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Anatomical History of a Single Coronary Artery Anomaly: A Case Report

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ABSTRACT

Single coronary artery (SCA) anomaly is a rare anomaly, where the right and left coronary arteries arise from a single ostium in the sinus Valsalva and feed the whole heart. Although asymptomatic in most cases, it can cause angina, syncope, myocardial infarction, and sudden cardiac death. It is essential to diagnose it, especially in terms of the risk of encountering clinical pictures, such as sudden death at a young age, and its association with other congenital anomalies. In this report, we present a patient with a SCA anomaly. During an emergency conventional angiography, a right coronary artery (RCA) arising from the left main coronary artery (LMCA) originating from a single ostium was detected in the aorta.

INTRODUCTION

The incidence of incidental and rare coronary artery anomalies (CAA) in patients undergoing coronary angiography (CAG) is estimated to be between 0.2% and 1.3% [Al-Sadawi 2019]. CAA are the second most common cause of sudden cardiac death among young athletes after hypertrophic cardiomyopathy [Al-Sadawi 2019; Satija 2012; Swaminath 2013]. SCA is a rare congenital anomaly in which there is an isolated coronary artery originating from a single coronary ostium and providing coronary blood supply to the entire myocardium. SCA is divided into different types based on origin, branching pattern and course. Although most patients with SCA are asymptomatic, some patients may present with life-threatening symptoms [Al Umairi 2019]. Currently, the prevalence of RCA origination from the left coronary sinus occurs between 0.019% and 0.49% [Al-Sadawi 2019; Lee 2009]. RCA originating from the LMCA constitutes only 0.65% of these anomalies [Lee 2009]. Although CAA are rare, they can be found in many configurations, where each case is unique in terms of origin, continuation, and termination of a major coronary artery. For example, abnormal arising of the LMCA or RCA from the aorta, which is the course between the aorta and the pulmonary trunk [Al-Sadawi 2019; Swaminath 2013]. This course is malignant because of the risk of arterial compression, which can lead to life-threatening consequences such as ischemic heart disease and sudden cardiac death in young adults. Because of the clinical significance of these anomalies, it is important to identify them as a measure of primary prevention [Lumia 2007]. Percutaneous coronary intervention (PCI) of SCA is technically challenging because it requires careful assessment of its origin and starting point, course of the vessel, and location of the lesion. Selection of the appropriate hardware is critical for a successful PCI and optimization with imaging and plaque modification [Vijayvergiya 2020].

CASE REPORT

A 46-year-old male patient presented to the emergency department of our hospital with occasional chest pain and shortness of breath. The patient's tests were negative for troponin levels, his electrocardiogram was unremarkable. Coronary angiography (CAG) was performed in the patient with 18 years of smoking, hypertension, and exertional angina as risk factors for coronary artery disease. Following emergency premedication and local anesthesia to the patient, the right femoral artery was entered by puncture, and selective

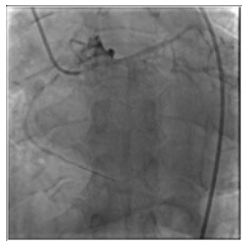

Figure 1. CAG demonstrating before PCI an anomalous RCA originating from the LMCA.

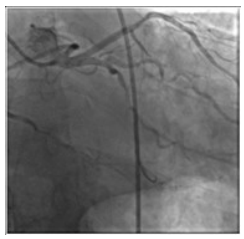

Figure 2. CAG demonstrating after PCI an anomalous RCA originating from the LMCA.

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Angiographic classification of Lipton's Modified Single coronary artery anomaly

	KFA	Definition
Osteal Placement	R (right)	Right sinus of Valsalva
	L (left)	Left sinus of Valsalva
Anatomical distribution	I	SCA follows normal right or left coronary artery course (RI or LI)
	II	After a SCA leaves the right or left coronary sinus, it crosses the base of the heart in a wide transverse trunk to supply the contralateral coronary artery.
	III	SCA originates from the right sinus, RCA after the exit, the LAD and Cx arteries arise from a SCA trunk separately, not from a single trunk.
Transverse Branch Course	A	In front of the great vessels (in front of the right ventricle)
	B	Between the aorta and the pulmonary artery
	P	Behind the great veins
	S	Septal type (through the interventricular septum), combined type

KFA, key for abbreviations

angiography and left ventriculography were performed in RAO (right anterior oblique) and LAO (left anterior oblique) positions. During diagnostic coronary angiogram, multiple attempts to cannulate the RCA with a right Judkins (4.5 cm) diagnostic catheter were unsuccessful and, instead, a right amplatz coronary catheter was used. In the left ventriculography, it was observed that the LMCA, which originated from the left sinus Valsalva abnormally, emerged 5 mm from the ostium, and the RCA coursed between the aorta and the pulmonary trunk (Figures 1 and 2). In the left coronary arteriography of the patient, LMCA plaqued, LAD (left anterior descending) plaqued, 99% thrombotic diffuse stenosis from middle to distal, circumflex artery (Cx) plaqued, diffuse 70% stenosis in the distal; and, in the right coronary arteriography, RCA plaqued, diffuse 99% stenosis from proximal to middle were detected (Figure 1). Percutaneous transluminal coronary angioplasty (PTCA) with a 2×15mm balloon to the stenosis in the LAD during CAG was applied to the patient, then 2.25×28mm, 2.5×28mm, 2.75×16mm stents were inserted and full patency was achieved. PTCA was performed with a 2×15mm balloon to the stenosis in the RCA, followed by 2.25×18mm, 2.25×24mm, and 2.5×14mm stents, providing full patency in all coronary arteries (Figure 2). We performed computed tomography angiography to re-evaluate the patient's coronary arteries before discharge, which supported the RCA from the LMCA originating from the single coronary ostium in the LSV (left sinus of Valsalva (Figures 3 and 4). The patient's treatment was arranged with

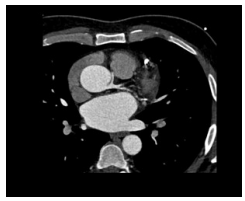

Figure 3. CT coronary angiography image of an anomalous origin of RCA from LMCA (View 1).

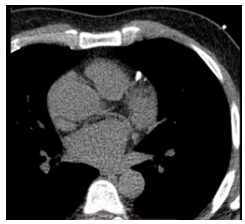

Figure 4. CT coronary angiography image of an anomalous origin of RCA from LMCA (View 2).

acetylsalicylic acid 100 mg/day, metroprolol succinate 25 mg/day, ticagrelor 90 mg twice a day, and he was discharged two days later with the recommendation of lifestyle changes. The patient was also referred to the smoking cessation outpatient clinic in our hospital. At the patient's first month follow up, we performed stress myocardial perfusion scintigraphy, which showed no ischemia and EF (ejection fraction) was in normal range. No arrhythmias occurred during Holter implantation, so no ICD (implantable cardioverter defibrillator) was required. Therefore, the cardiovascular surgeon will re-evaluate the patient's condition for coronary artery bypass grafting after the patient's follow-up angiography at the end of the first 6 months.

DISCUSSION

CAA is associated with abnormal regression or persistence of primitive coronary arteries in embryological development. Initially, six coronary arteries develop, three of which originate from the aorta and three from the pulmonary artery. Except for two aortic origins, they normally regress and disappear. The development of coronary artery anomaly depends on the abnormalities in this process [Vrancken 1997]. With the widespread use of CAG, it has started to be observed more frequently and its clinical importance has been better understood. Ten percent of those with coronary artery anomalies are accompanied by other congenital heart diseases [Yamanaka 1990]. Although most of the cases with CAA are asymptomatic, it can cause arrhythmia, heart failure, myocardial ischemia, angina, syncope, myocardial infarction, and sudden death. The symptomatic nature of the patients is mostly related to the myocardial tissue size at risk and the character of the anomaly [Frescura 1998; Roberts 1986]. There are many variations of a single coronary artery anomaly and 23 different forms have been described [Taylor 2001]. Its

incidence in various angiographic series have been reported to be between 0.02 and 0.04% [Shirani 1993]. It is observed in approximately 1/2250 of patients undergoing coronary angiography [Walker 2001]. In a study, 19% of sudden deaths in young athletes were associated with a single coronary artery anomaly, and it was reported as the second most common cause of sudden death in young athletes after hypertrophic cardiomyopathy [Maron 1996; Maron 2003]. The type in which the RCA originates from the left single coronary artery and travels between the aorta and pulmonary artery roots was found to be the one most associated with sudden death [Roberts 1986]. Since these cases frequently are lost in the early period, their prevalence in angiography series is low. Surgical treatment is required in symptomatic patients, in cases where the course of the artery is dangerous, and in patients diagnosed at a young age [Lipton 1979]. Surgical approach can be applied in the form of repositioning the anomalous artery as suitable to the coronary sinus or coronary bypass. However, coronary bypass is used as a more effective and reliable method [Koşar 2006]. Lipton et al divided single coronary artery cases into three groups angiographically [Lipton 1979]. They included Group 1: SCA follows the course of right or left coronary artery; Group 2: After the SCA leaves the right sinus of Valsalva (RSV) or LSV, it crosses the base of the heart in a wide transverse trunk to supply the contralateral coronary artery; and Group 3: This group applies to cases where a SCA originates from RSV, after exit LAD and Cx arise from the coronary artery trunk separately, not with a single trunk [Lipton 1979]. The Cx artery follows a retroaortic course and enters the left atrioventricular sulcus, while the LAD passes between the aorta and the main pulmonary trunk and enters the anterior interventricular sulcus [Lipton 1979]. SCA anomaly is a rare coronary anomaly in which all coronary artery branches originate from a single ostium. SCA anomalies are classified according to where they originate from the right or left coronary artery, anatomical distribution on the ventricular surface, and relationship with the great artery [Lipton 1979] (Table). In our case, it was a SCA anomaly, which sets an example for group 2. We wanted to draw attention to some points by presenting a case in which a SCA, the RCA, originated from the left system. This case shows a rare anomaly. However, it should be noted that it may have clinically important consequences, and therefore it is important to evaluate any dynamic external compressions. This finding is an extremely rare variant of a SCA. The reported incidence of a single coronary system originating from the left sinus of Valsalva is 0.04%, and an RCA originating from the left main trunk constitutes 0.65% of these [Walker 2001; Bromage 2011]. It also has been reported that myocardial ischemia develops in cases, where the RCA originates from LAD or Cx. In rare cases, cardiomyopathy and congestive heart failure have been reported as a result of ischemia in patients with a single coronary artery. PCI successfully can be applied to atherosclerotic lesions in single coronary artery anomalies, following the necessary preparations. Care should be taken when performing angioplasty. Temporary osteal obstruction caused by large diameter catheters used during the procedure may not be tolerated by these patients, chest pain, dizziness, dyspnea, hypotension, and hemodynamic deterioration may occur [Frescura 1998; Iyisoy 2002]. In the presence of ischemia, all treatment options, including surgery, should be considered for the treatment of ischemia. Unexpected complications are encountered during cardiac surgery in SCA anomaly. Cases that resulted in intraoperative mortality as a result of coronary artery ligation during mitral and aortic valve interventions have been reported [Taylor 2001; Iyisoy 2002].

CAG should be performed before cardiac surgery, especially in congenital heart diseases, to avoid unexpected complications.

CONCLUSION

Although SCA anomalies are rare anomalies, because they cause life-threatening conditions, treatment should be planned considering clinical symptoms, presence of atherosclerotic heart disease, and type of coronary anomaly. It is important that the interventional cardiologist and cardiovascular surgery determine the most appropriate treatment option together in these anomalies, which are more common in large centers where CAG frequently is performed.

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