Brief Communication

Quadricuspid Aortic Valve: Interesting Images

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ABSTRACT

The quadricuspid aortic valve is a rare congenital anomaly, usually associated with aortic regurgitation requiring surgical intervention. It may be associated with other congenital anomalies such as coronary anomalies, patent ductus arteriosus, ventricular septal defect, pulmonary stenosis, and subaortic stenosis. The diagnosis is generally established by either transthoracic or transesophageal echocardiography. Herein, we report a case of a 52-year-old woman who was diagnosed to have quadricuspid aortic valve by intraoperative transesophageal echocardiography.

Keywords: Aortic regurgitation, quadricuspid aortic valve, transesophageal echocardiography

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INTRODUCTION

Congenital quadricuspid aortic valve (QAV) is a rare malformation with an estimated incidence of 0.01% to 0.04%.[1] Most cases are discovered as incidental finding during aortic valve surgery or autopsy. The condition frequently progresses to aortic regurgitation, which generally manifests in the fifth decade of life and may require surgical treatment.

CASE REPORT

A 52-year-old woman with severe aortic regurgitation was admitted for aortic valve replacement. Preoperative transthoracic echocardiography has reported a tricuspid aortic valve; however, intraoperative transesophageal echocardiography (TEE) showed a QAV in mid-esophageal aortic valve short-axis view [Figure 1a and 1b, Video 1]. Color Doppler images of the aortic valve showing aortic regurgitation are shown in Videos 2 and 3. The left and right coronary cusps (1 and 2) were large, while the noncoronary and accessory cusps (3 and 4) were smaller in size, making it type "C" QAV according to the Hurwitz and Roberts classification [Figures 1 and 2].[2] The other cardiac valves, ascending aorta, left ventricular function, and left ventricular end-diastolic dimension were normal. The patient underwent aortic valve replacement with a mechanical prosthesis uneventfully.

DISCUSSION

The QAV, first reported by Balinton in 1862, is a rare congenital anomaly seen in 0.01–0.04% of the population.[1,3] Technological advances in echocardiography and magnetic resonance imaging have enabled the increased diagnosis of this anomaly even in asymptomatic individuals. The symptoms appear with an increase in the severity of aortic regurgitation and the development of left ventricular dysfunction. Embryologically, the fourth cusp arises during the early stage of truncal septation, resulting in either a different number of primordial aortic leaflets or abnormal cusp

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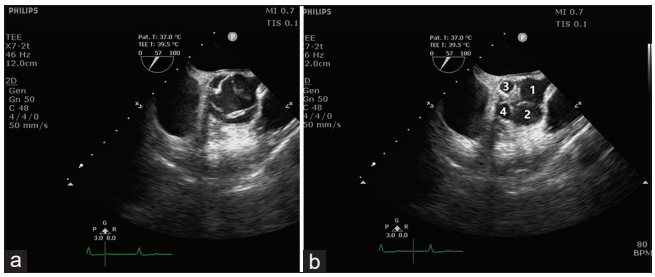

Figure 1: Mid-esophageal aortic valve short-axis view on transesophageal echocardiography showing a "rectangular" opening of the quadricuspid aortic valve during systole (a) and an "x" or cross-shaped appearance during diastole (b). Cusp 1: left coronary, cusp 2: right coronary, cusp 3: noncoronary, cusp 4: accessory cusp

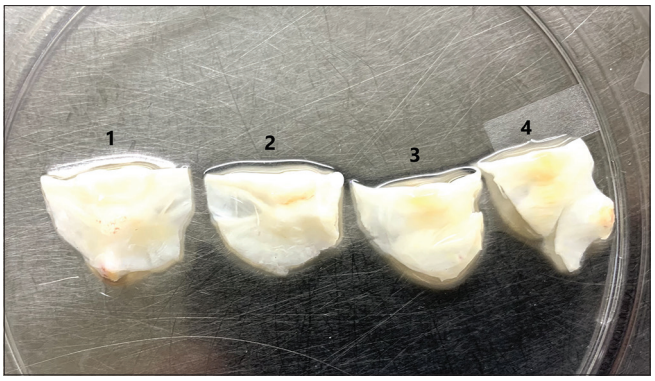

Figure 2: Photograph taken after excision of the cusps showing left coronary (1), right coronary (2), noncoronary (3), and an accessory cusp (4)

proliferation.[4] The proposed mechanism of aortic regurgitation includes age-related progressive leaflet fibrosis, unequal distribution of stress, and abnormal leaflet coaptation. The associated anomalies include coronary anomalies, patent ductus arteriosus, ventricular septal defect, pulmonary stenosis, and subaortic stenosis. These patients are at risk of coronary ostial obstruction or injury at the time of valve replacement due to the abnormal origin and distribution of coronary arteries.[5] Seven different anatomical types of QAV have been described by Hurwitz and Roberts.[1] The patient described here had type "C" QAV with two larger and two smaller cusps. Previous reports have described type A and type B as the most common variants, but it is unclear whether any specific subtype of QAV predisposes the patient to more severe aortic regurgitation.[5,6]

In conclusion, QAV is a rare congenital anomaly and the majority of these patients will require surgery for aortic regurgitation. Once diagnosed, patients should be closely followed so that aortic valve replacement or repair is done before left ventricular decompensation occurs.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initial s will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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