

Quadricuspid Aortic Valve Diagnosed by Transesophageal Echocardiography: A Case Report

Farahnaz Nikdoust, MD¹, Hakimeh Sadeghian, MD¹, Bahareh Eslami, MD¹,
Shahla Majidi, MD^{1*}, Dariush Javidi, MD²

¹Tehran Heart Center, Tehran University of Medical Sciences, Tehran, Iran.

²Shariati Hospital, Tehran University of Medical Sciences, Tehran, Iran

Received 14 December 2008; Accepted 19 April 2009

Abstract

Quadricuspid aortic valve (QAV) is an uncommon congenital anomaly which was an incidental finding during surgery or autopsy in the past. We present the case of a 44-year-old woman with moderately severe aortic regurgitation due to unequal cusp size QAV diagnosed via transesophageal echocardiography. Due to echocardiographic imaging improvement, the diagnosis of QAV is now easier and earlier than the past.

J Teh Univ Heart Ctr 2 (2010) 95-97

Keywords: Echocardiography, transesophageal • Aortic valve • Aortic valve insufficiency

Introduction

Quadricuspid aortic valve (QAV), a rare congenital heart defect with an incidence range of 0.008 % to 0.033%,^{1,2} was first reported by Balington et al.³ In former years, QAV was an accidental finding during surgery or necropsy, but recent improvements in echocardiographic imaging techniques have conferred an easier and earlier detection of QAV. Herein, we report a case of QAV diagnosed via transesophageal echocardiography (TEE).

Case report

A 44-year-old woman presented with a history of dyspnea on exertion (New York Heart Association functional class II) starting 7 years previously and aggravated in the previous 2-3 years. Her past medical history revealed hypertension and hypothyroidism, without a positive family history of heart diseases. She had been under observation for the past 3 years and undergone two transthoracic echocardiography

(TTE) examinations, in which only moderate to severe aortic insufficiency was detected. Because of the deterioration of her dyspnea, she was referred to Tehran Heart Center for TEE evaluations.

On physical examination, she had a grade II/VI decrescendo diastolic murmur in the second intercostal space of the right sternal border with no radiation and a wide pulse pressure. Blood pressure was 170/70 mmHg with a normal pulse rate, and lung auscultation was normal. Chest radiography showed a CT ratio at the upper limit of normal; and normal sinus rhythm, normal axis deviation, and left ventricular (LV) hypertrophy were detected on the electrocardiogram. Angiography reported an ejection fraction (EF) of 50%, severe aortic insufficiency, mild aortic root dilatation, LV pressure of 150/0-15, aortic pressure of 150/80, and normal coronary artery.

In the treadmill exercise test, she developed fatigue in stage III of the Bruce protocol (after 6 minutes).

In TTE, there was mild LV and left atrial dilation with normal LV systolic function (left ventricular end-diastolic diameter = 57 mm, left ventricular end-systolic diameter = 41 mm, left

*Corresponding Author: **Shahla Majidi**, Department of Cardiology, Tehran Heart Center, North Kargar Street, Tehran, Iran. 1411713138. Tel & Fax: +98 21 88029256. Email: shahla.majidi@yahoo.com.

ventricular EF = 55%), moderately severe aortic insufficiency with no aortic stenosis, moderate mitral regurgitation, normal right ventricle size and function, mild tricuspid regurgitation, and pulmonary artery pressure = 36 mmHg.

TEE was performed for further evaluations of the aortic and mitral valves with regard to the severity and mechanism of the insufficiency of the aortic and mitral valves. After precise evaluation, TEE confirmed moderate to severe aortic insufficiency (Figure 1) and moderate mitral regurgitation and also revealed QAV (Figure 2) and the prolapse of both mitral leaflets as the mechanisms of the valvular regurgitation, respectively. Apart from a small patent foramen ovale, no dilation of the sinus of Valsalva or ascending aorta was visualized.

Discussion

A rare congenital heart defect, QAV has an incidence range of 0.008 % to 0.033%.^{1,2} It was first reported by Balington et al. in 1826.³ QAV used to be an accidental finding during aortography,⁴ surgery, or autopsy; currently however, TTE and TEE are the methods of choice in the detection of QAV. It is deserving of note that similar to our case, TTE is reported to only detect aortic regurgitation and not QAV in some patients.⁵

According to the leaflet morphology, 7 types of QAV are described by Hurwitz and Roberts, named from A to G.² Because of 1 large cusp, 2 intermediate-sized cusps, and 1 small cusp, our patient was set in type D. In this case, the placement of the left coronary artery was normal.

Holm et al. reported a case with a large left main coronary artery originating unusually low in the aortic root near the posterior margin of the left cusp, which produced a “4-leaf clover” appearance.⁶ And Kaminishi et al. reported a widely patent left coronary ostium in their case.⁷

QAV is a single malformation which is rarely associated with other congenital defects, including paroxysmal supraventricular tachycardia, right double kidney with double renal pelvis and double proximal ureter, hypertrophic obstructive and non-obstructive cardiomyopathy, patent duct, atrial septal defect, ventricular septal defect, pulmonary valve stenosis, bicuspid pulmonary valve, and malformation of the mitral valve.⁸⁻¹²

Associated mitral valve anomalies which are reported include severe mitral regurgitation, prolapsed mitral valve causing a mild regurgitation, mitral regurgitation resulting from annulus dilatation and thickening of the anterior leaflet, severe mitral regurgitation due to infective endocarditis, ruptured mitral valve aneurysm, and hypoplastic anterior mitral leaflet.¹²⁻¹⁶

In the case presented herein, there was a concomitant patent foramen ovale and eccentric moderate mitral regurgitation due to prolapsed mitral valve leaflets. Aortic stenosis is rare,

and most dominant abnormalities due to QAV are different degrees of aortic insufficiency because of malcoaptation of the leaflets.^{11,17} Similarly, aortic valve leaflets could not meet each other centrally in our patient.

Conclusion

QAV is an uncommon congenital anomaly which was accidentally found in surgery or autopsy in former years. Aortic insufficiency is the most prevalent abnormalities associated with QAV. Nowadays, in tandem with advances in echocardiographic imaging, TEE helps us to identify QAV earlier and to manage it more appropriately.

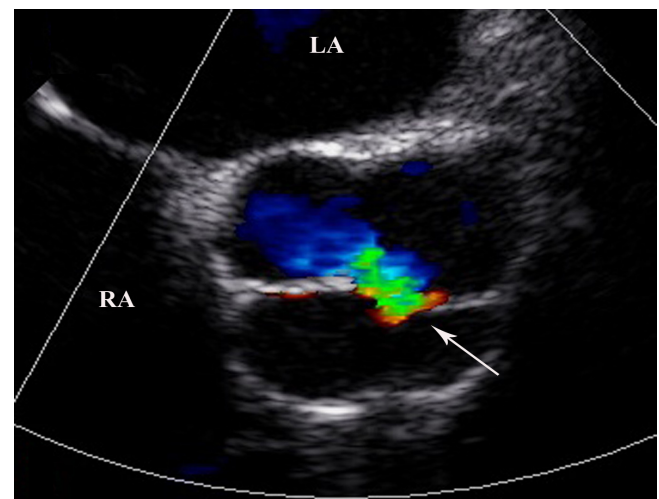


Figure 1. Moderately severe aortic regurgitation by transesophageal echocardiography (arrow) seen in the short-axis view of the aortic valve
LA, Left atrium; RA, Right atrium

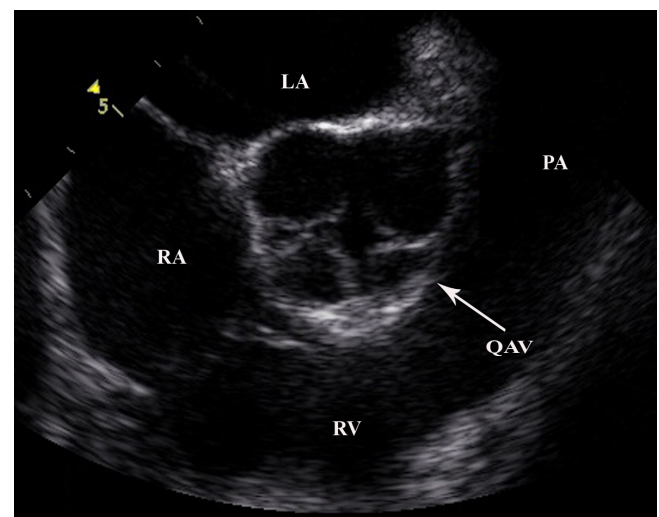


Figure 2. Quadricuspid aortic valve (arrow) by transesophageal echocardiography seen in the short-axis view of the aortic valve
LA, Left atrium; PA, Pulmonary artery; QAV, Quadricuspid aortic valve; RA, Right atrium; RV, Right ventricle



References

1. Simonds JP. Congenital malformation of aortic and pulmonary valves. *Am J Med Sci* 1923;166:584-595.
2. Hurwitz LE, Robert WC. Quadricuspid semilunar valves. *Am J Cardiol* 1973;31:623-626.
3. Balington, quoted by Robicsek F, Sanger PW, Daugherty HK. Congenital quadricuspid aortic valve with displacement of the left coronary orifice. *Am J Cardio* 1969;23:288-290.
4. Chang PC, Lin CC, Lee CS, Wang PH, Chen YF. Rare anatomic entity of quadricuspid aortic valve with aortic insufficiency: a case report. *Kaoshiung J Med Sci* 2007;23:422-425.
5. Formica F, Sangalli F, Ferro O, Paolini G. A rare cause of severe aortic regurgitation: quadricuspid aortic valve. *Interact Cardiovasc thorac surg* 2004;3:672-674.
6. Holm H, Jacobson S, Reul GJ, Stainback RF. Quadricuspid aortic valve. *Tex Heart Ins J* 2004;31:450-451.
7. Kaminishi Y, Terada Y, Nakamura K, Gomi S, Sato F, Mihara W, Enomoto Y, Sakakibara Y, Jikuya T, Atsumi N, Shigeta O, Mitsui T. Aortic valve regurgitation due to quadricuspid valve. A report of complicated case. *Kyobu Geka* 1997;50:63-66.
8. Vohra RK, Singh H, Failing CF. A quadricuspid aortic valve with atrial septal defect. *Echocardiography* 2006;23:865-868.
9. Nucifora G, Badano LP, Iacono MA, Fazio G, Cinello M, Marinigh R, Fioretti PM. Congenital quadricuspid aortic valve associated with obstructive hypertrophic cardiomyopathy. *J Cardiovasc Med (Hagerstown)* 2008;9:317-318.
10. Turhan S, Tulunay C, Sayin T, Ozdol C, Kilickap M, Dincer I. Two cases of quadricuspid aortic valve. *Eur J Echocardiography* 2006;7:330-331.
11. Janssens U, Klues HG, Hanrath P. Congenital quadricuspid aortic valve anomaly associated with hypertrophic non-obstructive cardiomyopathy: a case report and review of the literature. *Heart* 1997;78:83-87.
12. Podesta A, Dottori V, Parodi E, Crivellari R, Regesta T, Passerone GC. Quadricuspid aortic valve, paroxysmal supraventricular tachycardia and double right kidney: an uncommon association. *Minerva Cardioangiol* 2001;49:75-79.
13. Di Pino A, Gitto P, Silvia A, Bianca I. Congenital quadricuspid aortic valve in children. *Cardiol Young* 2008;14:1-4.
14. Irisawa T, Yoshiya K, Yokosawa T, Iwamatsu T, Arai K, Aoki T. A case of quadricuspid valve associated with mitral regurgitation. *Kyobu Geka* 1993;46:618-621.
15. Asami H, Asano H, Handa N, Nakamura S, Ogiwara M, Ueda K, Kyo S, Yokote Y, Omoto R. A surgical case of quadricuspid aortic valve associated aortic regurgitation and severe mitral regurgitation due to infective endocarditis. *Kyobu Geka* 1998;51:216-219.
16. Goh K, Yamamoto H, Inaba M, Kakuchi H, Sasajima T. Ruptured mitral valve aneurysm in patient with quadricuspid aortic valve. *J Cardiovasc Surg (Torino)* 2000;41:393-394.
17. Feldman BJ, Khanderia BK, Wames CA, Seward JB, Taylor CL, Tajik AJ. Incidence, description and functional assessment of isolated quadricuspid aortic valves. *Am J Cardiol* 1990;65:937-938.