OLGU SUNUMU • CASE REPORT

Kosuyolu Kalp Derg 2013;16(1):75-77 • doi: 10.5578/kkd.3937

Quadricuspid Aortic Valve

Quadricuspid Aort Kapak

Mehmet Taşar[1], Osman Tansel Darçın[1], Okay Güven Karaca[1], Mehmet Kalender[1], Ayşe Gül Kunt[1]

[1] Department of Cardiovascular Surgery, Konya Training and Research Hospital, Konya, Turkey
[1] *Konya Eğitim ve Araştırma Hastanesi, Kalp ve Damar Cerrahisi Kliniği, Konya, Türkiye*

ABSTRACT

Quadricuspid aortic valve is an extremely rare congenital defect. Isolated form appears frequently. It is seen with aortic valve regurgitation in adulthood. This case report presents a rare clinical finding of an isolated quadricuspid aortic valve with aortic regurgitation in an 64-year-old female who was referred for cardiac evaluation due to newly identified murmur.

Key Words: *Aortic valve; aortic valve insufficiency.*

Received: *20.06.2012* • ***Accepted:*** *05.07.2012*

ÖZET

Quadricuspid aort kapak oldukça nadir görülen konjenital bir defekttir. İzole formuna daha sık rastlanmaktadır. Genellikle erişkin dönemde aort kapak yetmezliği şeklinde karşımıza çıkar. Biz burada erişkin dönemde aort yetmezliği kliniği ile karşımıza çıkan izole quadricuspid aort kapak saptanan hastamıza yapılan biyolojik kapak replasmanı olgusunu sunuyoruz.

Anahtar Kelimeler: *Aort kapak; aort kapak yetmezliği.*

Geliş Tarihi: *20.06.2012* • ***Kabul Tarihi:*** *05.07.2012*

INTRODUCTION

Isolated quadricuspid aortic valve is an extremely rare congenital cardiac defect which has 0.008% prevalence reported in autopsy series[(1)]. In literature, 200 cases were reported; the largest serial includes 186 cases[(2)]. First case reported by Balington in 1862[(3)]. Quadricuspid aortic valve anomaly is seen generally isolated but in rare cases it may accompany coronary artery anomaly. Patients become symptomatic in adulthood but there are cases reported symptoms started in childhood[(4,5)]. Natural history progresses to aortic regurgitation. We present a patient with aortic regurgitation resulting from quadricuspid aortic valve replaced with a biological prosthetic valve in this report.

CASE REPORT

Sixty four years old female patient was admitted to our hospital with malaise and presyncope attack. She also described shortness of breath for a long while. Physi-

Yazışma Adresi/ Correspondence

Dr. Mehmet Taşar

Konya Eğitim ve Araştırma Hastanesi, Kalp ve Damar Cerrahisi Kliniği Konya-Türkiye

e-posta

mehmet.tasar@hotmail.com

cal examination revealed systolic aortic ejection murmur. Transthoracic echocardiography mentioned moderate-to-severe aortic regurgitation. Ejection fraction measured as 42%, EDLVD: 6.2 cm, ESLVD: 5.1 cm. Coronary artery angiography showed normal coronary morphology. We detected aortic valve with four commisures and four leaflets which had heavy calcifications intraoperatively (Figure 1). The aortic valve was resected and replaced with 23 sized biological prostheses. Patient discharged on postoperatif 7th day. She had control in postoperative 1st month. Transthoracic echocardiography revealed normal functioning aortic prosthetic valve. Now she is fine.

DISCUSSION

Quadricuspid aortic valve is a rare congenital heart defect that is usually found incidentally at the time of open heart surgery or at autopsy[6]. Transesophageal approach reveal aortic valvular morphology and leaflet functioning status better but we were able to detect moderate-to-severe regurgitant quadricuspid aortic valve with transthoracic echocardiography.

An isolated quadricuspid aortic valve is a rare clinical finding and cause significant aortic regurgitation in adulthood. It's believed that incomplete coaptation of leaflets and unequal stress to leaflets causing thinning are main reasons of aortic regurtation in quadricuspid aortic valves. In our case progressive calcification caused aortic valvular dysfunction.

In literature quadricuspid aortic valvular patients' mean age is 49 at diagnosis and 45.2% of them needed operation during their 5th-6th decades[7]. Our patient was diagnosed and treated during her 6th decade.

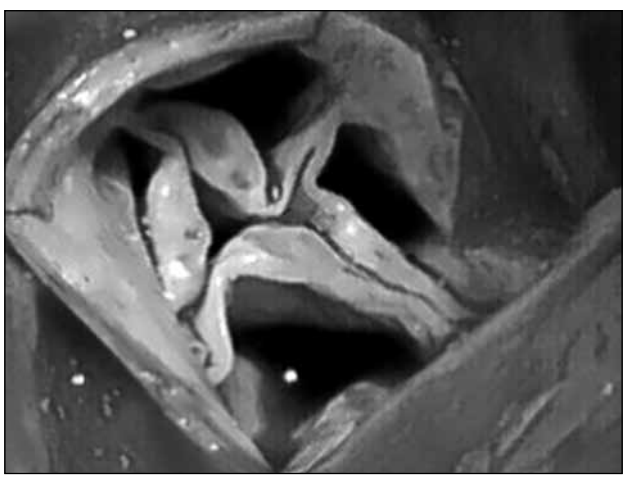

Figure 1. Intraoperative view of the quadricuspid aortic valve.

Hurwitz and Roberts classified this anomaly into seven anatomic variations (from A to G) according to the relative size of the four cusps[8]. B type is the most common one (three equal-sized cusps and one smaller cusp). Our patient had four equal-sized cusps (type A). Nakamura and colleagues designed a classification according to the position of the nondominant cusp[9]. The most common variation in their study was the presence of the supranumerary cusp between the right coronary and the noncoronary cusps.

18% of all quadricuspid aortic valvular cases have accompanying congenital and/or acquired cardiac anomaly including coronary artery anomaly and others. Also with decreasing frequency ventricular septal defect, atrial septal defect, sinus valsalva aneurysm, patent ductus arteriosus, Ehler Danlos' syndrome, subaortic fibromuscular stenosis, hypertrophic cardiomypathy, mitral valve pathologies, ascending aortic dilatation, pulmonic valve stenosis and left coronary atresia with supravalvular stenosis are additional other anomalies[10-12]. Therefore, patients with quadricuspid aortic valve should be examined for associated congenital anomalies during preoperative preparation and intraoperatively.

Structural valve degeneration may predispose to infective endocarditis. We did not encounter to any sign of infective endocarditis pre and post-operative period. Some pathologies such as bacterial endocarditis or rheumatic valves may mask quadricuspid aortic valve pathology and complicate the diagnosis[13].

In case of surgical indications biological or mechanical aortic prosthetic valve replacement is the most common method applied. It is reported that aortic valve sparing procedures is also available[14]. We have chosen biological valve for replacement because of patient's age and social status.

Although our patient's coronary ostiums were in their physiological positions, malpositioned coronary ostiums should be kept in mind.

As a result quadricuspid aortic valve is a rare condition and presents itself with aortic regurgitation in the adulthood. Preoperative transthoracic echocardiography is helpful in the diagnosis. Furthermore, advanced investigation is needed for accompanying anomalies. Surgical treatment either reconstruction or replacement is similar to tricuspid aortic valve disease.

CONFLICT of INTEREST

None declared.

REFERENCES

1. *Simonds JP. Congenital malformations of the aortic and pulmonary valves. Am J Med Sci 1923;166:584-95.*
2. *Tutarel O. Quadricuspid aortic valves and anomalies of the coronary arteries. J Thorac Cardiovasc Surg 2004;127:897.*
3. *Balington J, quoted by Robicsek F, Sanger PW, Daugherty HK, Montgomery CC. Congenital quadricuspid aortic valve with displacement of the left coronary orifice. Am J Cardiol 1969;23:288-90.*
4. *Di Pino A, Gitto P, Silvia A, Bianca I. Congenital quadricuspid aortic valve in children. Cardiol Young 2008;18:324-7.*
5. *Kurosawa H, Wagenaar SS, Becker AE. Sudden death in a youth. A case of quadricuspid aortic valve with isolation of origin of left coronary artery. Br Heart J 1981;46:211-5.*
6. *Timperley J, Milner R, Marshall AJ, Gilbert TJ. Quadricuspid aortic valves. Clin Cardiol 2002;25:548-52.*
7. *Tutarel O. The quadricuspid aortic valve: a comprehensive review. J Heart Valve Dis 2004;13:534-7.*
8. *Hurwitz LE, Roberts WC. Quadricuspid aortic valve. Am J Cardiol 1973;31:623-6.*
9. *Nakamura Y, Taniguchi I, Saiki M, Morimoto K, Yamaga T. Quadricuspid aortic valve associated with aortic stenosis and regurtitation. Jpn J Thorac Cardiovasc Surg 2001;49:714-6.*
10. *Vohra RK, Singh H, Siu BL, Failinger CF. A quadricuspid aortic valve with atrial septal defect. Echocardiography 2006;23:865-8.*
11. *Handoko ML, Paulus WJ. Polishing the diastolic dysfunction measurement stick. Eur J Echocardiogr 2008;9:575-7.*
12. *Janssesns 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;83-7.*
13. *Takeda N, Ohtaki E, Kasegawa H, Tobaru T, Sumiyoshi T. Infective endocarditis associated with quadricuspid aortic valve. Jpn Heart J 2003;44:441-5.*
14. *Van Son JAM, Reddy VM, Black MD, Rajasinghe H, Haas GS, Hanley FL. Morphologic determinants favoring surgical aortic valvuloplasty versus pulmonary autograft aortic valve replacement in children. J Thorac Cardiovasc Surg 1996;111:1149-57.*