QAV was reported in 1862.[6] Simonds et al reported an incidence of 0.008% by autopsy.[7] With improvement in ultrasonographic technology during the past several years, valvular anatomy can be assessed confidently in vivo. Herman et al described the first case of isolated QAV diagnosed by cross sectional echocardiography.[4] A retrospective analysis of 60446 echocardiograms revealed 8 QAVs (0.013%).[2] Among 225 patients who underwent aortic valve surgery for pure aortic regurgitation, 2 were found to have QAV (an incidence of 0.88%).[8] The present patient with isolated QAV will be second case reported in childhood in the literature.

Two-dimensional TTE has become the diagnostic test of choice, because the four cusps and their relative sizes can be easily assessed,[4] as shown in the present case. However, because of technical factors such as suboptimal echocardiographic windows or extensive aortic calcification, TTE may occasionally be limited in visualizing the aortic valve. Transesophageal echocardiography overcomes many of the imaging limitations of TTE and can clearly delineate the aortic valve morphology.[4,5]

The malformation has to be differentiated from "pseudo-quadricuspid" valve caused by endocarditis, rheumatic valve disease, or surgery. "True" QAV should bear a corpus arantii on each cusp. The occasional small accessory cusp in QAV may be overlooked even in necropsy or at surgery. On echocardiography QAV has usually been defined as a characteristic "**X**" shape in diastole and freely opening cusps like a rectangular configuration in systole.[2] Our case was showed a QAV with a characteristic "**X**" shape in diastole on echocardiography and classified as type A (four relatively equal cusps) in accordance with the Hurwitz and Roberts classification,[9] which described seven anatomic variants (type A – type G), as shown in Fig. 1.

QAV is often incompetent[4]; normally functioning valves have been found in only 16% of cases.[10] The present case with QAV has mild aortic regurgitation that was central related to incomplete coaptation of four cusps. Surgical and histologic findings have shown that fibrous thickening of valves, due to uneven distribution of mechanical stress, results in incomplete coaptation of the cusps, in aortic regurgitation and more vulnerability to infection resulting in endocarditis.[2,11] Doppler echocardiography is a sensitive technique that allows its detection even when it is not clinically significant. For these reasons the patients with QAV need periodical non-invasive evaluation.[11]

4. Conclusions

QAV is a rare congenital malformation that can be easily diagnosed by two-dimensional TTE because of the typical aspect of the four cusps in both diastole and systole. Mild aortic regurgitation is frequently associated with QAV. Consequently, presence of QAV should be investigated in all children determined aortic valve regurgitation.

Conflicts of interest

The authors have none to declare.

REFERENCES

1. Yener N, Resatoglu A, Elnur el AE, Yener A. Quadricuspid aortic valve: a chance for repair. *J Heart Valve Dis.* 2004;13:860–862.
2. Feldman BJ, Khandheria BK, Warnes CA, et al. Incidence, description and functional assessment of isolated quadricuspid aortic valves. *Am J Cardiol.* 1990;65:937–938.
3. Yildirim SV, Gümüş A, Coşkun I, Türköz R. Quadricuspid aortic valve: a rare cause of aortic regurgitation and stenosis. *Turk J Pediatr.* 2008;50:500–502.
4. Herman LR, Cohen IS, Glaser K, Newcomb 3rd EW. Diagnosis of incompetent quadricuspid aortic valve by two-dimensional echocardiography. *Am J Cardiol.* 1984;53:972.

5. Gouveia S, Martins JD, Costa G, et al. Quadricuspid aortic valve–10-year case series and literature review. *Rev Port Cardiol.* 2011;30:849–854.
6. Balington J. *London Medical Gazette*. July 1862.
7. Simonds JP. Congenital malformations of the aortic and pulmonary valves. *Am Med Sci.* 1923;166:584–595.
8. Olson LJ, Subramanian R, Edwards WD. Surgical pathology of pure aortic insufficiency: a study of 225 cases. *Mayo Clin Proc.* 1984;59:835–841.
9. Hurwitz LE, Roberts WC. Quadricuspid semilunar valve. *Am J Cardiol.* 1973;31:623–626.
10. Tutarel O. The quadricuspid aortic valve: a comprehensive review. *J Heart Valve Dis.* 2004;13:534–537.
11. Recupero A, Pugliatti P, Rizzo F, Arrigo F, Coglitore S. Quadricuspid aortic valve: a rare cause of aortic insufficiency diagnosed by Doppler echocardiography. Report of two cases and review of the literature. *Ital Heart J.* 2005;6:927–930.