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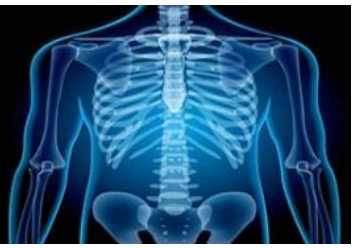

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ALCAPA in adulthood: A CT perspective

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Abstract
Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital abnormality of the coronary arteries. It typically presents with acute, non-exertional left-sided chest pain, accompanied by sweating and palpitations, often in the presence of a significant medical history. The condition is commonly diagnosed through advanced imaging techniques, such as CT coronary angiography or cardiac MRI, which provide detailed views of the origin and path of the abnormal coronary artery. Early identification and timely surgical intervention aimed at restoring normal coronary circulation generally lead to excellent outcomes, with gradual recovery of the heart muscle. This case report discusses a late presentation of ALCAPA syndrome.

Keywords: Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), Bland-White-Garland syndrome, coronary steal, inter- coronary collaterals, left-to-right shunt

Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) syndrome, also known as Bland-White-Garland syndrome, is an uncommon congenital heart defect, occurring in approximately 1 in 300,000 live births [1] and representing 0.25%-0.5% of all congenital heart conditions [2]. This condition typically presents as an isolated abnormality, although in 5% of cases**,** it may be associated with other heart defects, including Atrial Septal Defect (ASD)**,** Ventricular Septal Defect (VSD) and aortic coarctation [3].

ALCAPA syndrome causes the "coronary steal" phenomenon, where a left-to-right shunt disrupts normal perfusion to the left ventricle. It is one of the leading causes of myocardial ischemia and infarction in children. Without treatment, up to 90% of infants with ALCAPA will not survive past the first year of life [4]. In individuals who survive into adulthood, ALCAPA can result in complications such as myocardial infarction, left ventricular dysfunction, mitral regurgitation, and silent myocardial ischemia, all of which increase the risk of sudden cardiac death.

Case report

A 42-year-old female patient sudden, non-exertional left-sided chest pain, along with sweating and palpitations. Physical examination was unremarkable.

Echocardiography/ECG showed complete right bundle branch and left anterior fasicular block, ST depression (II, aVF, V2, V3, V4, V5, V6), deep negative T waves (V1, V2), findings suggestive of anteroseptal myocardial infarction.

A contrast-enhanced coronary CT angiography, performed on a Canon Aquilion Prime SP 160-slice CT scanner, revealed a clockwise rotation of aortic root with posterior non coronary cusp seen on the right side, with Right Coronary Artery (RCA) arising from 12 o' clock with severe dilatation and tortuosity of the RCA from its origin. Multiple tortuous inter-coronary collateral arteries were seen arising from RCA and its branches, coursing along the interventricular septum and epicardial surface of heart.

Left Main Artery (LM) was also severely dilated, tortuous and was seen arising from inferior aspect of Main Pulmonary Artery (MPA) in keeping with ALCAPA. There was lesser dense contrast opacification of the MPA at the level of LM suggesting flow via collaterals from posterior descending artery into Left Anterior Descending (LAD) into LM into MPA pointing towards left to right shunt. The MPA, left Pulmonary Artery (LPA) and Right Pulmonary Artery (RPA) are more opacified than the aorta.

Subsequent 2D echo revealed an ejection fraction of 30-35%, large septal collaterals in intraventricular septum and dilated RCA ostium.

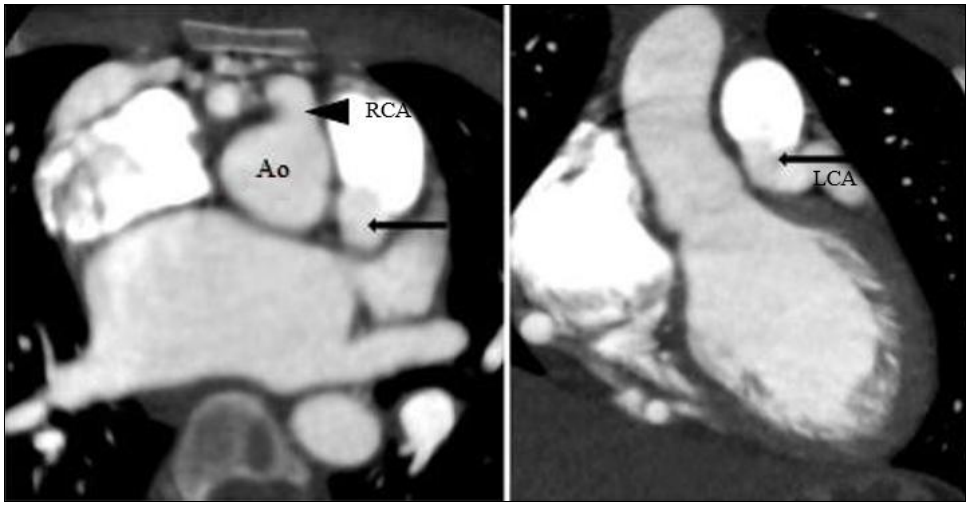

Fig 1: ALCAPA syndrome in a 42-year-old woman. Axial and coronal multidetector CT angiogram shows the origin of the LCA (arrow) from the main pulmonary artery *(PA)* and the dilated RCA (arrowhead). Ao = aorta

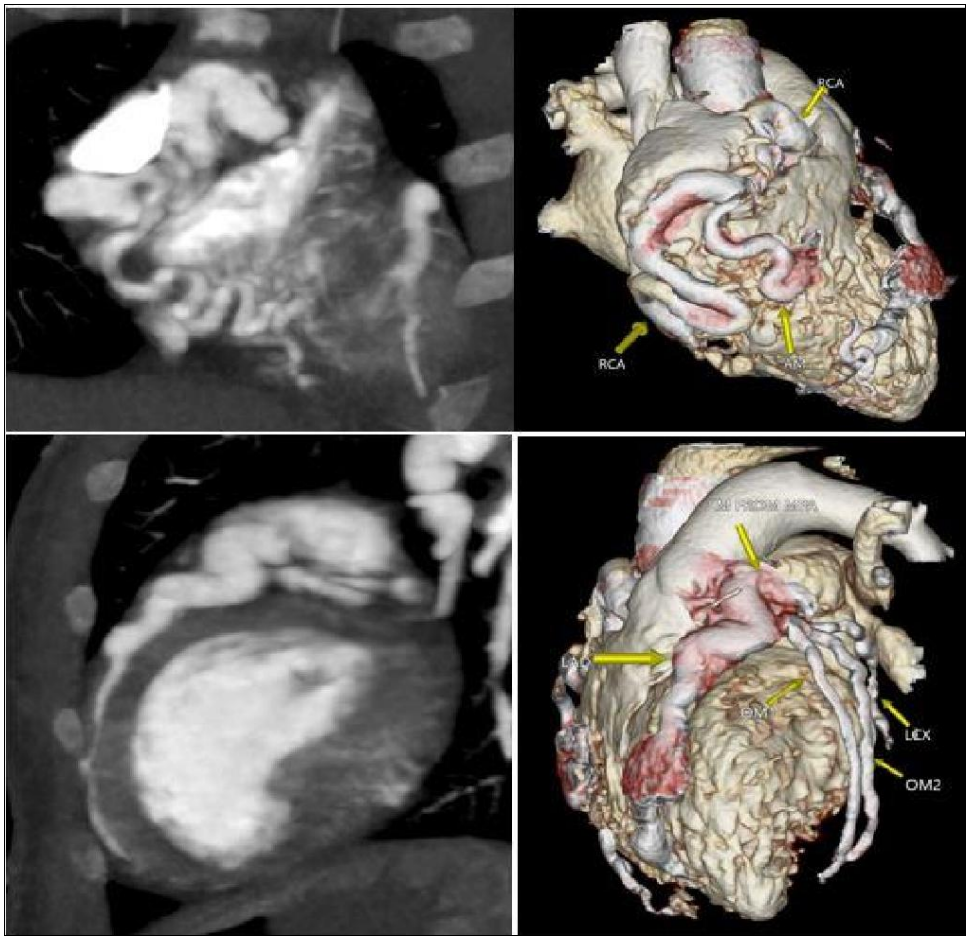

Fig 2: Multidetector CT angiogram (Maximum Intensity Projection images) and three-dimensional volume-rendered multidetector CT angiogram shows severely dilated and tortuous coronary arteries(arrows) with multiple tortuous inter-coronary collateral arteries seen arising from RCA and its branches

Discussion

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a uncommon congenital coronary anomaly. Without correction, most patients die in infancy due to severe myocardial ischemia. Survival into adulthood, as in the present case, is uncommon and depends on the development of extensive intercoronary collateral circulation from the right coronary artery and favorable hemodynamic adaptations that sustain left coronary perfusion.

Adult patients may present with variable symptoms, including exertional or non-exertional chest pain, palpitations, heart failure, or arrhythmias. Sudden cardiac death remains a recognized risk. In this case, the patient presented with acute chest pain and ECG abnormalities suggestive of myocardial infarction, highlights the diagnostic challenge.

Conventional angiography can demonstrate the course of anomalous coronary arteries but may be technically demanding or difficult to interpret. In contrast, both

multidetector CT and MRI provide high diagnostic accuracy in identifying the anomalous coronary origin and proximal course. MRI offers detailed assessment of myocardial viability and perfusion, whereas CT provides superior spatial resolution of coronary anatomy. In adults with ALCAPA, imaging typically demonstrates the left main coronary artery originating from the pulmonary artery, enlargement of the right coronary artery, and the presence of dilated collateral vessels along the epicardial surface and interventricular septum. In this case, CT angiography was pivotal, clearly delineating the anomalous origin, tortuous RCA and extensive collateral circulation.

Definitive treatment of ALCAPA requires surgical restoration of a two-coronary system. Surgical options include direct reimplantation of the left coronary artery into the aorta, aortocoronary bypass grafting using either saphenous vein or internal mammary artery, or the Takeuchi intrapulmonary tunnel procedure, with the choice depending on anatomic considerations. Early surgical intervention is strongly advised even in adult patients, as it prevents progression of left ventricular dysfunction, malignant arrhythmias, and sudden death.

Conclusion

This case highlights the rare survival of ALCAPA into adulthood, diagnosed through CT coronary angiography. The presence of a dilated RCA with extensive collateralization and retrograde flow into the pulmonary artery are key imaging features. Early recognition and surgical correction remain essential to improve long-term outcomes and prevent life-threatening complications. The uniqueness of this case lies in its late diagnosis, as the patient presented at the age of 42 years.

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