



ARCAPA Management in an 80-year-old Female with Complex Coronary Disease

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Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital anomaly, often discovered incidentally or during ischemic evaluation. We present a complex case of an 80-year-old female with multivessel calcific coronary artery disease (CAD), ischemic mitral regurgitation (MR), history of atrial fibrillation status post ablation who presented with dyspnea subsequently diagnosed ARCAPA. The patient underwent triple coronary artery bypass grafting (CABG), ligation of the anomalous RCA, mitral valve repair and left atrial appendage clip placement. This case highlights the diagnostic challenges and therapeutic decision-making in managing concurrent congenital and acquired cardiac pathology.

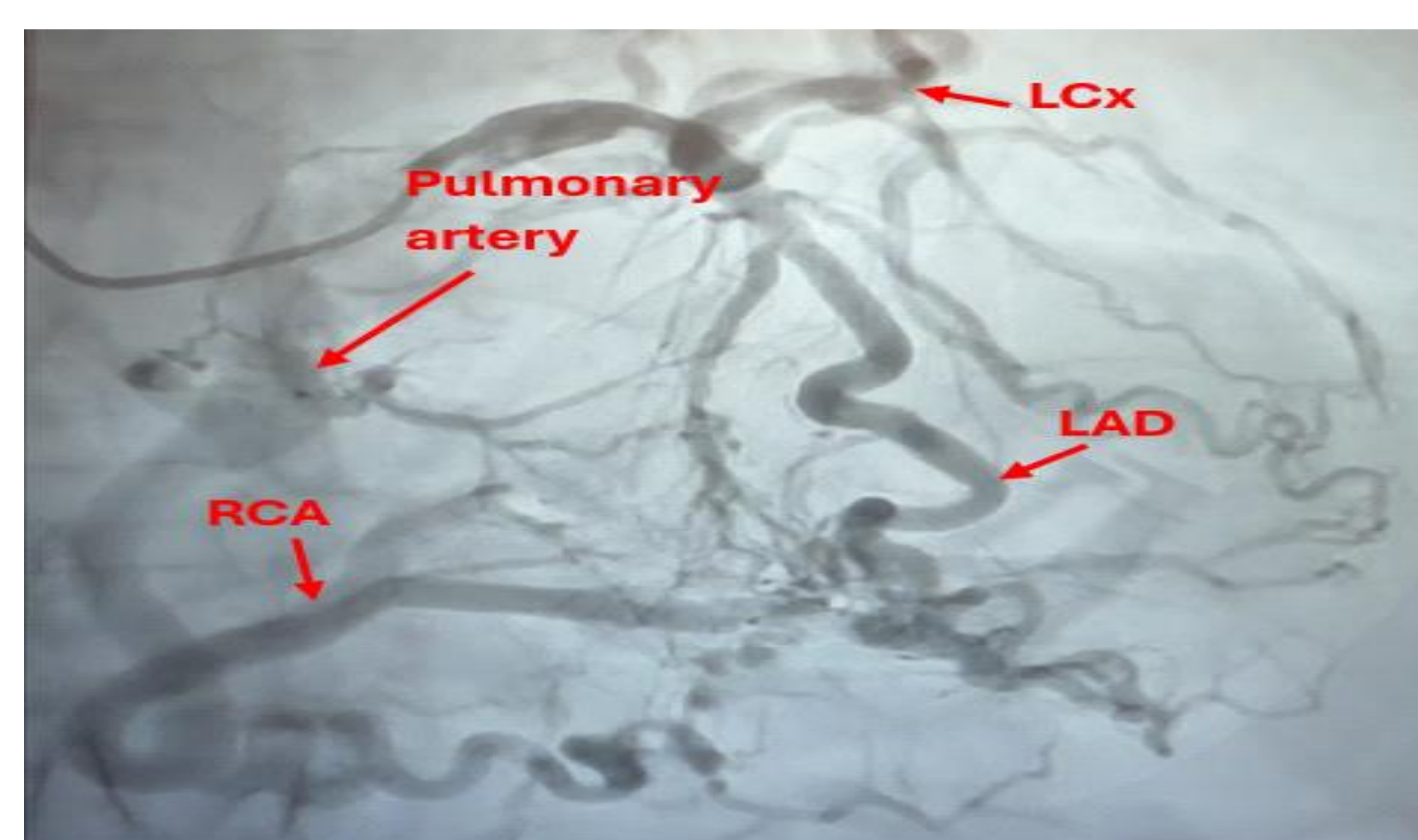
Anomalous origin of a coronary artery is a rare congenital condition, with reported incidence ranging from 0.64% to 1.6% in the general population. Among these anomalies, the origin of the right coronary artery originating from the pulmonary trunk (ARCAPA) is uncommon and occurs less frequently than anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) [1,2]. Clinical outcomes often depend on coronary dominance, as patients with a left-dominant system generally tolerate the anomaly better than those with right-dominant circulation [3]. We report the case of an 80-year-old female with a left-dominant coronary circulation who was found to have an anomalous RCA originating from the pulmonary trunk. The anomaly was identified with angiography in the setting of severe coronary artery disease and managed with coronary artery bypass grafting (CABG).

Case Presentation

- An 80-year-old female with a medical history of coronary artery disease, atrial fibrillation status post ablation, hypertension, dyslipidemia, and prior tobacco use, presented with chest pain, dyspnea on exertion, tachycardia, and atypical exertional angina prompting further cardiac evaluation.
- A chest computed tomography angiogram (CTA) was performed to rule out pulmonary embolism, which revealed extensive coronary calcification. Subsequent, left, and right heart catheterization demonstrated severe calcific 95% proximal circumflex artery stenosis, mild distal left main artery disease, minimally diseased LAD, stenosis of obtuse marginal branch, anomalous RCA originating from the main pulmonary artery with retrograde filling, moderate pulmonary hypertension, and ischemic mitral regurgitation.
- The RCA drained predominantly into the left pulmonary artery with evidence of a left-to-right shunt confirmed via oxygen saturation step-up. LV systolic function was fair with diastolic dysfunction (LVEDP 20 mmHg). There was no prior history of significant MR, suggesting new onset ischemic MR, likely secondary to the circumflex lesion.

Discussion

Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital anomaly (~0.002% incidence), far less common than ALCAPA. While often asymptomatic in childhood due to collateral development, it can later lead to ischemia, heart failure, or sudden cardiac death. Diagnosis relies on multimodal imaging, with angiography confirming retrograde RCA filling and drainage into the pulmonary artery. Surgical correction is universally recommended, regardless of symptoms, to prevent long-term complications. Options include RCA reimplantation or bypass grafting with ligation of the anomalous pulmonary connection, the latter being essential to eliminate coronary steal. In our case, CABG with mitral valve repair and left atrial appendage clip was performed along with RCA ligation due to multivessel disease. This highlights the importance of early recognition, thorough imaging, and a comprehensive surgical approach that addresses both congenital and acquired pathology.



ARCAPA, though rare and often asymptomatic, poses significant long-term risks including myocardial ischemia, heart failure, and sudden cardiac death. This case underscores the necessity of maintaining a high index of suspicion in patients with unexplained ischemia or arrhythmia and highlights the indispensable role of multimodal imaging for definitive diagnosis. Once identified, surgical intervention is essential, regardless of symptom burden. Timely ligation of the anomalous vessel and revascularization of the affected myocardium are critical to prevent coronary steal and ensure adequate perfusion. A comprehensive surgical approach addressing both congenital anomalies and coexisting structural heart disease maximizes patient survival and quality of life.



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