Quadricuspid Aortic Valve: A Rare Intraoperative Diagnosis by Transesophageal Echocardiography

Abstract

Quadricuspid aortic valve (QAV) is a rare congenital anomaly frequently associated with other anomalies particularly coronary anomalies. It may be detected on transthoracic or transesophageal echocardiography. We present here a case report of a 27-year-old male patient with a QAV, the valve being regurgitant and requiring aortic valve replacement. It has been reported as isolated case reports in the literature and various theories exist to the development of QAV. The diagnosis requires a high degree of suspicion and a detailed assessment, and if asymptomatic, then patients need to be carefully followed up for the development of aortic regurgitation.

Keywords: *Congenital, coronary ostium anomaly, echocardiography, quadricuspid aortic valve, regurgitation*

Anupam Das, Ummed Singh[1], Palleti Rajashekar

Departments of Cardio Thoracic Vascular Surgery and [1]Cardiac Anaesthesia, All India Institute of Medical Sciences, New Delhi, India

A 27-year-old male patient presented with gradually progressive palpitations and exertional dyspnea for 5 years. Clinical examination revealed normal vital signs with a blood pressure of 138/38 mmHg. Cardiac examination showed apical impulse in the left sixth intercostal space 1.5 cm lateral to the midclavicular line. Auscultation revealed a grade 4/6 diastolic murmur in the second right intercostal space. Electrocardiogram showed normal sinus rhythm with normal axis. The chest radiograph showed cardiomegaly with a cardiothoracic ratio of 0.6. Preoperative two-dimensional echocardiography revealed severe aortic regurgitation in a tricuspid aortic valve with left ventricular ejection fraction of 50%. Intraoperatively, the transesophageal echocardiography revealed and confirmed the presence of a quadricuspid aortic valve (QAV) with the accessory cusp being rudimentary and placed between the right and left coronary cusps [Video 1 and Figure 1]. Cusps were found to be normal with no evidence of thickening and calcification. After the establishment of cardiopulmonary bypass with standard aortic and two-stage venous cannulation, ostial cardioplegia was planned. Aortotomy was done after application of the aortic cross clamp and anatomy of the valve was assessed. Ostial cardioplegia was administered in the left coronary ostium [Figure 2]. The right coronary ostium was found to be anomalously present near the accessory fourth cusp [Figure 3]. The cusps were found to be normal, but coaptation was found to be inadequate. The right aortic sinus was found to be dilated. Aortic valve replacement was done with mechanical prosthesis after native valve excision. The patient recovered uneventfully and was discharged on the 6th postoperative day.

Discussion

QAV is a rare congenital anomaly with an incidence of <1.5%.[1] The anomaly usually presents with aortic regurgitation requiring surgical intervention in most instances. It is frequently associated with coronary ostial anomalies[1] and may present in adulthood. First reported by Balinton in 1862,[2] QAV was previously detected only during autopsy. With technological advances in the form of transesophageal echocardiography, cardiac magnetic resonance imaging, there has been increased diagnosis of this congenital anomaly even in asymptomatic individuals. Coronary ostial anomalies associated with the QAV has previously been reported by Miyata *et al*.[3] Hurwitz and Roberts[4] classified this QAV into seven different anatomical variations: type A-four equal cusps; type B-three equal cusps and one smaller cusp; type C-two equal larger cusps and two equal smaller cusps; type D-one large, two intermediate, and one small cusp; type E-three equal cusps and one larger cusp; type F-two equal larger cusps

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Address for correspondence:
Dr. Anupam Das, B-2/56-B, Lawrence Road, Keshav Puram, New Delhi - 110 035, India.
E-mail: dranupamdas21@gmail.com

Video Available on: www.annals.in

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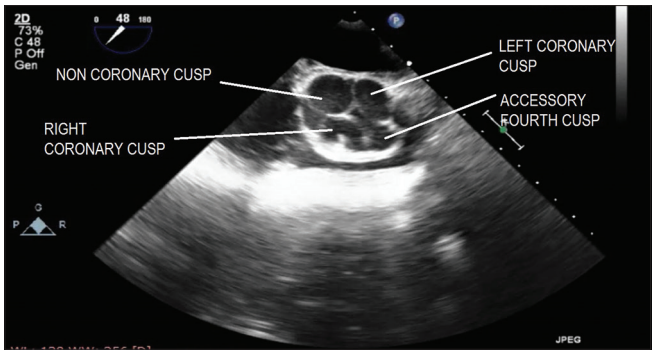

Figure 1: Two-dimensional transesophageal echocardiography (midesophageal aortic valve short axis view) showing the quadricuspid aortic valve

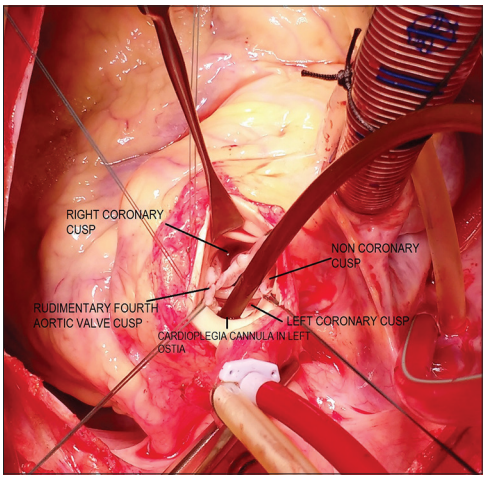

Figure 2: Surgeon's view of the quadricuspid aortic valve with the cardioplegia cannula in the left coronary ostium

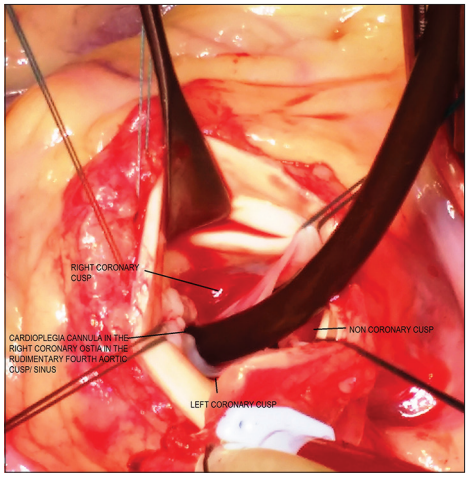

Figure 3: Cardioplegia being delivered in the right coronary ostium located near the accessory cusp of the aortic valve

and two unequal smaller cusps; and type G-four unequal cusps. Our case report was the type B QAV which is also the most commonly encountered type and is associated with unequal shear stress leading to regurgitation. QAV may often be associated with patent ductus arteriosus, ventricular septal defect, pulmonary valve stenosis, subaortic fibromuscular stenosis, and anomalies of the coronary arteries.[1] Transthoracic echocardiography plays an important role in detecting congenital anomalies of the aortic valve preoperatively. However, it may be suboptimal for recognizing QAV and associated malformations due to the poor acoustic window. Transesophageal echocardiography provides reliable imaging of the aortic valve as in our case. The importance of diagnosis of the QAV lies in the fact that most patients will eventually require surgery for aortic regurgitation before the left ventricle decompensation sets in. Associated anomalies should be kept in mind and looked for so that no residual lesion is left behind. Aortic valve replacement is required in most cases and can be performed without any deviation from the standard procedure. Although a congenital anomaly, QAV may not present itself before fourth or fifth decade when it may result in significant regurgitation. Although the risk of endocarditis is still not clear, confirmed cases of endocarditis affecting QAVs have been reported.[5] Therefore, whether detected incidentally and found to be normally functioning, either on echocardiography or intraoperatively, it is critical to follow-up these patients closely to detect the development of regurgitation or endocarditis at the earliest before significant left ventricular compromise develops.

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Conflicts of interest

There are no conflicts of interest.

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