Radiology Case Reports 19 (2024) 3080–3083

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journal homepage: www.elsevier.com/locate/radcr

Case Report

# Anomalous origin of the circumflex artery from the right pulmonary artery ☆

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ARTICLE INFO

*Article history:*
Received 27 February 2024
Accepted 20 April 2024

*Keywords:*
Cardiac
Anomalous coronary artery
Circumflex artery
Computed tomography

ABSTRACT

Anomalous origin of the circumflex artery from the pulmonary artery (ACxAPA) is a rare but clinically significant condition in which the circumflex artery arises from either the main pulmonary artery or one of its main branches. Untreated patients with ACxAPA may develop severe heart failure or sudden cardiac death. Diagnosis is established with either catheter or CT angiography. We present a case of an adult male with no prior known cardiac history who was found to have ACxAPA after presenting to our institution in acute decompensated heart failure.



## Introduction

Anomalous origins of the coronary arteries are a well-known entity, and may be broadly categorized as anomalous aortic origin, or rarely, pulmonary arterial origin [1,2]. Of the pulmonary artery variants, the anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) has been described most frequently [2]. Anomalous origin of the right coronary artery (RCA), left anterior descending artery (LAD), and circumflex artery (LCx) from the pulmonary artery are extremely rare [1,2]. Additionally, the anomalous origin of the circumflex artery from the pulmonary artery (ACxAPA) is often associated with other congenital cardiac defects, some of which include coarctation of the aorta, bicuspid aortic valve, and atrial septal defects [1–3]. The clinical presentation of ACxAPA is variable depending on the degree of compensatory vessel collateralization and the presence of other cardiac abnormalities [2]. Diagnosis is established with direct visualization of the coronary circulation, either by catheter or CT angiography [1,2,4,5]. Only a handful of case reports describing patients with ACxAPA have been published, most of which have had additional congenital defects [1], and fewer in which ACxAPA was the only cardiovascular defect [6,7].

We present a 42-year-old man, with no known congenital cardiovascular abnormalities, who was diagnosed with ACxAPA after presenting to our institution for symptoms related to acute decompensated heart failure with reduced ejection fraction.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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https://doi.org/10.1016/j.radcr.2024.04.061

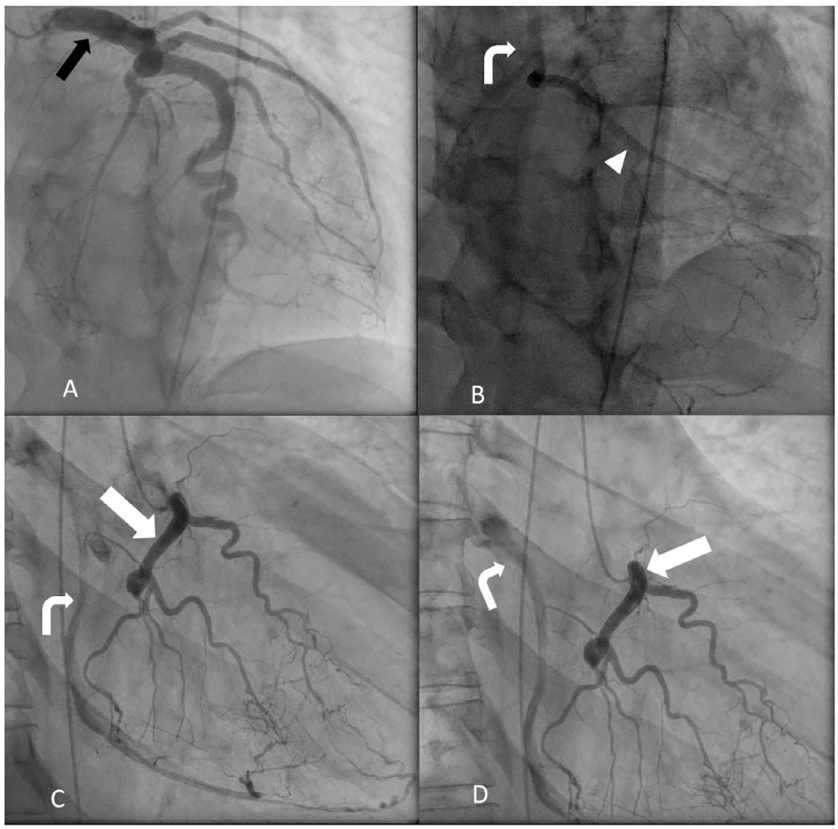


**Fig. 1 – Catheter coronary angiography in our patient with anomalous origin of the left circumflex artery (LCx) from the right pulmonary artery (PA). (A) Dilated, tortuous left anterior descending coronary artery (LAD) (black arrow). (B) Late filling of the anomalous LCx (curved arrow) and posterior descending artery (arrowhead) after contrast has been washed out of the LAD, compatible with fistulous connections. Note this is a left dominant coronary system with the posterior descending coronary artery arising from the LCx. (C) Right coronary artery (RCA) (white arrow) with numerous fistulous connections to the anomalous LCx (curved arrow). (D) Robust collateralization between the RCA (white arrow) and the LCx (curved arrow). Contrast can be seen emptying into the right PA at the origin of the LCx.**

## Case presentation

A 42-year-old man with a history of hypertension presented to the emergency department for one month of worsening shortness of breath. Initial laboratory results revealed troponins elevated to 246 ng/L (normal ≤15 ng/L) with repeat at 194 ng/L, D-dimer 1742 ng/mL (normal ≤500 ng/mL), and BNP 9570 pg/mL (normal ≤72 pg/mL). Electrocardiogram showed left ventricular hypertrophy but no ST segment elevation. Chest radiograph showed an enlarged heart and evidence of increased pressure pulmonary edema. Chest CTA with pulmonary embolism protocol showed no pulmonary embolism but revealed significant left ventricular enlargement and evidence of increased pressure pulmonary edema. Subsequent echocardiogram confirmed a severely enlarged, hypokinetic left ventricle with an ejection fraction of 15% consistent with acute decompensated heart failure with reduced ejection fraction.

To further elucidate the etiology of heart failure, the patient underwent cardiac catheterization (Fig. 1) which revealed significantly dilated left and right coronary arteries and the early appearance of abnormal venous drainage suspicious for arteriovenous communication. There was no significant coronary artery stenosis. Prospectively gated coronary CTA was then performed (Fig. 2), demonstrating an anomalous origin of the LCx from the right pulmonary artery (PA), 2.3 cm from the bifurcation. The LAD and RCA were markedly dilated and tortuous with normal origins. Coronary venous drainage was normal, and no other cardiac anomaly was visualized. The suspected etiology of our patient's heart failure was vascular steal from fistulous connections between the LAD, RCA, and LCx. The patient was initiated on guideline directed medical therapy (GDMT) and underwent implantable cardioverter-defibrillator placement. At the time of this report, he was still undergoing evaluation for cardiac transplant.

## Discussion

The term congenital coronary artery anomaly covers an array of anatomic variations, from anomalous origins to variations in termination [4,8]. A basic understanding of coronary artery embryology is important to better understand these anomalies. During early cardiac development, cells in the atrioven-

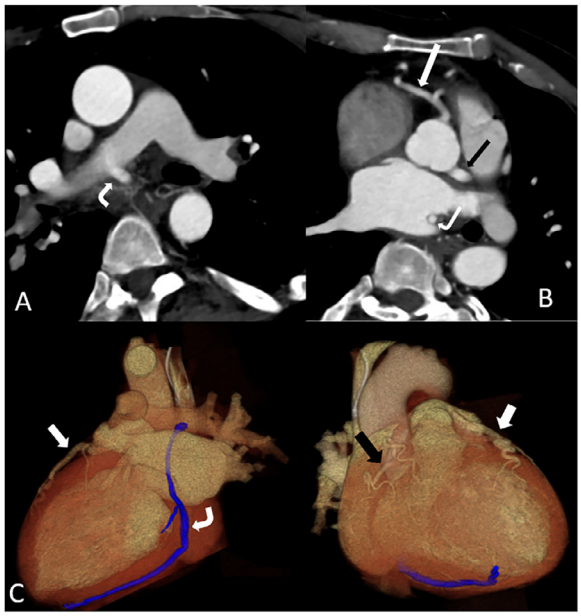


**Fig. 2 – Prospectively gated cardiac CTA of the chest in an axial oblique plane at the level of the pulmonary artery (PA) bifurcation (A) and aortic root (B). (A) Anomalous origin of the left circumflex artery (LCx) (curved arrow) from the right PA. Contrast from the LCx can be seen mixing with pulmonary arterial blood at the site of origination. (B) Normal origins of the left anterior descending artery (LAD) (black arrow) and right coronary artery (RCA) (white arrow). The anomalous LCx (curved arrow) can be seen coursing along the left side of the left atrium, between the left atrial appendage and left pulmonary vein ostia. (C) 3-Dimensional volume rendered reconstructions of the cardiac CTA showing the anomalous LCx (curved arrow) arising from the right PA and coursing along the left side of the left atrium, between the left atrial appendage and left pulmonary vein ostia and distally along the left atrioventricular grove. The LAD (black arrow) and RCA (white arrow) are dilated and tortuous.**

tricular epicardium undergo epithelial to mesenchymal transition, creating migratory mesenchymal cells which eventually differentiate to form the endothelial lining and smooth muscle cells of the coronary vessels [9]. Vasculogenesis continues and vessels ultimately spread into the myocardium [9]. Finally, the proximal coronary arteries grow into the aorta under the guidance of several chemical signals, creating the source of oxygenated blood for the myocardium [9,10]. It is proposed that alterations in this chemical signaling process is the mechanism behind anomalous origins of the coronary arteries [10].

The clinical presentation of ACxAPA varies from asymptomatic to severe heart failure or sudden cardiac death [2]. The suggested mechanism behind symptom development in ACxAPA is due to steal phenomenon, similarly to what has been described in ALCAPA [1,2,4]. Because blood pressure in the PA is less than the aorta, an anomalous LCx offers a lower resistance pathway for blood flowing through the LAD and RCA collaterals. Subsequent shunting of blood results in myocardial ischemia, which may lead to heart failure if not promptly recognized and treated.

The development of collateral circulation may prove beneficial in earlier stages of the disorder. If collateral circulation is sufficient, oxygenated blood still reaches the myocardium supplied by the LCx and symptom onset is delayed unless myocardial demand exceeds supply [2]. On the other hand, in patients who do not develop robust collateralization, the myocardium normally supplied by the LCx becomes ischemic from the lack of oxygenated blood supply and symptoms present earlier [1]. It is suspected that despite robust collateralization, repeated episodes of steal eventually lead to the onset of severe heart failure in our patient.

The diagnosis of coronary artery anomalies is performed with coronary catheterization or with less invasive measures such as coronary CTA [1,2,4]. Findings that suggest anomalous origin of a coronary artery from the PA include dilated and tortuous coronary vessels and collateralization, often with retrograde flow into the PA [1,11]. According to the American Heart Association and American College of Cardiology 2018 guidelines for management of adults with congenital heart diseases, surgical management is recommended when an anomalous left coronary artery originates from the PA, or in symptomatic patients with anomalous RCA from the PA [12]. The most common surgical procedure is reimplantation of the vessel onto the aorta to restore flow to the affected myocardium; however other options such as coronary artery bypass graft with ligation of the anomalous artery have also been described [1,5]. Despite these recommendations, there are no specific guidelines regarding the management of patients with ACxAPA.

In conclusion, anomalous origins of the coronary arteries are rare, but potentially life-threatening conditions requiring a high degree of suspicion and advanced imaging to diagnose. This case highlights the value that advanced imaging studies, such as cardiac gated CTA, provide in the evaluation of these disorders.

## Patient consent

The patient provided written informed consent for the publication of this case report.

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