Case Report



Quadricuspid aortic valve: a word of caution!

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ABSTRACT

Quadricuspid aortic valve is a rare cardiac malformation that usually appears as an isolated anomaly. It may cause either pure regurgitation or regurgitation accompanied by stenosis. Herein, we report a 54-year-old male patient who was referred to us for the surgical management of severe aortic valve regurgitation. Although preoperative echocardiography showed tricuspid aortic valve, we detected quardicuspid aortic valve intraoperatively. Aortic valve was replaced with great caution to the coronary ostia and conduction system. Postoperative course was uneventful. As quadricuspid aortic valve might be encountered incidentally during surgery, surgeons should be aware of anatomical difficulties with this morphology.

Keywords: Mechanical valve; quadricuspid, aortic valve; valve replacement.

Quadricuspid aortic valve (QAV) is a rare developmental cardiac anomaly. Its estimated incidence is 0.0031-0.043% of all congenital heart diseases.^[1] It usually appears as an isolated anomaly; however, several other malformations may accompany, the most common being coronary artery ostial anomalies.^[1,2] Advances in diagnostic modalities enabled us to diagnose this anomaly with using noninvasive techniques; however, most of the reported cases have still been diagnosed incidentally at autopsies or during cardiac surgery.^[2] It frequently leads to aortic regurgitation or aortic stenosis with regurgitation and eventually requires surgery. Therefore, prompt detection is of utmost importance in this population.

CASE REPORT

A 54-year-old man was admitted to our clinic for aortic valve replacement. His medical history revealed acute rheumatic fever at the age of 12 years. He was New York Heart Association (NYHA) class II. Transthoracic echocardiography showed severe aortic regurgitation and mild mitral regurgitation with a left ventricular end-diastolic diameter of 56 mm and an ejection fraction of 69%. Aortic valve was reported as tricuspid and heavily calcified. Preoperative coronary angiography revealed normal coronary arterial anatomy with mild atherosclerosis. Aortic root angiography confirmed severe aortic regurgitation. Elective surgery was planned for the valve replacement. A written informed consent was obtained from the patient.

Following a median sternotomy, cardiopulmonary bypass was initiated with aortic and single atrial cannulation. Following antegrade cardioplegic arrest, a transverse aortotomy was made and aortic valve was inspected. As opposed to preoperative echocardiography, aortic valve was quadricuspid with mild thickening and calcification. The valve had one large, two intermediate and one small cusp (Type D in Hurwitz and Roberts' anatomical classification). Accessory cusp was located between the right and left coronary cusps (Figure 1). There was no abnormality regarding coronary arteries. Native valve was excised and a 25 mm mechanical bileaflet valve was implanted. Postoperative period was uneventful and he was discharged in the fifth postoperative day. Followup echocardiography revealed that the valve was functioning well, the left ventricular dimensions and functions were preserved.

DISCUSSION

Among patients undergoing aortic valve replacement, the incidence of QAV ranges from 0.55% to 1.46%.^[2] Since QAV is not considered in the differential diagnosis of aortic valve regurgitation, preoperative diagnosis with transthoracic echocardiography

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Figure 1. Accessory cusp (AC) located between right coronary cusp (RCC) and left coronary cusp (LCC). Note that non-coronary cusp (NCC) is large, RCC and LCC are intermediate, and the accessory cusp is small (Type D).

is highly challenging. In a series of 627 patients undergoing aortic valve surgery, only three QAV cases were encountered and none of them had been diagnosed preoperatively.^[1] Therefore, authors concluded that real-time three-dimensional transesophageal echocardiography might be helpful in diagnosis.

Hurwitz and Roberts' anatomical classification is the most commonly used systematization tool, which classifies QAV into seven types depending on the relative size of the valve leaflets (Type B is the most common). The most common positions of the accessory cusp is between the right and non-coronary cusps.^[3] Our case is unique and extremely rare considering the size and positional classification of QAV (Type D and accessory cusp between right and left coronary cusps).

Review of the literature revealed that several cardiac abnormalities may coexist with QAV: anomalies of the coronary arteries, ventricular septal defect, patent ductus arteriosus, and pulmonary valve stenosis. As one of the most common accompanying anomaly, displacement of the coronary ostia has been reported in approximately 10% of cases.^[1,3] From the surgical point of view, it is critical to inspect each

coronary ostium to prevent ostial obstruction at the time of valve surgery. In our case, no accompanying ostial anomaly was detected.

Another pitfall for surgeon is the downward displacement of the annulus in QAV setting, which may cause complete heart block during valve replacement.^[4] In this case, it is recommended to place supra-annular sutures, which lie anteriorly high within the membranous septum. We also placed valve sutures high in between non-coronary and right coronary cusps in our case. The patient had no rhythm disturbances during the postoperative course.

Despite aortic valve replacement for QAV is a traditional treatment of choice for patients with aortic regurgitation, there are only few reports utilizing aortic valve repair techniques (tricuspidization or bicuspidization) in QAV cases.^[5] Short-term results are satisfactory; however, further studies and continued follow-up are warranted to elucidate long-term results.

In conlusion, cardiac surgeons keep in mind that QAV is a rare developmental anomaly but possesses low risk in terms of surgical outcome.

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REFERENCES

- 1. Hayakawa M, Asai T, Kinoshita T, Suzuki T. Quadricuspid aortic valve: a report on a 10-year case series and literature review. Ann Thorac Cardiovasc Surg 2014;20:941-4.
- 2. Gouveia S, Martins JD, Costa G, Paramés F, Freitas I, Rebelo M, et al. Quadricuspid aortic valve--10-year case series and literature review. Rev Port Cardiol 2011;30:849-54.
- 3. Timperley J, Milner R, Marshall AJ, Gilbert TJ. Quadricuspid aortic valves. Clin Cardiol 2002;25:548-52.
- 4. Pirundini PA, Balaguer JM, Lilly KJ, Gorsuch WB, Taft MB, Cohn LH, et al. Replacement of the quadricuspid aortic valve: strategy to avoid complete heart block. Ann Thorac Surg 2006;81:2306-8.
- Song MG, Yang HS, Lee DH, Shin JK, Chee HK, Kim JS. Mid-term results in patients having tricuspidization of the quadricuspid aortic valve. J Cardiothorac Surg 2014;9:29.