Cor et Vasa

Available online at www.sciencedirect.com

ScienceDirect

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

Kasuistika | Case report

Quadricuspid aortic valve: A case report

Shi-Min Yuan[a], **Song-Li Yan**[b]

[a] *Department of Cardiothoracic Surgery, The First Hospital of Putian, Teaching Hospital, Fujian Medical University, Putian, Fujian Province, China*

[b] *Department of Ultrasonography, The First Hospital of Putian, Teaching Hospital, Fujian Medical University, Putian, Fujian Province, China*

ARTICLE INFO

Article history:
Received: 20. 1. 2016
Accepted: 5. 3. 2016
Available online: 31. 3. 2016

Klíčová slova:
Aortální insuficience
Echokardiografie
Kontrolní vyšetření

Keywords:
Aortic valve insufficiency
Echocardiography
Follow-up studies

SOUHRN

Čtyřcípá aortální chlopeň se vyskytuje vzácně. Sedmačtyřicetiletá žena si stěžovala na mírnou dušnost při zátěži a byla odeslána do naší nemocnice. Transthorakální echokardiografie prokázala čtyřcípou aortální chlopeň s mírnou aortální insuficiencí. Při kontrolním vyšetření po devíti měsících se ženě vedlo dobře, bez progrese výše popsaného mírného symptomu. V tomto článku je popsán velmi vzácný případ čtyřcípé aortální chlopně typu D. Transthorakální echokardiografie může poskytnout přesné anatomické informace o čtyřcípé aortální chlopni. Při mírných symptomech je nutno pacienty pravidelně sledovat.

ABSTRACT

Quadricuspid aortic valve is rare. A 47-year-old female complained of mild exertional dyspnea and was referred to this hospital. Transthoracic echocardiography showed a quadricuspid aortic valve with mild aortic regurgitation. She was doing well without any progression of her mild symptom at 9-month follow-up. In this report, a very rare type D quadricuspid aortic valve is described. Transthoracic echocardiography may provide with exact anatomies of the quadricuspid aortic valve. Patients with mild symptoms may warrant a close follow-up.

Introduction

Quadricuspid aortic valve (QAV) is a rare congenital variant of the aortic valve and is often incidentally found during surgery or autopsy. Due to the extensive use of echocardiography, QAV is now increasingly disclosed preoperation or antemortem [1].

Case report

A 47-year-old female complaint of mild exertional dyspnea and referred to us for a check-up on September 9, 2014. Transthoracic echocardiography revealed normal dimensions of heart chambers. Meanwhile, a QAV was conformed with one large, two intermediate and one small

Address: Song-Li Yan, MD, Department of Ultrasonography, The First Hospital of Putian, Teaching Hospital, Fujian Medical University, 389 Longdejing Street, Chengxiang District, Putian 351100, Fujian Province, China, e-mail: shiminyuan@126.com
DOI: 10.1016/j.crvasa.2016.03.001

Please cite this article as: S.-M. Yuan, S.-L. Yan, Quadricuspid aortic valve: A case report, Cor et Vasa 58 (2016) e513–e514 as published in the online version of *Cor et Vasa* available at http://www.sciencedirect.com/science/article/pii/S0010865016300169

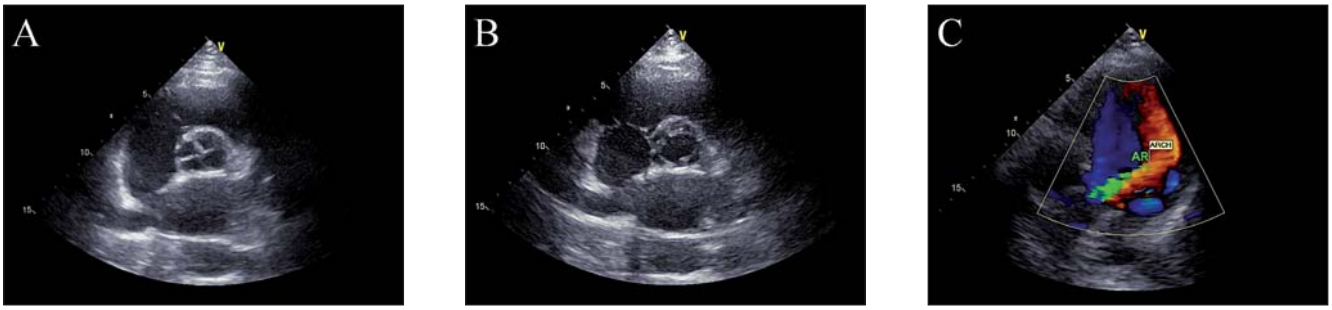

Fig. 1 – Echocardiographic visualization of the quadricuspid aortic valve showing one large, two intermediate and one small cusps: (A) on the diastolic and (B) systolic phases, and (C) mild eccentric jet of aortic regurgitation. AR – aortic regurgitation.

cusps on the diastolic and systolic phases (Figs. 1A and 1B). There was mild eccentric jet of aortic regurgitation (Fig. 1C) but no significant aortic valve gradient. The patient was on a close follow-up. She was doing well without any progression of her mild symptom 9 months later.

Discussion

The incidence of QAV was 0.008% in autopsy series and 0.043% in patient population by TTE [2]. The embryology of QAV remains unknown. However, it has been proposed that any developing problems leading to the division of one of the three mesenchymal ridges would be the underlying mechanism [1].

Hurwitz and Roberts [3] classified QAVs into 7 types according to the sizes of the valve leaflets: A (four equal cusps), B (three equal cusps, one small cusp), C (two equal larger and two equal small cusps), D (one large, two intermediate and one small cusps), E (three equal cusps, one larger cusp), F (two equal larger and two unequal smaller cusps), and G (four unequal cusps), 85% of which were types A, B and C. The present patient showed a type D QAV.

On the short axis view of the aortic valve in diastole, the commissural lines formed by the adjacent cusps shows an "X" configuration other than the "Y" configuration of the normal tricuspid aortic valve [1]. Coronary ostium displacement might also be shown on transesophageal echocardiography [4]. Apart from TTE and TEE, cardiac magnetic resonance imaging and multidetector computed tomography are also helpful in the diagnosis of QAV in the current era [2].

Aortic insufficiency, which is considered the result of fibrous thickening with incomplete coaptation of the valve leaflets, is the most predominant complication of QAV accounting for 75% of the cases [1]. Besides, 9% show both aortic stenosis and regurgitation and 16% have normal-functioning QAVs [2]. Unlike in bicuspid aortic valve, aortic root dilation is only occasionally seen in QAV patients [5]. QAVs with four equal leaflets are not more likely to function normally than do those with unequal leaflet sizes [1]. QAV may be affected by infective endocarditis [6–8]. Often does the valve function of the QAVs deteriorate during adulthood and a surgical treatment is warranted during the sixth decade of life [1]. Aortic valve replacement is indicated for the patients with severely regurgitant or stenosed QAVs [2, 5].

QAV is rare. Transthoracic echocardiography may provide with exact anatomies of the QAV. Patients with mild symptoms may warrant a close follow-up.

Conflict of interest
No conflict of interest.

Funding body
None.

Ethical statement
I declare, on behalf of co-author, that the research was conducted according to Declaration of Helsinki.

Informed consent
I declare, on behalf of co-author, that informed consent was obtained from the patient participating in this study.

References

[1] J. Timperley, R. Milner, A.J. Marshall, T.J. Gilbert, Quadricuspid aortic valves, Clinical Cardiology 25 (2002) 548–552.
[2] C.B. Chapman, T. Kohmoto, A.F. Kelly, et al., Cardiac computed tomography and quadricuspid aortic valve: a case report, Wisconsin Medical Journal 109 (2010) 219–221.
[3] L.E. Hurwitz, W.C. Roberts, Quadricuspid semilunar valve, American Journal of Cardiology 31 (1973) 623–626.
[4] T. Watanabe, Y. Hosoda, S. Sasaguri, Y. Aikawa, A quadricuspid aortic valve diagnosed by transesophageal echocardiography: report of a case, Surgery Today 28 (1998) 1102–1104.
[5] P. Garg, H. Kamaruddin, R. Orme, V. Watt, Type F congenital quadricuspid aortic valve: a very rare case diagnosed by 3-dimenional transoesophageal echocardiography, Open Cardiovascular Medicine Journal 8 (2014) 23–25.
[6] I. McColl, Pericarditis due to a mycotic aneurysm in subacute bacterial endocarditis: report of a case affecting congenitally stenosed quadricuspid aortic valve, Guy's Hospital Reports 107 (1958) 34–47.
[7] T. Matsukawa, S. Yoshii, R. Hashimoto, et al., Quadricuspid aortic valve perforation resulting from bacterial endocarditis – 2-D echo- and angiographic diagnosis and its surgical treatment, Japanese Circulation Journal 52 (1988) 437–440.
[8] M.T. Dotti, N. De Stefano, S. Mondillo, et al., Neurological involvement and quadricuspid aortic valve in a patient with Ehlers–Danlos syndrome, Journal of Neurology 246 (1999) 612–613.