Case Report

Congenital absence of the right coronary artery, with the LCX supplying the entire RCA territory

Hossein Nough [1], *Mohammadtaghi Sarebanhassanabadi* [1], *Mahmoud Emami* [1], *Parisa Peigan* [1,*], *Fatemeh Rahmanidehnavi* [2]

[1] Yazd Cardiovascular Research Center, Non-communicable Diseases Research Institute, Shahid Sadoughi University of Medical Sciences, Yazd, Iran
[2] Department of Cardiology, Clinical Research Center, Paster Educational Hospital, Bam University of Medical Sciences, Bam, Iran

*** Corresponding Author:**
Address: Yazd, Afshar Hospital, Jomhouri Blvd, Yazd, Iran. **Postal code:** 8917945556; **Email:** parisapaygan@gmail.com

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Abstract

Objectives: Congenital absence of the right coronary artery (RCA) is an exceedingly rare coronary anomaly in which the left coronary system supplies the entire myocardium. We report a case of congenital absence of the RCA with a superdominant left circumflex artery (LCX) supplying the entire RCA territory in a patient presenting with stable angina, highlighting the diagnostic and clinical importance of recognizing this unusual coronary anatomy.

Methods: A 57-year-old man with hypertension and type 2 diabetes mellitus presented with a one-month history of typical anginal chest pain. Clinical evaluation included electrocardiography, transthoracic echocardiography, single-photon emission computed tomography myocardial perfusion imaging (SPECT MPI), and coronary angiography to assess myocardial ischemia and coronary anatomy.

Results: SPECT MPI demonstrated a small-to-moderate reversible perfusion defect in the apical region. Coronary angiography revealed no identifiable RCA ostium in the right coronary sinus, confirming congenital absence of the RCA. Selective left coronary angiography demonstrated a markedly enlarged, superdominant LCX that coursed through the left atrioventricular groove, crossed the crux, and extended into the right atrioventricular groove, supplying the territories normally perfused by the RCA. A myocardial bridge was also observed in the mid-left anterior descending artery without significant compression at rest, and no significant obstructive coronary artery disease was identified. Medical treatment with bisoprolol was initiated, and the patient remained asymptomatic during follow-up.

Conclusions: Congenital absence of the RCA with compensatory perfusion by a superdominant LCX is a rare coronary anomaly that may present with angina and evidence of myocardial ischemia. Accurate delineation of coronary anatomy is essential to establish the diagnosis, avoid unnecessary attempts to cannulate a nonexistent RCA ostium, and guide future interventional or surgical management.

Keywords: Congenital Absence of Right Coronary Artery, Single Coronary Artery, Coronary Artery Anomaly

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Introduction

Anamoly1

Congenital absence of the right coronary artery (RCA) is a rare coronary anomaly, with a reported incidence of approximately 0.024%–0.066% in angiographic studies (1-3). It represents a distinct and uncommon variant of single coronary artery anomalies, in which the left coronary system supplies the entire myocardium (4). Absence of LCX, in which the RCA supplies the LCX territory, is another type of single coronary artery (17).

Anamoly 2

Although often considered benign and asymptomatic, this anomaly may have important clinical implications (1, 4). In some cases, it has been associated with serious complications, including acute myocardial infarction, stroke, and sudden cardiac death (5). Here, we report a case of congenital absence of the RCA, in which the left circumflex artery (LCX) supplied the entire RCA territory.

Case Report

The patient was a 57-year-old man who presented to the cardiology clinic with a one-month history of typical anginal chest pain, classified as functional class I–II. His medical history was notable for hypertension of six years' duration and type 2 diabetes mellitus for seven years, both managed with pharmacological therapy. On physical examination, the only remarkable finding was a fourth heart sound (S4) at the apex. A fourth heart sound (S4) was detected at the apex. The patient's resting electrocardiogram (ECG) showed normal sinus rhythm without evidence of acute ischemic changes. Transthoracic echocardiography showed preserved left ventricular systolic function with a normal ejection fraction and grade 1 diastolic dysfunction. To further evaluate for myocardial ischemia, a myocardial perfusion scan using technetium single-photon emission computed tomography (SPECT MPI) was performed, revealing a small-to-moderate reversible perfusion defect in the apical region, consistent with ischemia. Based on the patient's symptoms and stress test findings, coronary angiography was performed. The procedure revealed a myocardial bridge in the mid-segment of the left anterior descending artery (LAD), causing insignificant compression at rest. During catheterization of the aortic root, the catheter was advanced into the right coronary sinus; however, no right coronary artery (RCA) ostium was identified, confirming its congenital absence. Contrast injection into the left coronary artery demonstrated a markedly enlarged, superdominant left circumflex artery (LCX). This vessel coursed through the left atrioventricular groove, crossed the crux, and extended into the right atrioventricular groove. Branches arising from the LCX supplied the entire territory typically perfused by the RCA, including the right ventricular branches and the conus artery. Thus, the LCX provided complete compensatory perfusion in the absent RCA territory Figures 1 to 4.

OUTCOME AND FOLLOW-UP

Medical treatment with bisoprolol 5 mg /day was recommended for patient. In follow up he was asymptomatic.

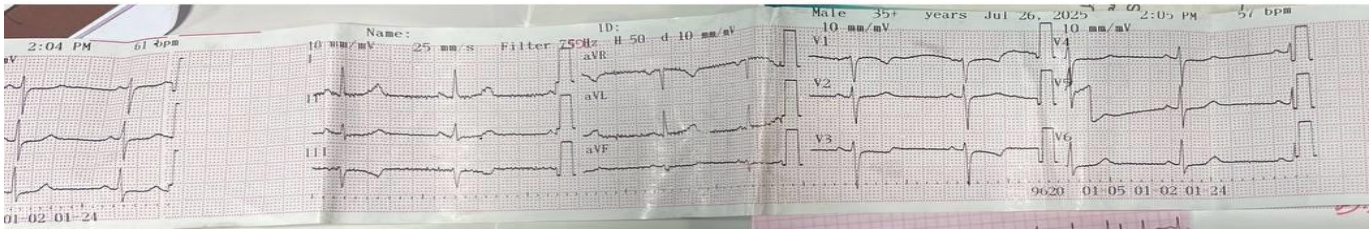

Fig 1. The rhythm was consistent with sinus bradycardia originating from the sinus node at 59 bpm. The PR interval was slightly prolonged at 205 ms, indicating a first-degree atrioventricular (AV) block, which is generally considered benign. The QRS and QTc intervals were within the normal range. The cardiac electrical axis was normal, with no evidence of bundle branch block, chamber enlargement, or acute ischemia. R-wave progression was also normal.

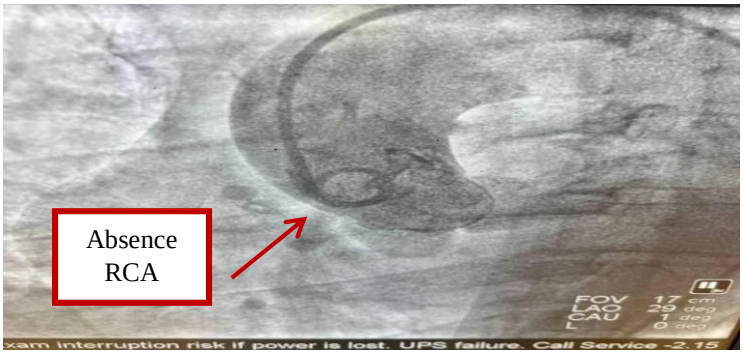

Fig 2. Coronary angiography demonstrated a single coronary artery, characterized by congenital absence of the right coronary artery (RCA). The left main coronary artery bifurcated into a normal left anterior descending (LAD) artery and a superdominant left circumflex (LCX) artery. The superdominant LCX extended distally to supply the entire RCA territory, including the posterior descending and posterior left ventricular branches. The entire coronary system was patent and free of significant atherosclerotic disease.

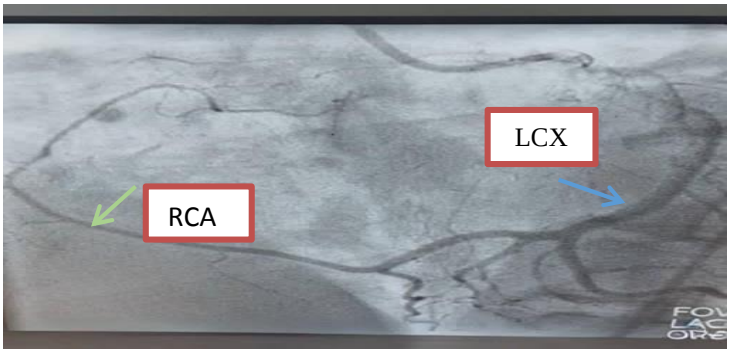

Fig 3. Selective angiography of the left coronary system demonstrated the left main artery, left anterior descending artery, and a superdominant left circumflex (LCX) artery. The superdominant LCX was markedly elongated, coursing through the atrioventricular groove to the crux of the heart. This vessel exclusively perfused the inferior and posterior myocardial territories normally supplied by the right coronary artery. The image, obtained in a left anterior oblique cranial projection (41°/14°), showed patent arteries without significant stenosis.

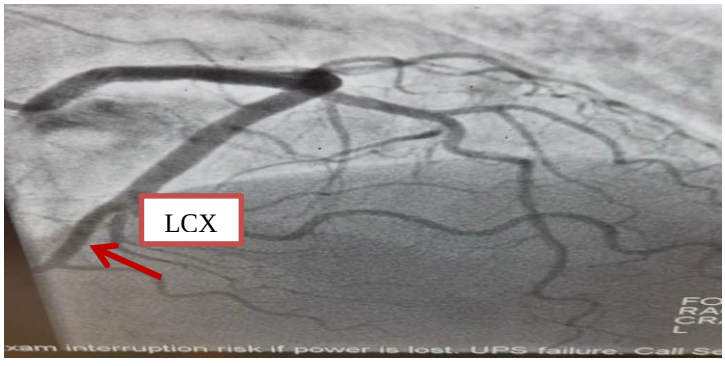

Fig 4. This angiogram depicts the left coronary system originating from the left main artery, which then bifurcates. The image demonstrates the left anterior descending (LAD) artery and a markedly large, dominant left circumflex (LCX) artery. The enlarged LCX reflects a compensatory blood flow pattern, supplying regions normally perfused by the absent right coronary artery. In this right anterior oblique (RAO) cranial view, the visualized arteries appear free of significant stenosis.

Discussion

Congenital absence of the right coronary artery (RCA) is an exceptionally rare coronary anomaly in which the entire myocardium is supplied solely by the left coronary system, most often through a markedly enlarged and superdominant left circumflex artery (LCX) (1). This condition represents a distinct subtype of single coronary artery malformation, with an estimated incidence of 0.024% to 0.066% in angiographic series (3). Under normal coronary anatomy, the RCA supplies the right ventricle, the atrioventricular (AV) node, the sinoatrial (SA) node (in approximately 60–70% of individuals), the posterior descending artery (PDA), and the conus artery (6). In the absence of the RCA, however, the LCX assumes responsibility for perfusing these critical territories, typically extending across the crux of the heart into the right atrioventricular groove, where it gives rise to the PDA and right ventricular branches (7, 8). In the present case, a 57-year-old man with stable angina was diagnosed with congenital absence of the right coronary artery (RCA), fully compensated by a superdominant left circumflex artery (LCX). Coronary angiography demonstrated the absence of a right coronary ostium in the right sinus of Valsalva. Selective injection of the left coronary system revealed a markedly enlarged, tortuous LCX coursing through the left atrioventricular (AV) groove, crossing the crux of the heart, and continuing into the right AV groove to give rise to the posterior descending artery, posterolateral branches, conus artery, and right ventricular branches territories normally supplied by the RCA (4). This anatomical configuration represents a single coronary artery anomaly, classified as type L-III according to the Lipton classification, in which the LCX arises from the left main coronary artery and extends to the entire inferior wall (9). Although many patients with this anomaly remain asymptomatic and are diagnosed incidentally during coronary angiography, some may present with angina, dyspnea, arrhythmias, or even sudden cardiac death, particularly during exertion (10). The potential pathophysiological mechanisms of ischemia in such cases include the susceptibility of the elongated compensatory vessel to spasm, external compression, or hemodynamic compromise due to its extended and often acutely angulated course (11, 12). Moreover, because myocardial perfusion relies entirely on a single arterial system, the development of atherosclerotic disease in the left coronary artery carries an especially high risk of catastrophic ischemic events (13). In our patient, single-photon emission computed tomography (SPECT) myocardial perfusion imaging (MPI) revealed a small-to-moderate reversible perfusion defect in the apical region. While this finding may be attributable to microvascular dysfunction associated with long-standing type 2 diabetes mellitus and hypertension, both well-recognized risk factors for coronary microangiopathy, it may also reflect subtle hemodynamic inefficiency resulting from the anomalous coronary anatomy (5). Moreover, a myocardial bridge was identified in the mid–left anterior descending (LAD) artery; however, it did not produce significant systolic compression at rest, making it an unlikely primary contributor to ischemia (14). The possibility of coronary steal associated with the myocardial bridge cannot be entirely excluded, particularly under stress conditions, although it remains speculative in this case. Accurate recognition of such anomalies is essential for optimal clinical management. First, it prevents unnecessary and potentially harmful repeated angiographic attempts to cannulate a non-existent RCA ostium. Second, it has important implications for revascularization strategies. During percutaneous coronary intervention (PCI), stent placement must be carefully planned to avoid compromising the superdominant LCX, particularly at bifurcation sites. Similarly, in coronary artery bypass grafting (CABG), detailed knowledge of anomalous anatomy is critical to prevent inadvertent LCX ligation and ensure precise graft placement (15). Moreover, non-invasive imaging modalities, particularly coronary computed tomography angiography (CTA), play a pivotal role in initial detection and detailed anatomical characterization of anomalies, therefore facilitating accurate pre-procedural planning (16). Because this anomaly may remain clinically silent for decades, clinicians should maintain a high index of suspicion in patients presenting with unexplained angina, even when conventional cardiovascular risk factors are present.

Conclusion

Congenital absence of the right coronary artery is an exceedingly rare anomaly that may present with angina pectoris. In such patients, precise delineation of coronary anatomy using coronary angiography or computed tomography (CT) angiography is essential for accurate diagnosis and appropriate treatment planning. This case underscores the importance of considering congenital coronary anomalies as a potential cause of myocardial ischemia, even in elderly individuals with conventional atherosclerotic risk factors.

Acknowledgements
Congenital absence of the right coronary artery (RCA) is a rare coronary anomaly, with a reported incidence of approximately 0.024%–0.066% in angiographic studies.

Conflicts of Interest Congenital absence of the right coronary artery, with the LCX supplying the entire RCA territory.

Funding
Congenital Absence of Right Coronary Artery, Single Coronary Artery, Coronary Artery Anomaly.

Ethical statements
Ethics with code of: IR.SSU.MEDICINE.1405.0

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