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AN UNUSUAL RIGHT CORONARY ARTERY EMERGING FROM DISTAL CIRCUMFLEX ARTERY

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ABSTRACT

Introduction: Coronary artery anomalies (CAAs) affect around 1% of the general population, ranging from 0.3%-5.6% in studies on patients undergoing coronary angiography and in approximately 1% of routine autopsies. One of the rarest coronary artery anomalies is the single coronary artery (SCA). The prevalence of SCA is approximately 0.024% to 0.066% in patients who undergo coronary angiography. The SCA are asymptomatic in general. In some cases, the disease may appear with life-threatening symptoms.**Case Description:** A 53-year-old male was admitted to the hospital due to chest pain. He underwent coronary angiography. The angiography showed a very short left main coronary artery diving into LAD and LCX arteries in several millimeters. He had a big which had the same diameter in the distal part as the proximal part. After giving a big obtuse marginal branch, a smaller obtuse marginal branch, and a posterolateral branch, the circumflex continued to the right side of the heart with a slightly smaller diameter than the preceding parts. It was found that the patient had an anatomically different RCA, which originates from the distal circumflex artery. According to Lipton et al., our case represents an L-I type.**Conclusion:**A single coronary artery is commonly associated with other congenital cardiac anomalies, such as bicuspid aortic valve, coronary arteriovenous fistula, and transposition of the great arteries.SCA is a rare coronary artery anomaly. A detailed anatomical description of this anomaly will help diagnose coronary artery abnormalities.

Keywords: Coronary artery anomalies (CAAs), Single coronary artery (SCA), Coronary angiography, Lipton classification

Introduction:

The congenital anomalies and acquired diseases of the heart may cause sudden cardiac deaths(Schiavone, Gobbi et al. 2021). Coronary artery anomalies (CAA), which are rare congenital anomalies, may be asymptomatic and may remain silent or cause life-threatening symptoms, including arrhythmias, syncope, myocardial infarction, and sudden death(Lau, Lee et al. 2023).

In recent years, coronary artery anomalies (CAA) have become increasingly important due to their relation to sudden cardiac death, syncope, myocardial infarction (MI), and angina pectoris(Sidhu, Wander et al. 2019). CAA can be detected by autopsy, coronary angiography, echocardiography, computed tomography angiography, or cardiac magnetic resonance imaging. They are often detected incidentally at 0.6%-1.3% in coronary angiography and can be detected as isolated anomalies or can be detected with complex cardiac abnormalities, including tetralogy of Fallot, transposition of the major vessels, single ventricle and the coarctation of aorta(Barriales Villa, Morís et al. 2001, Pourafkari, Taban et al. 2014).

The CAA classification is divided into three groups based on the coronary artery anatomy. Group 1 according to the origin and course of the coronary artery. Group 2: coronary artery anomalies of intrinsic coronary artery anatomy, including myocardial bridging and the aneurysm. Group 3: according to the termination of the coronary artery(Angelini 2007, Kayalar, Burkhart et al. 2009, Ghadri, Kazakauskaite et al. 2014).

In the single coronary artery (SCA) anomaly, the heart's blood supply is provided by an isolated coronary artery originating from a single ostium. SCA is a rare form of coronary artery anomaly with an incidence of 0.03%-0.4% of the population(Harikrishnan, Jacob et al. 2002, Cademartiri, La Grutta et al. 2008, Kayalar, Burkhart et al. 2009).Coronary artery anomalies are the second most common cause of sudden cardiac death (Maron, Haas et al. 2009). An atherosclerotic lesion in individuals with a single coronary artery may cause ischemia in a large myocardial region or whole heart. This condition is one of the causes of sudden death secondary to exercise, particularly in young individuals(Yin, Jin et al. 2017).

Embryology and anatomy of the coronary arteries

Coronary artery anomaly is associated with abnormal regression or persistence of primitive coronary arteries in the embryological period. It is seen in 1-2% of the general population.

Coronary arteries are formed before the development of arterial orifice in aortic sinuses. Primitive vessels developing from the subepicardial space invade the myocardium and spread to the myocardial surface. These sprouted vessels are then connected with the aortic root. Initially, six coronary arteries develop, three of which originate from the aorta and three from the pulmonary artery. Except for two coronary arteries that generally arise from the aorta, they disappear by regression. The persistent arteries are called

the right and left coronary arteries. In one-third of humans, small accessory branches of the coronary arteries may exist (Eralp, Lie-Venema et al. 2005).

The coronary arteries, responsible for the arterial supply of the heart, are the first branch of the aorta ascendens. They are located in the subepicardial connective tissue and on the surface of the heart. Coronary arteries are not the end arteries, but functionally, they behave like end arteries. They are vasa vasorum of the ascending aorta. In regular anatomy, the right and left coronary arteries (RCA and LCA, respectively) originate from the right and left sinus Valsalva of the aortic root. RCA originates lower than the left main coronary artery (LMCA) origin(Kini, Bis et al. 2007). The right sinus is located anterior, and the left sinus is located posteriorly on the aortic root. RCA supplies blood to the right atrium, right ventricle, and often the left ventricular posterior wall. LMCA is a short-course artery, 5-20 mm in length. It is located at the posterior part of the pulmonary trunk and is divided into the left anterior descending artery (LAD) and left circumflex arteries (LCx). The LAD lies in the anterior interventricular groove along the ventricular septum and gives the septal perforator branches that feed the anterior 2/3 of the anterior ventricular septum(Tomanek 2005).

The LCx lies in the posterior atrioventricular groove on the opposite side and has a course similar to the RCA. The diameter and length of this artery vary. The caliber of the arteries at their origins may vary between 1.5 and 5.5 mm according to the type of measurements (i.e., arterial casts or angiograms) (Badshah, Qadir, et al. 2015, Baz, Refi, et al. 2024). Sometimes, there may be another third branch of the LMCA called ramus intermedius. The presence of ramus intermedius, the most common variation of the LMCA, occurs in approximately 1/3 of the population, and this pattern is called the trifurcation of LMCA. Less than 1% of the population does not have an LMCA (Mahalingam, Gawandalkar et al. 2016). The arterial supply of the rest of the LV is provided by the dominant artery (Wu, Kheiwa et al. 2024).

Case Description:

A 53-year-old non-smoker male, with no family history of CAD and no chronic disease, was unexpectedly admitted to the hospital due to atypical chest discomfort and pain. His regular peripheral pulses and electrocardiogram led to a coronary angiogram for CAD evaluation. The angiography revealed an abnormal coronary artery pattern, with the patient having only LMCA, originating from the left ostium. The LMCA, divided into LAD and LCX arteries, was unusually short (Figure 2). The LAD, after giving a large obtuse marginal artery branch, also gave a smaller one and continued in the course of the right coronary artery towards the right side of the heart. The angiography further revealed that the distal part of the left circumflex artery was maintained in accordance with the course of the right coronary artery.It was found that the patient had an anatomically different RCA, which originates from the distal circumflex artery.

Discussion:

Coronary artery disease (CAD) is the most common cause of cardiovascular deaths (Brown, Gerhardt et al. 2024). Coronary artery anomaly is associated with abnormal regression or persistence of primitive coronary arteries in the embryological period. It is seen in 1-2% of

the general population(Kanagala, Gupta et al. 2023).

Coronary arteries are formed before the development of arterial orifice in aortic sinuses. Primitive vessels developing from the subepicardial space invade the myocardium and spread to the myocardial surface. These sprouted vessels are then connected with the aortic root. Initially, six coronary arteries develop, three of which originate from the aorta and three from the pulmonary artery. Except for two coronary arteries that generally arise from the aorta, they disappear by regression. The persistent arteries are called the right and left coronary arteries. Small accessory branches of the coronary arteries may exist in one-third of humans(Tomanek 2005).

The coronary arteries, responsible for the arterial supply of the heart, are the first branch of the aorta ascendens. They are located in the subepicardial connective tissue and on the surface of the heart. Coronary arteries are not the end arteries, but functionally, they behave like end arteries. They are vasa vasorum of the ascending aorta(Fioranelli, Gonnella et al. 2009). In regular anatomy, the right and left coronary arteries (RCA and LCA, respectively) originate from the right and left sinus Valsalva of the aortic root. RCA originates lower than the left main coronary artery (LMCA) origin. The right sinus is located anterior, and the left sinus is located posteriorly on the aortic root. RCA supplies blood to the right atrium, right ventricle, and often the left ventricular posterior wall. LMCA is a short-course artery, 5-20 mm in length. It is located at the posterior part of the pulmonary trunk and is divided into the left anterior descending artery (LAD) and left circumflex arteries (LCx). The LAD lies in the anterior interventricular groove along the ventricular septum and gives the septal perforator branches that feed the anterior 2/3 of the anterior ventricular septum(Ogobuiro, Wehrle et al. 2024).

The LCx lies in the posterior atrioventricular groove on the opposite side and has a course similar to the RCA. The diameter and length of this artery vary. The caliber of the arteries at their origins may vary between 1.5 and 5.5 mm according to the type of measurements (i.e., arterial casts or angiograms) (Badshah, Qadir, et al. 2015, Baz, Refi, et al. 2024). Sometimes, there may be another third branch of the LMCA called ramus intermedius. The presence of ramus intermedius, the most common variation of the LMCA, occurs in approximately 1/3 of the population, and this pattern is called the trifurcation of LMCA. Less than 1% of the population does not have an LMCA (Mahalingam, Gawandalkar et al. 2016). The arterial supply of the rest of the LV is provided by the dominant artery (Wu, Kheiwa et al. 2024)

Figures:

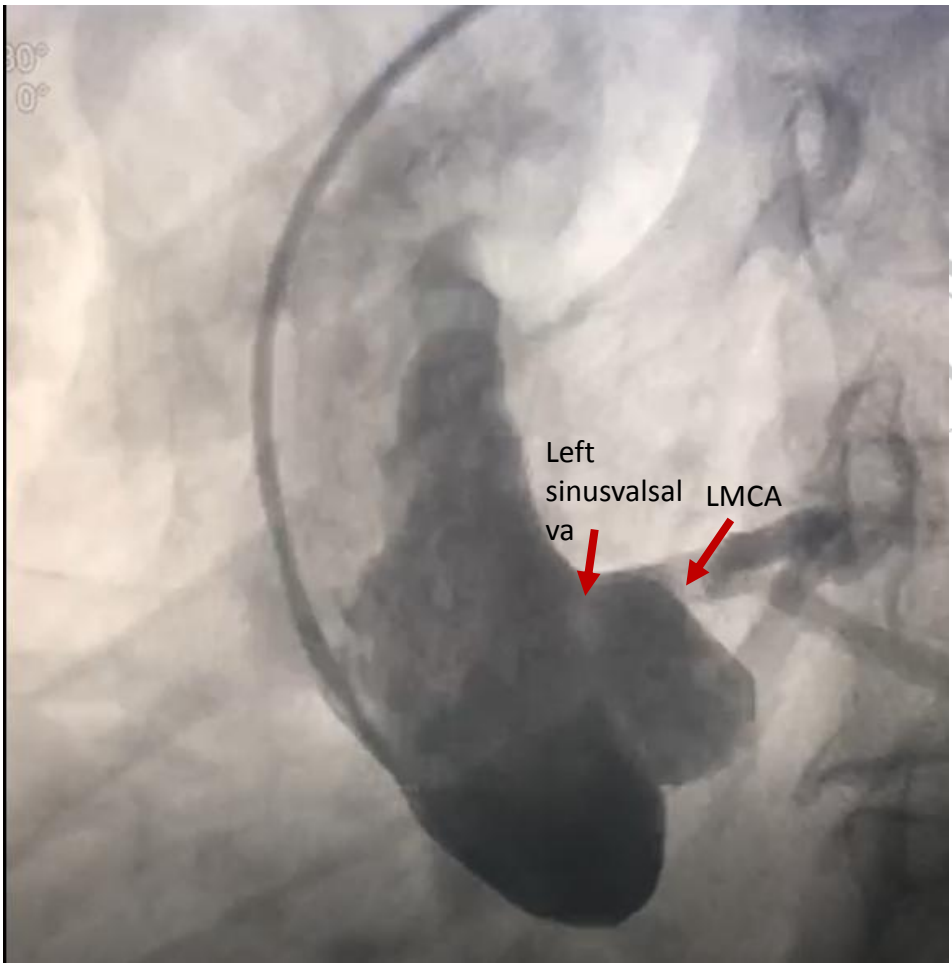

Figure 1. Aortic root angio revealing LMCA arising from left sinus of Valsalva and there is no right coronary ostium. LMCA: Left main coronary artery

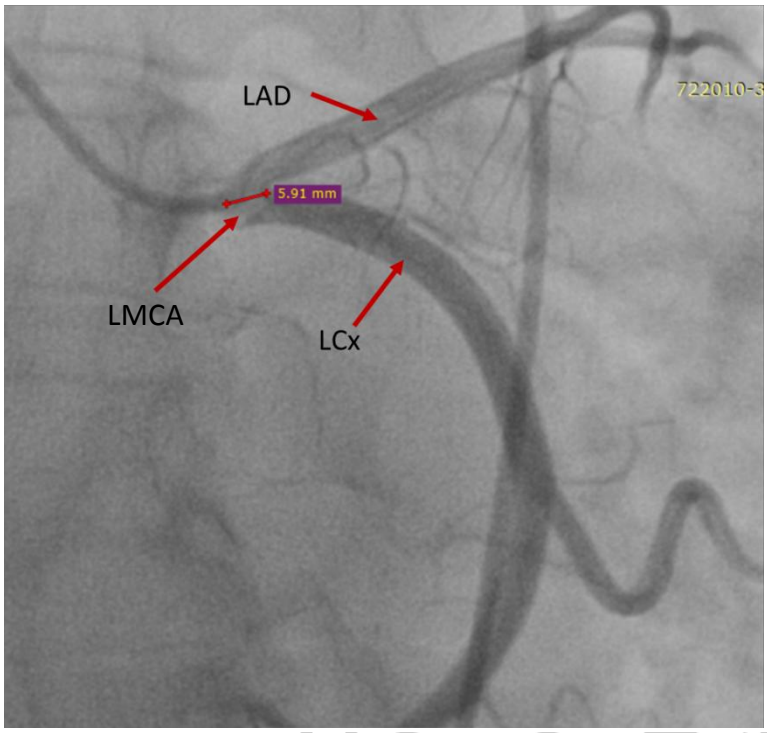

Figure 2. Coronary angiography image shows LMCA and its branches, LAD and LCx. LMCA: Left main coronary artery, LAD: Left anterior descending artery, LCx: Left circumflex artery

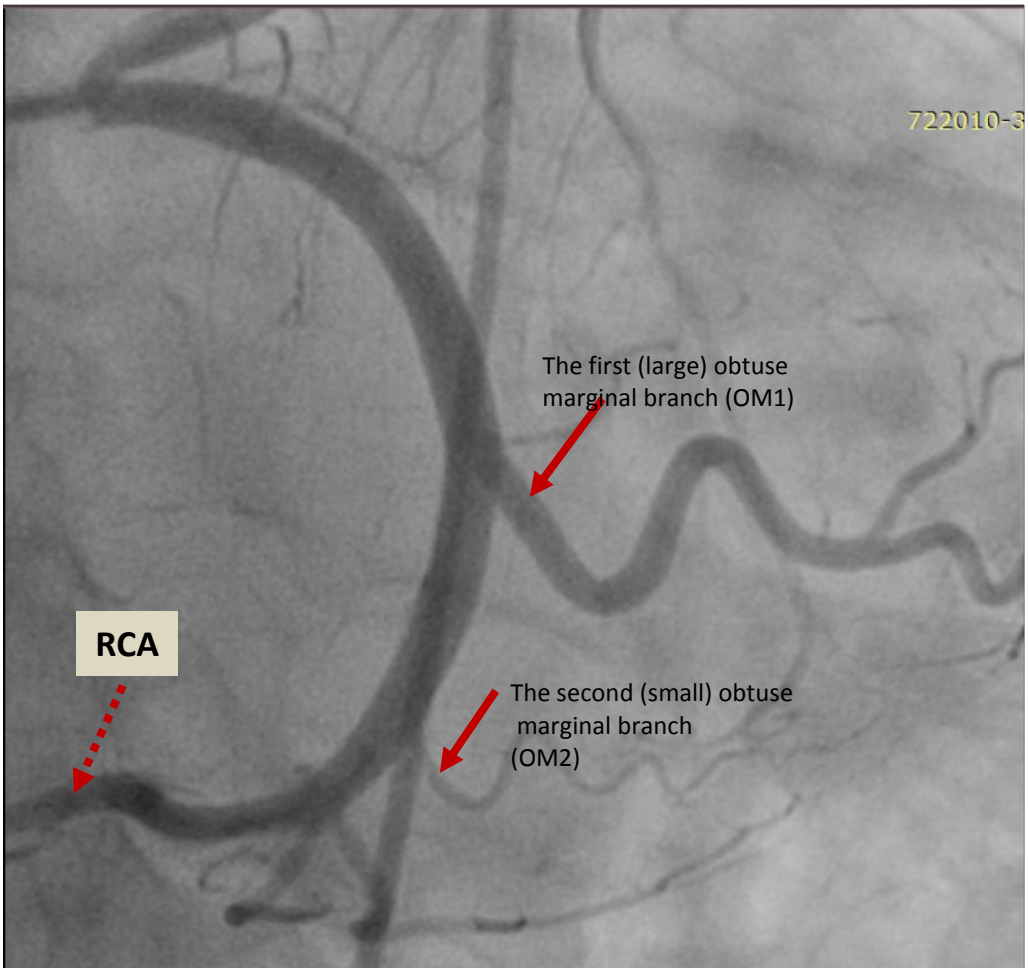

Figure 3. Coronary angiography image shows LCx and its branches which are OM1 and OM2. Dotted line indicates the distal part of the LCx was maintained in accordance with the course of the RCA.

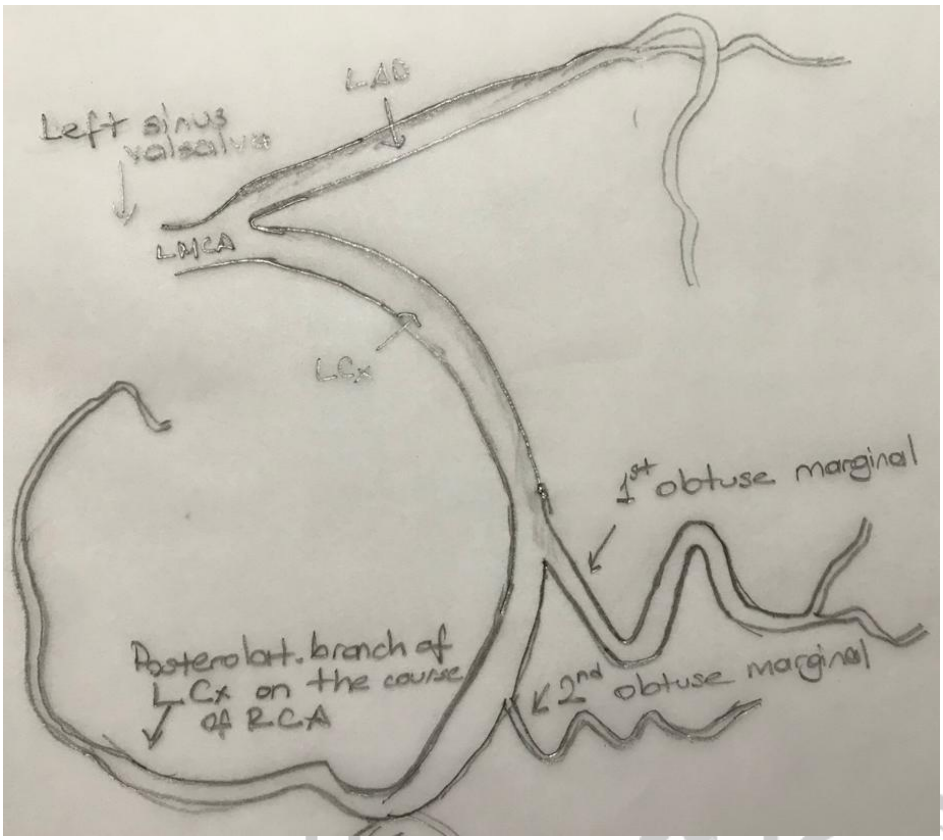

Figure 4. Drawing shows single coronary artery and its branches, LAD, LCx OM1, OM2 and RCA which is the distal part of the LCx.

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