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Case Report

An Anomalous Right Coronary Artery Draining into the Left Ventricle Through a Fistula

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Abstract

Background: Of the three main epicardial coronary arteries, the right coronary artery, left anterior descending artery, and the left circumflex artery, there are many variables between individuals regarding arterial course, distribution of side branches, and termination points. The term 'Coronary Artery Anomaly' (CAA) is reserved for congenital alterations in origin or course of the epicardial arteries that occur in less than 1% of the population. While the majority are asymptomatic, such anomalies can significantly increase the risk of myocardial ischemia and sudden cardiac death. With the increasing use of Coronary Computed Tomography Angiography (CCTA), the rate of detection of anomalous coronary arteries is increasing every year. The concurrent presence of a coronary cameral fistula further escalates clinical complexity requiring a multidisciplinary approach to management.

Case Report: A 61-year-old female with a past medical history of hypertension, hypothyroidism, migraine, Non-Sustained Ventricular Tachycardia (NSVT), and Supraventricular Tachycardia (SVT), presented to the emergency department with chest pain and palpitations. Coronary angiography was performed revealing a large Right Coronary Artery (RCA) with a proximal branch giving rise to the Left Circumflex Artery (LCX), with an additional branch supplying the Left Anterior Descending (LAD) territory. The RCA and LCX arteries terminated distally in the left ventricular cavity forming a coronary cameral fistula. No flow-limiting stenosis was observed. Intervention versus medical management was discussed with a multidisciplinary team, and the decision was made to proceed with medical management. The patient's symptoms improved with the medical management.

Conclusion: A thorough diagnostic evaluation and a multidisciplinary approach in managing rare coronary anomalies are critical. Medical management, risk factor modification, and regular follow-up are essential components of long-term care for patients with complex coronary artery anomalies.

Keywords: Coronary Artery Fistula; Case Report; Chest Pain

Introduction

Coronary Artery Anomalies (CAAs) are uncommon congenital abnormalities identified in approximately 1% of individuals undergoing coronary angiography [1,2]. These anomalies can manifest across a broad spectrum of clinical presentations, ranging from asymptomatic findings to severe complications, including myocardial ischemia and sudden cardiac death [2]. It is particularly rare to find an anomalous origin of a coronary artery. These cases can be associated with significant morbidity and mortality due to the potential for compromised myocardial blood flow [3]. The simultaneous occurrence of an anomalous coronary artery and a coronary artery fistula is even more uncommon. Coronary cameral fistulas are abnormal connections between a coronary artery and a cardiac chamber. They are extremely rare, detected in approximately 0.2% of the general population undergoing coronary angiography. The most common form of coronary cameral fistula occurs with the RCA

terminating in the right ventricle. Left atrial or left ventricular involvement is exceedingly rare [3,4]. The combination of these two anomalies can complicate the clinical presentation and management strategy. In this case report, we present a 61-year-old female with a complex cardiac anomaly involving an anomalous LCx arising from the RCA, giving off a branch supplying LAD territory and terminating into the LV [5,6].

Ethical Statement

The project did not meet the definition of human subject research under the purview of the IRB according to federal regulations and therefore was exempt.

Case Presentation

A 61-year-old female with a past medical history of hypertension, hypothyroidism, migraine, Non-Sustained Ventricular Tachycardia (NSVT), and Supraventricular Tachycardia (SVT), presented to the hospital with chest pain and palpitations. She described the chest pain as left-sided tightness and squeezing without radiation, non-exertional, non-pleuritic, and non-positional. Symptoms self-resolved while she was in the emergency department.

Her symptoms had been ongoing for approximately one year, initially occurring once or twice a week, then increasing in frequency recently over the past three to four weeks to several times a day. She was evaluated by an outpatient cardiologist for her recurrent palpitations and chest pain and an echocardiogram, and Holter monitor were performed. CCTA was ordered, however, she had yet to complete it at the time of her presentation to the emergency department. While the echocardiogram results were unremarkable, the Holter monitor revealed some runs of SVT, likely atrial tachycardia, lasting less than two minutes. She denied any family history of congenital heart disease, sudden cardiac death, or premature coronary artery disease. Her EKG showed sinus rhythm without ST elevation or depression.

A chest X-ray was unremarkable. High-sensitivity troponin assays and Brain Natriuretic Peptide (BNP) levels were within normal limits. Due to the quality and the refractory nature of chest pain despite increasing intrahospital antianginal therapy cardiac catheterization was performed revealing a large Right Coronary Artery (RCA) with anomalous origin of Left Circumflex Artery (LCX) arising from a proximal branch of the RCA. The LCX gave off an additional branch from which the Left Anterior Descending artery (LAD) arose. The RCA and LCX arteries terminated distally in the left ventricular cavity forming a coronary cameral fistula (Fig. 1).

Notably, there were no significant stenosis or plaques observed in any of the coronary arteries. The patient's Left Ventricular Ejection Fraction (LVEF) was within the normal range of 60-65%, with mildly elevated Left Ventricular End-Diastolic Pressure (LVEDP) of 17 mmHg. Further characterization was performed with Coronary Computed Tomography Angiography (CCTA) redemonstrating the anomalous origin of the left-sided coronary arteries stemming from a large RCA with an anterior, pre-pulmonic course.

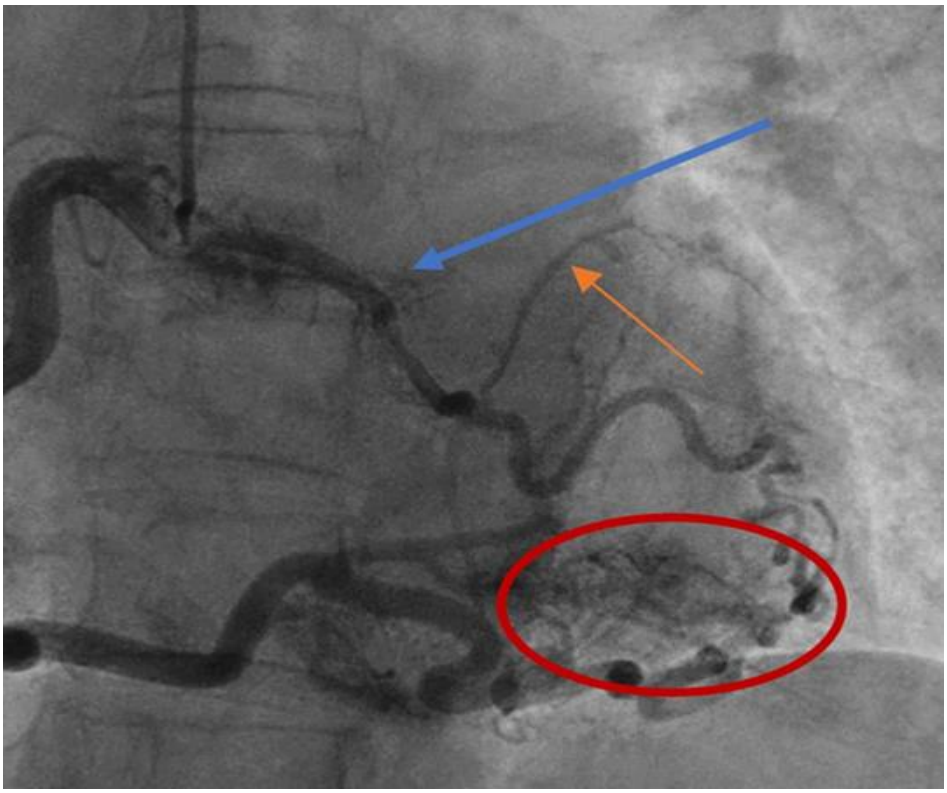

Figure 1: Coronary angiography: Left circumflex artery (blue arrow) arising from proximal branch of the RCA. Small branch of LCx (orange arrow) supplying LAD territory. Fistulous connection of distal RCA and LCx into LV (red circle).

Treatment

Given the complexity of the coronary artery anomaly and the absence of significant ischemia, a multidisciplinary team comprising of interventional cardiology, adult congenital cardiology, and cardiothoracic surgery recommended against surgical intervention. It was determined that coronary artery bypass grafting of the left-sided system would likely result in competitive flow without substantial benefit. Moreover, a distal insertion into LV cavity was not well visualized on CCTA, ECHO or MRI. A multi-slice CT did show anomalous insertion into LV cavity, however no single image was able to be obtained demonstrating the insertion. Given high spatial resolution no single slice was possible to capture the RCA appropriately. Consequently, the patient was managed medically on a regimen of aspirin, beta-blocker, and a calcium channel blocker. Her palpitations and chest pain resolved, and continues to do well upon follow up visits. She was counseled on cardiac risk factor modification to mitigate the risk of future cardiovascular complications. Follow up was at 6 months, and the patient was chest pain free at that time on anti-anginal medical therapy.

Discussion

This case highlights a rare and complex cardiac anomaly involving an anomalous origin of the LCx coronary artery from a large RCA and a coronary artery fistula between distal RCA and LCX into the Left Ventricle (LV). This specific combination of anomalies is exceedingly uncommon, with no documented cases in literature yet [7,8]. The clinical presentation of patients with coronary artery origin and insertion anomalies can vary widely, depending on the presence of a fistula, patient age, activity level, baseline cardiac health, and specific coronary blood flow insertion point. The presenting symptoms may range from asymptomatic to severe symptoms such as angina, heart failure, or sudden cardiac death [9-14]. Coronary cameral fistulas are particularly risky. Coronary cameral fistulas are abnormal connections between a coronary artery and a cardiac chamber. In the case of coronary cameral fistulae, myocardial ischemia, and thus chest pain, may result as the presence of a fistula may lead to a steal phenomenon, in which blood is diverted away from the myocardial tissue [15]. Anomalous coronary arteries and fistulas can predispose patients to arrhythmias due to abnormal electrical pathways and myocardial ischemia [2]. Given such anomalies' complexity and potential severity, several recommendations can be made for future clinicians. Early and precise identification is crucial for appropriate management, especially if the patient is a reproductive-age woman desiring a pregnancy. Advanced imaging techniques such as Coronary Computed Tomography Angiography (CCTA) and Magnetic Resonance Angiography

(MRA) are often utilized for accurate diagnosis and detailed anatomical assessment of coronary anomalies. Management of such patients should involve a multidisciplinary team including cardiologists and cardiothoracic surgeons to formulate an optimal treatment strategy. Treatment should be tailored to the patient's specific anatomy and clinical presentation. Options may include surgical correction, percutaneous intervention, or conservative management with regular follow-up to monitor for the development of complications such as myocardial ischemia, arrhythmias, or heart failure [9]. In this particular case of a complex coronary artery anomaly, another reason for medical over surgical initial management suggested coronary artery bypass grafting of the left-sided system would likely result in competitive flow without substantial benefit. It is essential to educate patients about their condition, potential symptoms, and when to seek medical attention. Additionally, genetic counseling may be recommended for families with a history of congenital heart defects.

Conclusion

This case report discusses a unique presentation of an anomalous coronary artery and concurrent coronary cameral fistula. It highlights the importance of thorough diagnostic evaluation and a multidisciplinary approach in managing rare coronary anomalies. Medical management, risk factor modification, and regular follow-up are essential components of long-term care for patients with complex coronary artery anomalies. Further studies are needed to establish standardized guidelines for the management of such rare conditions. This case highlights the dilemma patients and caregivers face with difficult management decisions regarding invasive versus medical or watchful waiting.

Conflict of Interests

The authors have no competing interests that are relevant to the content of this article to declare.

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Statements and Declarations

All authors had access to the data and a role in writing the manuscript, with no disclaimers and patient consent was obtained.

Financial Disclosure

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Data Availability

Data Availability Statement: All data generated or analyzed in this study are included in this article. Access to data is possible with permission from the responsible author.

Consent for Publication

Informed consent was obtained from the patient for publication of this case report and is stated in the manuscript.

Authors Contribution

All authors contributed equally for this paper.

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