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Diagnostic challenge in an elderly patient with ARCAPA

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Introduction

Anomalous Origin of Right Coronary Artery from the Pulmonary Artery (ARCAPA) is an extremely rare congenital coronary anomaly with an estimated prevalence of less than 0.002% of the general population. Unlike its more common counterpart, Anomalous Left Coronary Artery from the Pulmonary Artery (ALCAPA), ARCAPA often remains asymptomatic until adulthood due to the development of extensive collateral circulation from the left coronary system. This collateralization can mask the true nature of the anomaly on conventional coronary angiography, leading to misdiagnosis as chronic total occlusion (CTO) of the right coronary artery (RCA), particularly in elderly patients.

The clinical presentation of ARCAPA varies widely, ranging from incidental findings on imaging studies to myocardial ischemia, heart failure, or sudden cardiac death. According to a systematic review by Guenther et al., only 223 cases had been reported in the literature, with a slight male predominance (54%) and a bimodal age distribution at diagnosis, with peaks near birth and between 40-60 years of age. This bimodal distribution highlights the diagnostic challenges associated with this condition, as many cases remain undiagnosed until adulthood when symptoms become apparent or when patients undergo coronary imaging for unrelated cardiac conditions.

Case presentation

A 69-year-old male with a past medical history of hypertension, insulin-dependent diabetes mellitus type 2, and chronic kidney disease stage 3 was admitted for NSTEMI in January 2022. He underwent left heart catheterization (LHC). He was found to have CTO of RCA with well collateralized from the left coronary artery system, 60% stenosis of the proximal left circumflex artery (LCX), and non-obstructive left anterior descending artery (LAD). The patient was treated medically without percutaneous coronary intervention (PCI). The patient had worsening chest pain on exertion in April 2023. He subsequently has an abnormal myocardial perfusion study consistent with a medium-sized region of severe inferior myocardial ischemia. Repeated LHC in July 2023 shows CTO of proximal to mid-LCX with unsuccessful PCI, unchanged CTO of RCA, and non-obstructive LAD lesions. However, there is a concern about a fistula between the RCA and the right atrium.

Investigations: Upon evaluation at our institution in August 2023, the cardiothoracic surgery team ordered a coronary computed tomography angiography (CCTA) to better characterize the coronary anatomy before proceeding with surgical intervention. The imaging findings fundamentally changed the diagnosis and management approach for this patient.

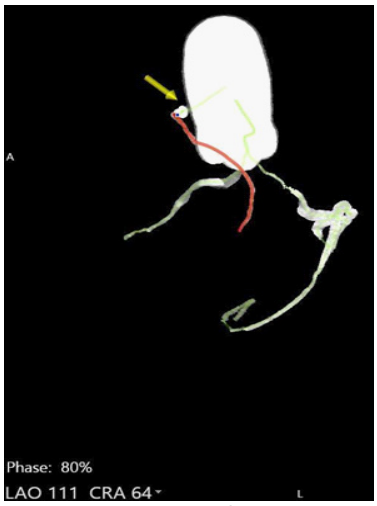

Figure 1: Coronary angiogram showing extensive collaterals. Angiographic image demonstrating extensive collateralization from left coronary system to distal RCA. The yellow arrow indicates the point where the RCA ostium should normally connect to the aorta but is absent.

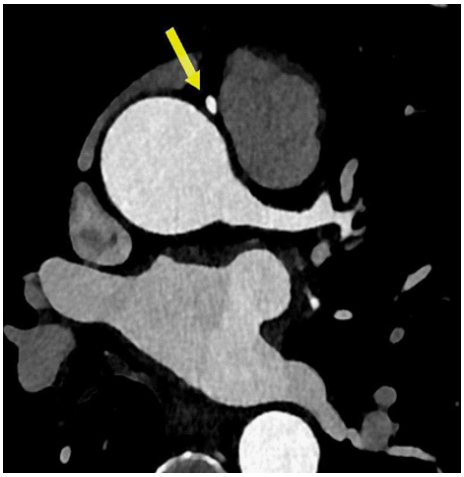

Figure 2: Axial CCTA showing anomalous RCA origin. Axial CCTA view revealing the origin of the RCA (yellow arrow) from the pulmonary artery rather than the expected aortic origin, confirming ARCAPA diagnosis.

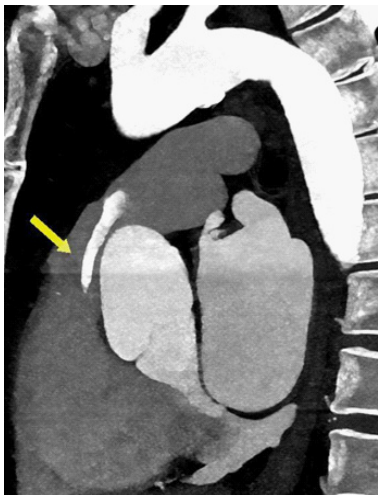

Figure 3: Sagittal CCTA demonstrating ARCAPA. Sagittal CCTA view showing the RCA (yellow arrow) arising from the pulmonary artery with retrograde flow of contrast back to the main pulmonary artery.

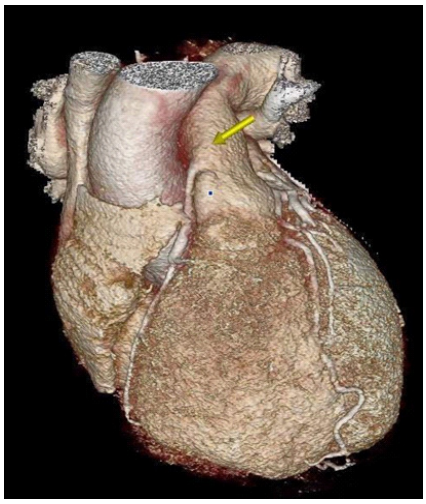

Figure 4: 3D reconstruction confirming anomalous anatomy. Three-dimensional CCTA reconstruction confirming the anomalous RCA (yellow arrow) originating from the pulmonary trunk rather than the aortic root.

The CCTA revealed that what had previously been interpreted as CTO of the RCA on conventional angiography was actually ARCAPA. The right coronary artery was found to originate from the pulmonary artery rather than the aorta, with extensive collateral circulation from the left coronary system. This explained the unusual angiographic appearance that was previously misinterpreted as a CTO.

Additional findings on the CCTA included:

- Soft plaques throughout the mid and distal RCA causing up to severe stenosis (75-99%).
- Complete occlusion of the proximal LCX, with the distal LCX in the interventricular groove remaining patent and perfused by collateral flow.
- Irregular soft plaques in the distal LAD causing severe stenosis (75-99%), with patent proximal and mid LAD segments.

The CCTA definitively ruled out the previously suspected coronary fistula between the RCA and the right atrium. This imaging modality was crucial in establishing the correct diagnosis of ARCAPA, which significantly altered the surgical approach.

Discussion

This case highlights several important aspects of ARCAPA, particularly in elderly patients. First, the misdiagnosis of ARCAPA as CTO of the RCA on conventional angiography is a common pitfall. The extensive collateral circulation that develops from the left coronary system to supply the RCA territory can mimic the appearance of a CTO with retrograde filling. This misdiagnosis can lead to inappropriate management decisions and delay definitive treatment.

The presentation of ARCAPA in elderly patients differs significantly from that in younger patients. While infants and children with ARCAPA may present with heart failure, myocardial infarction, or sudden death, elderly patients often have a more indolent course due to the protective effect of well-developed collateral circulation. However, as atherosclerotic disease pro-

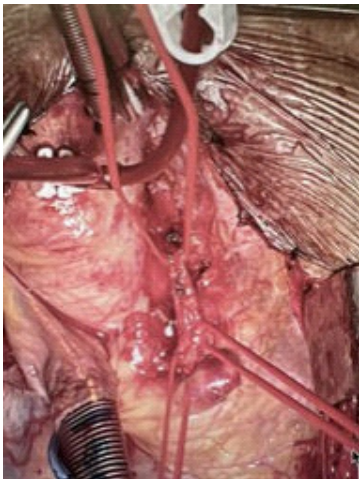

Figure 5: Intraoperative view showing the surgical field with exposure of the anomalous right coronary artery.

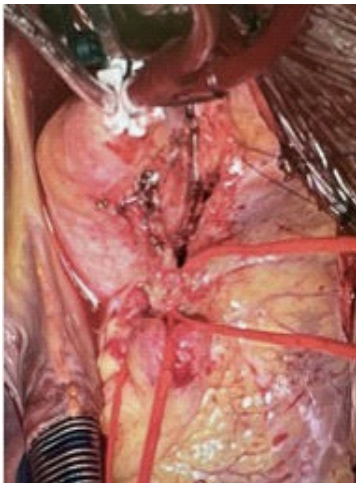

Figure 6: Completion of the RCA translocation from the pulmonary artery to the aorta.

gresses in these collateral vessels, as observed in our patient, symptoms of myocardial ischemia may develop or worsen.

CCTA played a pivotal role in establishing the correct diagnosis in our patient. While conventional angiography has been the traditional gold standard for coronary artery assessment, it has limitations in defining the origin of coronary arteries, particularly in complex congenital anomalies. CCTA offers superior visualization of coronary origins and their spatial relationships with adjacent cardiac structures, making it the imaging modality of choice for suspected coronary anomalies.

The management of ARCAPA involves surgical correction, even in asymptomatic patients, due to the risk of sudden cardiac death. Various surgical techniques have been described, including direct reimplantation of the RCA into the aorta, creation of an intrapulmonary tunnel, or CABG with ligation of the anomalous RCA. The choice of technique depends on the anatomical details and the presence of concomitant cardiac lesions.

Information/management

The patient underwent surgical correction via median sternotomy with translocation of the aberrant right coronary artery from the pulmonary artery to the aorta in August 2023. Surgical correction is recommended for ARCAPA regardless of symptom status due to the associated risk of sudden cardiac death. The primary goals of surgery are to eliminate left-to-right shunting and establish dual coronary circulation.

Conclusion

This case illustrates the diagnostic challenge of ARCAPA in elderly patients and the crucial role of advanced cardiac imaging, particularly CCTA, in establishing the correct diagnosis. The misdiagnosis of ARCAPA as CTO of the RCA on conventional angiography is a common pitfall that can lead to inappropriate management decisions.

The case emphasizes the importance of considering congenital coronary anomalies in the differential diagnosis of apparent CTO, especially when extensive collateralization is present. It also highlights the value of a multidisciplinary approach involving cardiologists, radiologists, and cardiac surgeons in the management of complex cardiovascular conditions.

Early recognition and appropriate surgical correction of ARCAPA can significantly improve patient outcomes, as demonstrated in our case. The successful translocation of the aberrant RCA from the pulmonary artery to the aorta resulted in marked improvement of the patient's cardiac symptoms and established normal dual coronary circulation.

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