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Case Report

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# Case Report: Understanding Malignant Coronary Artery Anomalies

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## ABSTRACT

**Background:** The right coronary artery (RCA) plays a crucial role in providing innervation to the right ventricle and right atrium, including key nodes involved in heart rhythm regulation. Anomalous origin of the RCA, particularly from the left coronary sinus, termed malignant RCA, is a rare congenital condition. Similarly, abnormalities involving the left circumflex artery (LCX), which supplies blood to the lateral and posterior walls of the left ventricle, can also pose significant clinical risks, particularly in cases where the LCX is anomalously connected or obstructed. Given the rarity of these conditions, we report three cases of hospitalized adults with malignant courses of their coronary arteries (two RCAs and one LCX).

**Case report**

**Case 1:** A 53-year-old male with diabetes presented with severe chest pain persisting for 3 days. Diagnostic tests revealed ST segment elevation in inferior leads on electrocardiogram (ECG) and elevated troponin T levels. Coronary angiography revealed an anomalous RCA origin, along with significant occlusion and stenosis in other coronary arteries, indicating a malignant course. The patient underwent coronary revascularization and was scheduled for coronary artery bypass grafting (CABG).

**Case 2:** A 69-year-old female with multiple comorbidities, including diabetes and hypertension, presented with continuous chest pain. ECG indicated left bundle branch block (LBBB), and troponin T levels were elevated. Imaging confirmed a malignant course of the RCA with calcification atheromatous plaque. Despite diagnosis, the patient declined intervention.

**Case 3:** A 76-year-old male presented with chest pain following a road traffic accident where he sustained impact to his chest. Initial tests showed an abnormal ECG and elevated troponin, prompting further investigation. Imaging revealed a sternal fracture and pericardial fluid but no major cardiac events. A coronary CT scan uncovered several significant coronary artery abnormalities, including a calcified obtuse marginal artery, a hypoplastic aberrant left anterior descending artery, and an abnormal left circumflex artery.

**Conclusion:** These cases highlight the clinical challenges posed by anomalous coronary arteries and underscore the importance of timely diagnosis and management. Treatment strategies should be tailored considering individual patient factors and preferences, with a focus on optimizing outcomes and improving quality of life.

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## Introduction

Coronary artery anomalies (CAAs) are a group of congenital conditions characterized by abnormal origin or formation of any of the three main epicardial coronary arteries, including the right coronary artery (RCA), left anterior descending (LAD), and left circumflex coronary artery (LCX). Both LAD and LCX originate from the common tract (left main coronary artery) [1].

CAAs were first recognized in the 18th century, but the first scientific data was published in 1969 and revised in the year 2000 [2,3]. The prevalence of CAAs is estimated to be about 7.9% (as reported in a cohort study done on 1759 patients). Another study estimates the prevalence of CAAs to be 1.3% among patients undergoing coronary angiography [4]. Approximately 80% of anomalies are benign; the remaining 20% can cause symptoms and may be associated with significant disease [5]. Most of the CAAs are sporadic findings but could also be associated with other cardiac pathologies or congenital heart diseases (Tetralogy of Fallot, Transposition of the Great Vessels).

An anomalous right coronary artery is a rare entity, accounting for 0.92% of cardiac patients, resulting from dysfunction during embryonic development. This anomaly was first described in 1948 by White and Edwards. Malignant RCA is defined as mechanical compression of the RCA between the aorta and the pulmonary artery, which then follows its normal destination. This abnormality is rare, accounting for 0.1% of coronary angiographies [6-8].

Anomalous origin of the Left Circumflex Artery (LCX) is even rarer than anomalous RCAs. The LCX typically branches from the left main coronary artery and supplies blood to the lateral and

posterior walls of the left ventricle. Anomalous LCX, where the artery arises from an abnormal origin such as the right coronary sinus or right coronary artery, is a rare congenital condition.

Anomalous coronary arteries are often asymptomatic, discovered incidentally during Computed Tomography Coronary Angiogram (CTCA) or during Percutaneous Coronary Intervention (PCI), but can present with exertional chest pain, syncope, arrhythmia, heart failure, and sudden cardiac death. Major risk factors for developing symptoms include a history of myocardial infarction, arrhythmia, and an inter-arterial course (malignant course). Management techniques usually differ according to the subtype of the anomaly; however, surgical management is widely used in those patients with an increased risk of Sudden Cardiac Death (SCD).

## Case Presentation

### Case 1

A 53-year-old male presented to the emergency department with severe chest pain and associated tightness, accompanied by sweating, which commenced three days prior to admission. The patient had initially disregarded the symptoms; however, the recurrence of similar pain earlier on the day of admission prompted a visit to another facility.

An Electrocardiogram (ECG) at the clinic showed ST-segment elevation in the inferior leads. Following the administration of aspirin and clopidogrel, he was advised to seek further evaluation at our hospital. Clinical findings upon evaluation included mild pain rated at 1-2/10, a heart rate of 56–60 bpm, and hypotension with a blood pressure of 96/70 mmHg. Arterial blood gas analysis indicated elevated lactate, hyperglycemia, significantly reduced oxygen saturation, and hypercapnia. Laboratory tests revealed elevated white blood cells, neutrophils, monocytes, and a significantly increased troponin T level at 940 ng/L (normal: <14 ng/L).

Additional abnormalities included hypochloremia,hyponatremia, an elevated Creatine Kinase-Muscle Brain (CK-MB) level of 88.7 U/L (normal: <6.23 U/L), and an HbA1c of 8.5% (normal: <5.7%). The initial diagnosis was ST-Segment Elevation Myocardial Infarction (STEMI), implicating the right coronary artery. A coronary angiogram illustrated severe diffuse stenosis of the Left Anterior Descending Artery (LAD) starting from the proximal segment and an occluded Mid-Right Coronary Artery (RCA), which was noted to be anomalous, originating from the Left Coronary Cusp (LCC). The patient underwent successful Primary Coronary Intervention (PCI) with the placement of one Drug-Eluting Stent (DES) in the RCA without complications. Subsequent outpatient follow-up and a CT coronary angiogram revealed a malignant course of the RCA.

Although Coronary Artery Bypass Grafting (CABG) was recommended to revascularize the LAD and to perform unroofing of the anomalous RCA, the patient opted against the surgery, preferring to seek further treatment in his home country. Discharge advice and follow-up plans were meticulously outlined, emphasizing a low-fat, low-salt, and diabetic-friendly diet, adherence to prescribed medications, and strict avoidance of medication discontinuation without cardiological consultation. The patient was instructed to immediately call emergency services in case of symptom recurrence and to adhere to driving restrictions for one week. Recommended laboratory follow-ups included a complete blood count, creatinine, electrolytes, HbA1c, and lipid profile after one month.

### Case 2

A 69-year-old female was admitted to the Emergency Department (ED) due to central chest pain that began two hours before her presentation. This episode occurred while she was visiting a relative in the hospital. The patient's medical history included diabetes mellitus, hypertension, and the presence of varicose veins. A noteworthy finding on admission was a new Left Bundle Branch Block (LBBB) on the Electrocardiogram (ECG), which had not been documented previously. Upon evaluation in the ED, the patient's chest pain, initially rated at 4/10, subsided to 0 following aspirin administration.

This visit marked her second presentation to the ED in three weeks with similar symptoms. During her first visit, she was discharged for outpatient cardiology follow-up after tests showed negative troponin levels and a non-dynamic ECG. However, she was admitted again two days prior to the current admission with a diagnosis of ST-Segment Elevation Myocardial Infarction (STEMI), but she declined coronary angiography. The patient reported experiencing these symptoms for the past four days, along with difficulty breathing, primarily at night, not exacerbated by exercise or walking. Sitting upright alleviated her pain and shortness of breath.

A diagnosis of Non-ST-Segment Elevation Myocardial Infarction (NSTEMI) was made, with active medical issues including newly diagnosed LBBB, hypertension, hypothyroidism, type 2 diabetes mellitus with complications, and dyslipidemia, targeting a Low-Density Lipoprotein (LDL) goal of below 100 mg/dL. The patient also reported respiratory symptoms such as choking and chest tightness. Further examination revealed a sinus rhythm with left axis deviation and the new onset of LBBB compared to an ECG performed three weeks earlier. The patient's obesity was noted as a significant physical characteristic.

Laboratory findings included an elevated D-dimer level of 1.65 μg/mL (normal: <0.5 μg/mL), hyperglycemia with a random glucose level of 325 mg/dL, and a slight elevation in troponin T at 15 ng/L. Electrolyte imbalances included mildly low sodium and chloride levels, with an elevated blood urea nitrogen level of 46 mg/dL. Arterial blood gas analysis showed hypercapnia, hypoxemia, and compensated respiratory acidosis with elevated bicarbonate levels. The oxygen saturation was notably low, and the HbA1c level was elevated at 8.0%. An echocardiogram revealed left ventricular concentric remodeling with a relative wall thickness of 48% and normal systolic function. Despite initial reluctance, the patient was admitted and followed the STEMI pathway, including a planned primary Percutaneous Coronary Intervention (PCI). However, she ultimately refused PCI, necessitating a CTCA.

The CTCA uncovered a malignant course of the Right Coronary Artery (RCA) between the aorta and the right ventricle, along with small calcific atheromatous plaques in the proximal RCA, Left Anterior Descending (LAD) artery, and Mid-Left Circumflex (LCX) artery without significant stenosis. A clinical follow-up was planned for the patient.

### Case 3

A 76-year-old male presented to the ED with musculoskeletal chest pain post Road Traffic Accident (RTA) in which he sustained direct impact to his chest from the steering wheel. He reported mild chest pain with superficial contusions. His medical history includes dyslipidemia and prediabetes. Upon further examination, he was found to have an abnormal ECG and elevated troponin level of

15. Chest X-Ray was performed and it showed no pneumothorax, pleural effusion, or any obvious rib fractures. However, it was suggestive of fracture in the upper sternum. Serial ECG indicated ischemic changes with negative serial troponins and no chest pain or palpitations. An echocardiogram demonstrated a thin rim of pericardial fluid around the right atrium, which showed no progression upon repetition.

Given the patient's background of dyslipidemia and prediabetes, and the need for further evaluation, a CT angiography was ordered to assess for coronary artery disease. The CT angiogram resuts were normal, promting the decision to perform a coronary CT scan to check for any deformities or abnormalities within the vessels. The coronary CT revealed several significant findings:

- A calcified vessel extending from the distal third of the right coronary artery along the inferior atrioventricular groove and arching towards the left ventricle, consistent with an obtuse marginal artery.
- The left main coronary artery was not visualized.
- A small-caliber vessel arising from the right main coronary artery, coursing anterior to the coronary cusp into the interventricular groove, with speckled calcification in the distal third, suggesting a hypoplastic aberrant left anterior descending artery.
- A small-caliber vessel originating from the right coronary cusp, arching superiorly around the pulmonary artery and extending into the interventricular groove, with calcification observed at this level, likely representing the left circumflex artery.

## Discussion

Coronary arteries cover the heart from the outside, and some small branches dive inside the heart. Those arteries deliver oxygen and nutrients to the heart muscle so it can work efficiently. Coronary arteries are divided into two main arteries: the left main coronary artery (which supplies the left atrium and left ventricle) and the right coronary artery (which supplies the right atrium and right ventricle). The left main coronary artery is divided into two branches: the left anterior descending artery (which supplies blood to the front and left sides of the heart), and the left circumflex artery, which travels through the atrioventricular groove and then gives the obtuse marginal branches (which supply the lateral and posterolateral walls of the left ventricle) [9,10].

The right coronary artery is divided into smaller branches: the right posterior descending artery (which supplies blood to the inferior aspect of the heart), the right marginal artery (which supplies the lateral portion of the right ventricle), the sinoatrial nodal artery (which supplies the sinoatrial node), and the atrioventricular nodal artery (which supplies the AV node). However, in 90% of people, the AV node is supplied via the septal perforating branch [11]. Anomalies in the right coronary artery will have different pathways after they originate from the left coronary sinus. The most common presentation is the interarterial, which is further classified into high interarterial course or malignant RCA (RCA passes through the aorta and pulmonary artery) and low interarterial course (RCA passes through the aorta and right ventricular outflow tract). The high interarterial course is seen in the first two patients. It was first described in 1982 in medical literature by James E. Cheitlin, and is associated with major adverse cardiac events (MACEs) [12,13].

In contrast, the Left Circumflex Artery (LCX) plays a vital role in supplying blood to the lateral and posterior regions of the left ventricle. Anomalies or significant issues with the LCX, such as those observed in the third patient, can have serious implications for cardiac function. The LCX is essential as it supplies a substantial portion of the myocardium, and any occlusion or structural anomaly can result in localized ischemia, arrhythmias, or even infarction [14].

MACEs, related to RCA or LCX, can occur due to one or more of the three suggested theories: The first theory is ostial stenosis; some studies show an association with sudden cardiac death [15]. The second theory is mechanical compression that happens during exertion, and the third theory is vasospasm of the anomalous RCA [16]. In 1992, Taylor et al. examined the heart specimens of 242 autopsies that had isolated coronary anomalies; 20% had anomalous RCA originating from left coronary cusps, and 25% of the ARCA had died suddenly [17]. Therefore, management guidelines and options were established, which include: (1) Surgical Intervention: various surgical techniques are available to address this condition. Common approaches include unroofing the intramural segment of the artery, which is often preferred, and reimplantation of the anomalous artery into the appropriate aortic sinus. These techniques aim to relieve the compression that the RCA might be subjected to during physical exertion [18]. (2) Non-Surgical Management: In some cases, particularly where surgery is not deemed appropriate or is too risky, medical management might be considered. This can include the use of medications such as beta-blockers to manage the symptoms and reduce the risk factors associated with sudden cardiac events. Lifestyle modifications, including avoiding strenuous or competitive sports, may also be recommended to reduce the risk of cardiac events [17]. (3) Follow-up and monitoring: Regular follow-up using imaging techniques like CT angiography or MRI is crucial for monitoring the condition and assessing the effectiveness of the treatment approach. This is especially important for asymptomatic individuals who are at risk but may not display overt symptoms [19].

## Conclusion

Managing anomalous RCA or LCX, particularly with a risky interarterial path, is challenging due to its rarity and potential for severe outcomes like sudden cardiac death. Advanced imaging guides treatment, from conservative to surgical approaches.

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