A CASE OF RIGHT CORONARY ARTERY ATRESIA WITH DOMINANT LEFT CIRCUMFLEX CORONARY ARTERY – SINGLE LEFT CORONARY ARTERY OR CORONARY OSTIAL ATRESIA?

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DOI: https://doi.org/10.32896/cvns.v8n1.11-18
Received: 02.02.2026
Revised: 24.03.2026
Accepted: 29.03.2026
Published: 31.03.2026

ABSTRACT

Coronary artery anomalies are uncommon and are frequently detected incidentally during coronary angiography or computed tomography coronary angiography (CTCA). Coronary ostial atresia and single coronary artery are particularly rare entities and may pose a diagnostic dilemma because of overlapping imaging features. We report a case of a 74-year-old woman who presented with intermittent, non-exertional chest tightness and underwent CTCA for pre-operative cardiac assessment. Imaging demonstrated non-visualization of the right coronary artery (RCA) ostium. The left circumflex artery was dominant and continued along the left atrioventricular groove into the right atrioventricular groove, supplying the right coronary artery territory. Mild non-obstructive atherosclerotic plaques were present in the left anterior descending and left circumflex arteries. No significant myocardial ischemia or structural cardiac abnormality was identified. This case highlights the challenge of differentiating a single coronary artery variant from congenital or acquired right coronary ostial atresia on CTCA. Recognition of key anatomical features is essential, as accurate classification has important implications for prognosis and clinical management. CTCA provides excellent spatial resolution for non-invasive evaluation of coronary artery anomalies, allowing precise anatomical delineation. Given the patient's mild symptoms and absence of significant coronary obstruction, conservative medical management was adopted.

Keywords: Coronary Vessel Anomalies; Coronary Artery Atresia; Single Coronary Artery; Coronary Angiography; Tomography, X-Ray Computed

INTRODUCTION

Coronary artery anomalies are rarely identified, with literature reporting an incidence of approximately 0.6–1.5% in coronary angiograms (1). Among these anomalies, coronary ostial atresia and single coronary arteries (SCA) are least frequently observed (2). Right coronary ostial atresia without associated congenital anomalies in adults remains particularly uncommon (3). This case reports non-visualization of the right coronary artery (RCA) ostium, with the dominant left circumflex artery (LCx) continuing through intercoronary communication along the right atrioventricular groove (AV groove) to supply RCA territory. This being a case of single coronary artery is considered, as such coronary course has not been previously reported in local literature.

CASE REPORT

A 74-year-old female with underlying hypertension, dyslipidemia, hypothyroidism and COPD presented with intermittent chest tightness lasting a few minutes per episode, which was infrequent. There were no other associated symptoms, and episodes occurred both at rest and during exertion. She remained functionally active, exercising three times weekly with 30-minute walks. Clinical examination revealed no significant findings. Electrocardiogram was normal. Echocardiogram demonstrated normal left ventricular size and systolic function, impaired relaxation pattern of diastolic dysfunction with elevated LV filling pressure, no regional wall motion abnormality, and no significant valvular disease. A CT coronary angiography was performed for cardiac assessment prior to her cataract surgery to exclude coronary artery disease. CT showed non-visualization of the RCA ostium with tiny vessel arising from the right coronary sinus (FIGURE 1). The left circumflex (LCx) continued in left atrioventricular (AV) groove to the right AV groove supplying distal RCA territory (FIGURE 2). Otherwise, the left anterior descending coronary artery (LAD) has calcified and non-calcified plaques from proximal to distal causing mild stenosis. The LCx has calcified and non-calcified plaques at proximal segment causing mild stenosis. The CT essentially revealed a left dominant system with non-visualization of the RCA ostium and non-obstructive coronary artery disease. This rare presentation raises diagnostic consideration of single left coronary artery versus right coronary ostial atresia, which will be discussed based on distinguishing radiological features.

DISCUSSION

Non-invasive imaging such as CT Coronary Angiography (CTCA) is now more preferred in evaluation of the coronary arteries, particularly in ruling out coronary artery disease in patients with acute chest pain, pre-operative assessment to rule out coronary stenoses, assessing patency of bypass grafts and stents, and in evaluation of coronary artery anomalies (4). Several studies have highlighted the importance of understanding coronary artery anomaly key imaging features to ascertain its type which guide management in symptomatic patients with coronary anomalies (5). In the present case, the patient had intermittent chest pain and her CTCA demonstrated non-visualization of the RCA ostium, with a dominant left circumflex artery (LCx)

continuing along the left atrioventricular groove into the right atrioventricular groove to supply the RCA territory. These findings raise the diagnostic consideration of either a single coronary artery (SCA) or coronary ostial atresia as differential diagnosis. Differentiating between SCA and coronary ostial atresia involves assessment of coronary origin, course, and collateral circulation patterns (2). In SCA, particularly single left coronary artery variants, typical CT features include absence of a separate RCA ostium, supply of the RCA territory from branches of the left coronary system, and a continuous arterial course originating from either left coronary sinus or a common trunk (3). Compensation by collateral vessels such as Vieussens' arterial ring or other intercoronary collateral pathways may also be seen as described by Angelini *et al*. In our case, CTCA revealed non-visualization of the RCA ostium with collateral supply from the LCx to the RCA territory, which corresponds with features of a single left coronary artery variant. However, coronary ostial atresia whether congenital or acquired can still be considered as differential diagnoses in this case, considering her late symptoms onset and presence of coronary atherosclerotic disease (5). Lipton *et al* described classification of the coronary anomalies, which guides in recognizing these anomalies and their management. The classification of SCA is based on the origin and its course in relation to the ascending aorta and pulmonary trunk (6). Our case revealed that the LCx supplying the RCA territory through extension of collaterals from LCx in right atrioventricular groove as seen on the CTCA. Using the Lipton-Yamanaka classification, the present case exhibits an anomalous RCA coming from the LCx which corresponds to a left type I (L-I type) variant of SCA. This subtype is rare, approximately about 0.016% (6, 7). Generally, it is regarded as benign and normally treated conservatively. SCA has spectrum of manifestations such as angina, syncope, myocardial infarction, malignant arrhythmia and cardiac failure (8). Higher risk of sudden cardiac death reported in SCA patients (9). SCA with origin of RCA from LCx has been reported to have an incidence of 0–0.035% (2). There are cases reported with associated congenital cardiac anomalies specifically valvular anomalies (10).

On the contrary, for coronary ostial atresia – a rare cardiac anomaly (2), CT imaging features described by Gupta *et al* include non-visualization of RCA ostium with a stump at the expected location of RCA ostium (presence of remnant indicates a congenital anomaly rather than total absence of the artery), collateralization from left coronary system similar to left SCA collateralization to supply RCA territory, presence of proximal RCA collateral pathways where collaterals from LAD or LCx directly supply the territory normally supplied by RCA and in certain cases, RCA might have atypical course or anomalously originate from another coronary artery either from LAD or LCx. Following right coronary ostial atresia, this results in spectrum of compensatory mechanisms and clinical manifestations. Patients typically do not experience any symptoms with the development of collateral circulation like Vieussens' arterial ring, mostly from the left coronary artery (11). The degree of collateral growth and the existence of other cardiovascular problems, results in variation of the clinical presentations from moderate exertional

symptoms to severe ischemia episodes (11). Right coronary ostial atresia should be considered different from RCA arising from single coronary trunk, even though the LCA supplies circulation in both conditions (2). This is because in the prior condition, RCA circulation is completely dependent on LCA patency (3). In the present case, we still cannot ascertain if RCA was filling retrogradely almost up to its proximal part by collateral extension from LCx continuing in the right AV groove as depicted by Gupta *et al* in their conventional angiogram. Additionally, on our CTCA, ostial stump of the RCA was not visualized. Lipton *et al* have suggested that in congenital ostial atresia or hypoplasia, a small stump may be detected (6) though this may not always be the case (2). Nevertheless, distinguishing congenital atresia of the ostium from acquired ostial atresia is also important to highlight. Few features are suggestive of congenital coronary ostial atresia most importantly the presence of only 1–2 full diameter connecting collateral vessels without narrowing at the junction of the two vessels (3). On the contrary, acquired cases feature a rich network of septal, infundibular, and atrial collaterals that progressively enlarge postnatally, exhibiting collaterals that are smaller than distal recipient vessels. (2). Additionally, absence of typical clinical angina and lack of evidence of myocardial scarring is highly indicative of coronary ostial atresia (2). Several acquired causes which include atherosclerosis, syphilis, Kawasaki, and Takayasu arteritis (3) must be also ruled out in an adult patients when assessing coronary ostial atresia in CTCA. Overall, the CTCA findings are most consistent with a Lipton L-I type single coronary artery based on non-visualization of RCA ostium, dominant LCx continuation into the right AV groove and supply of RCA territory without an independent RCA. Right coronary ostial atresia remains a close differential due to overlapping collateralization patterns.

CONCLUSION

This case highlights the diagnostic challenge of distinguishing a Lipton L-I type single coronary artery from right coronary ostial atresia on CT coronary angiography. In our patient, the findings were more suggestive of a single coronary artery variant because the dominant left circumflex artery continued into the right atrioventricular groove to supply the RCA territory, and non-visualization of RCA ostial stump. Right coronary ostial atresia remained a close differential diagnosis because of overlapping collateral supply patterns. This distinction is clinically important, as accurate anatomical classification may influence prognostic assessment, surveillance, and management decisions. (4). Management of coronary anomalies varies based on symptoms, ranging from conservative treatment to surgical intervention (12). Accurate differentiation between these entities is important because single coronary artery variants are generally benign and managed conservatively, whereas coronary ostial atresia may require closer surveillance depending on collateral adequacy and ischemic risk. Given the patient's mild symptoms and absence of hemodynamically significant stenosis, conservative medical management was appropriate.

DATA AVAILABILITY

Further information regarding the data used for this work can be obtained upon reasonable request.

ACKNOWLEDGEMENT

We extend our sincere appreciation to our esteemed colleagues in for their unwavering support and invaluable guidance.

FUNDING:

This work received no external funding.

CONFLICT OF INTEREST:

The authors have no conflicts of interest to declare and is in agreement with the contents of the manuscript.

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FIGURE LEGENDS:

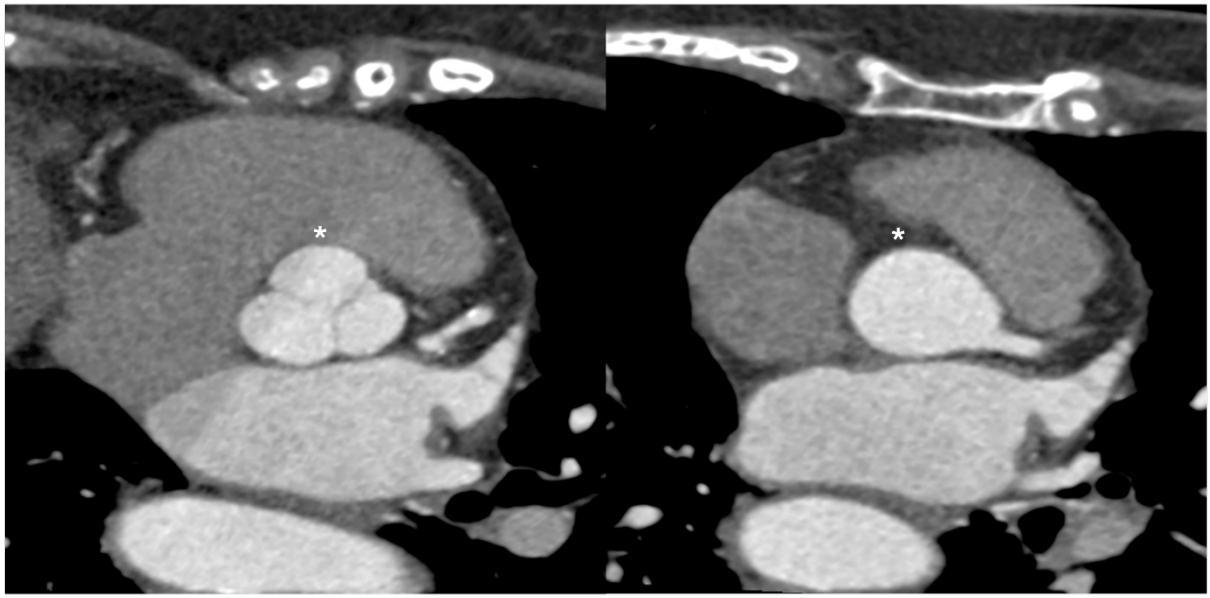

Figure 1. Axial CT coronary angiography image demonstrating non-visualization of the right coronary artery ostium (*).

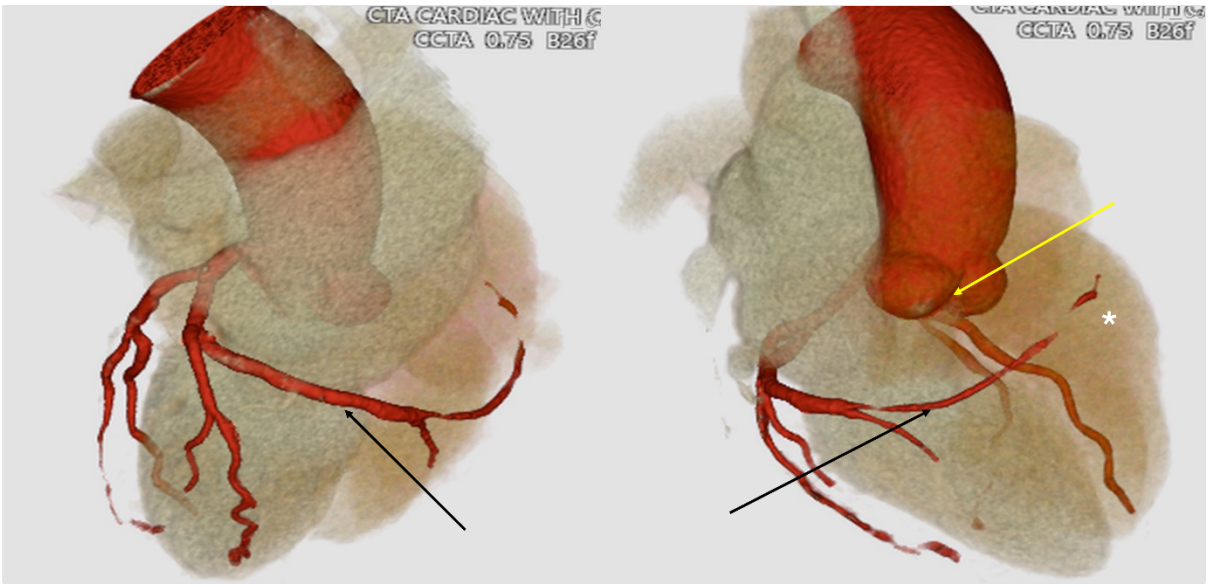

Figure 2. Volume-rendered CT coronary angiography image showing non-visualization of the right coronary artery ostium (yellow arrow). The left circumflex artery (black arrow) continues along the left atrioventricular groove into the right atrioventricular groove to supply the distal right coronary artery territory (*).