

LEFT MAIN CORONARY ARTERY ARISING FROM THE RIGHT SINUS OF VALSALVA WITH BICUSPID AORTIC VALVE RARE CORONARY ARTERY ANOMALY: A CASE REPORT

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Koroner anomaliler ile birlikte olan bikuspid aort kapakları, sık görülen malformasyon değildir.

Bu yazıda, her iki anomalinin bulunduğu bir kadın hasta sunulmuştur. Hasta başarılı aort valv replasmanına gitti.

INTRODUCTION

Coronary anomalies are rare congenital cardiac malformations that occur in less than 1% of the population. When present, these malformations are usually an ectopic origin of the right or left coronary artery from the aorta. In rare circumstances, coronary anomalies can result in myocardial malperfusion with devastating clinical consequences¹. In cases of anomalous origin of the left coronary artery (LCA) from the right sinus, the clinical significance is mainly determined by the course of the artery. Cases of sudden death, especially of young people during exercise are mainly reported in intertruncal course between aorta and pulmonary artery. Symptoms of this type of anomaly are disturbances of rhythm, exercise related angina pectoris, syncope and sudden death, even as first manifestation².

The bicuspid aortic valve (BAV) affects 1 to 2% of the population and may be complicated by aortic stenosis or aortic insufficiency and infective endocarditis. Bicuspid aortic valve is associated with abnormalities of the aortic wall such as coarctation of the aorta, aortic dissection, and aortic aneurysm. Most patients with BAV will develop some complication during life³. Patients with BAV may have a coexisting coronary anomaly³⁻⁵. However, this condition is not common.

A case with anomalous origin of the Left main coronary artery from the ascending aorta above the right

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sinus of Valsalva is described. This patient was also found to have a bicuspid aortic valve.

CASE

A forty years old female patient has applied to our hospital with complaints of chest pain and palpitation. Her blood pressure 180 / 110 mmHg, physical examination revealed a grade 5 systolic murmur on aortic area. Aortic systolic murmur did not show longitudinal expansion. Transthoracic echocardiography was performed, showed a severe bicuspid aortic valve stenosis with mean pressure gradient of 70 mm Hg and left ventricular hypertrophy. Left ventricular function was normal. Transesophageal echocardiography (TEE) showed BAV (Figure-1). Cardiac catheterization confirmed the diagnosis of severe aortic stenosis with peak systolic gradient of 90 mm Hg. Selective coronary angiography, ventriculography and aortography have been performed. No origin of the coronary ostium arising from the left sinus Valsalva was imaged. However, origin of left main coronary artery arises from the right sinus Valsalva. Origin of right coronary artery was normal (Figure-2,3). Significant stenosis was not encountered in any of the coronary arteries imaged. Our patient was discharged with oral anticoagulant therapy after successful aortic valve replacement surgery.

DISCUSSION

The bicuspid aortic valve affects 1 to 2% of the population and may be complicated by aortic stenosis or aortic insufficiency and infective endocarditis. In this patient, severe aortic stenosis was determined, although aortic insufficiency was not. Bicuspid aortic

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Figure 1: Transesophageal echocardiography showing bicuspid aortic valve

Figure 2: Abnormal exit of the left coronary artery and post-stenotic dilatation in aortography

Figure 3: Coronary angiograph shows left main coronary artery arises from the right sinus Valsalva and right coronary artery

valve is associated with abnormalities of the aortic wall such as coarctation of the aorta, aortic dissection, and aortic aneurysm³. Presented patient has not aortic dissection and aortic aneurysm.

Coronary anomalies are rare congenital cardiac malformations that occur in less than 1% of the population¹. Cases of anomalous origin of the left coronary artery from the right sinus of Valsalva are classified according to their course into four categories. 1-Intertruncal course with predisposition to sudden death, especially during exercise in childhood and adolescence. Slit like origin and intertruncal compression during systolic pressure rise in the aortic root are thought to be the mechanical factors for sudden death. Several reports on sudden death in this

anomaly have been published. 2- Anterior free wall course before the right ventricular outflow; according to the literature this course is clinically insignificant. 3- A posterior course with the left coronary artery or one of the great branches passing behind the aorta. This course is thought to be clinically insignificant as well. 4- An intertruncal-septal course through the crista supraventricularis^{2,6} Cheitlin et al⁷. have pointed out the pathologic significance of a single coronary artery or of both coronary arteries from the right sinus of Valsalva when the branch or artery that supplies the left coronary distribution courses leftward between the aorta and pulmonary artery so that it can be compressed. They showed a striking incidence of sudden unexplained death in adolescents and young adults, and a strong relation of death to heavy exertion. For this reason, any child or young person with angina pectoris, myocardial infarction or cardiac syncope should have this anomaly ruled out by coronary angiography. Left coronary artery corresponds to type 3 since it indicates posterior pattern. These are generally clinically unimportant cases and do not cause perfusion disorder hemodynamically. Patients with BAV disease may have a coexisting coronary anomaly^{3,5}. In our case with BAV both right and left main coronary arteries arise from right sinus Valsalva.

The patients with BAV when undergo valve repair or replacement, there is a potential risk of coronary injury during surgery⁸. The preoperative determination of the presence and location of an anomalous coronary artery, this state may reduce the risk of injury at the time of surgery.

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