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Case Report

Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA): A Case Report

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A B S T R A C T

Anomalous origin of the left coronary artery from the pulmonary artery, commonly designated as ALCAPA or Bland-White-Garland syndrome, constitutes a rare yet clinically critical congenital coronary anomaly. The condition is characterized by the emergence of the left coronary artery from the pulmonary arterial trunk rather than from the aortic sinus, resulting in compromised myocardial perfusion. The ensuing coronary steal phenomenon and progressive left-to-right shunting predispose affected patients to myocardial ischemia, ventricular dysfunction and, in the absence of treatment, a high risk of early mortality. Although the anomaly may occur in isolation, it is occasionally associated with additional congenital cardiac malformations.

We report the case of a one-year-old infant in whom ALCAPA was identified through multimodality imaging, including echocardiography and multislice computed tomography coronary angiography. Imaging demonstrated marked dilatation of the right coronary artery, extensive intercoronary collateralization and an associated juxtaposition of the left atrial appendage. This report highlights the diagnostic value of advanced cardiac imaging in defining coronary anatomy and guiding definitive surgical management while underscoring the importance of early detection in improving clinical outcomes.

Keywords: Anomalous origin; Left coronary artery; Pulmonary artery

Introduction

Congenital anomalies involving the coronary arteries encompass a heterogeneous spectrum of abnormalities affecting their origin, anatomical course or termination[1]. Among these entities, anomalous origin of the left coronary artery from the pulmonary artery represents one of the most hemodynamically significant despite its low incidence. Epidemiological estimates suggest that ALCAPA accounts for approximately 0.25% to 0.5% of congenital heart defects, with an overall prevalence approaching one case per 300,000 live births[2].

The pathophysiological consequences of this anomaly are profound. In normal physiology, both coronary arteries arise from the high-pressure, oxygen-rich aortic root. In ALCAPA, however, the left coronary artery originates from the low-pressure pulmonary circulation. Following the postnatal decline in pulmonary arterial pressure and oxygen saturation, myocardial perfusion through the left coronary system becomes progressively inadequate. This results in chronic subendocardial ische-

mia, left ventricular dysfunction and varying degrees of mitral insufficiency secondary to papillary muscle ischemia[3].

Clinical expression depends largely on the development of collateral vessels between the right and left coronary systems. In the absence of sufficient collateralization, most infants develop heart failure early in life and mortality rates remain extremely high without surgical correction. Contemporary advances in noninvasive cardiac imaging, particularly multislice CT coronary angiography, have substantially improved diagnostic accuracy and preoperative anatomical assessment.

Case Presentation

A one-year-old infant was referred to our tertiary paediatric cardiology centre for evaluation of persistent respiratory distress and failure to thrive. The child had been born at term following an uncomplicated pregnancy, with no antenatal suspicion of congenital heart disease. Over the preceding months, the parents had noted progressive feeding intolerance, excessive diaphoresis during crying and poor weight gain compared with age-matched peers.

On clinical examination, the infant appeared tachypnoeic with mild subcostal retractions. Peripheral pulses were palpable and symmetrical, though mild tachycardia was present. Cardiac auscultation revealed a grade III/VI systolic murmur best heard along the left parasternal border, radiating toward the apex. No cyanosis was observed, but signs of early congestive heart failure were clinically suspected.

Chest radiography **(Figure 1)** demonstrated cardiomegaly accompanied by increased pulmonary vascular markings suggestive of pulmonary congestion. Electrocardiography **(Figure 2)** revealed nonspecific ST-segment and T-wave abnormalities raising suspicion of myocardial ischemia. Transthoracic echocardiography **(Figure 3)** Transthoracic echocardiography provided crucial anatomical and functional information. The examination demonstrated marked dilatation of the left ventricle with globally reduced systolic function. The left ventricular walls appeared hypokinetic, particularly in the anterolateral segments. Increased echogenicity of the mitral papillary muscles was noted, a finding highly suggestive of ischemic injury and papillary muscle fibrosis, which correlated with the presence of mild-to-moderate mitral regurgitation on colour Doppler imaging. Careful assessment of the coronary origins revealed anomalous emergence of the left coronary artery from the main pulmonary artery rather than from the left aortic sinus. Colour Doppler interrogation showed retrograde flow from the left coronary system into the pulmonary artery, further supporting the diagnosis. The right coronary artery appeared dilated, reflecting compensatory increased flow and collateral development.

For comprehensive anatomical evaluation, a contrast-enhanced multislice CT coronary angiography was performed under controlled sedation using paediatric acquisition protocols.

Discussion

ALCAPA remains one of the most clinically consequential congenital coronary anomalies due to its direct impact on myocardial perfusion. In early infancy, pulmonary arterial pressures are relatively high, permitting antegrade perfusion of the left coronary artery[4]. However, as pulmonary vascular resistance decreases after birth, perfusion pressure falls and blood preferentially flows from the left coronary system into the pulmonary artery. This reversal produces myocardial ischemia and establishes a left-to-right shunt.

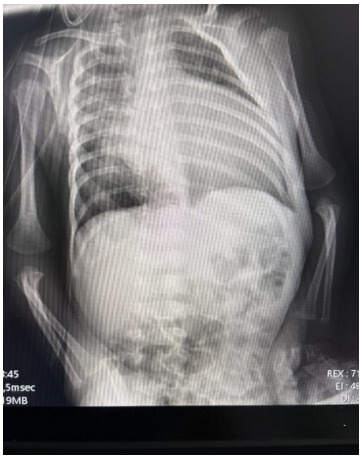

Figure 1: Frontal chest radiograph demonstrating cardiomegaly associated with pulmonary vascular congestion.

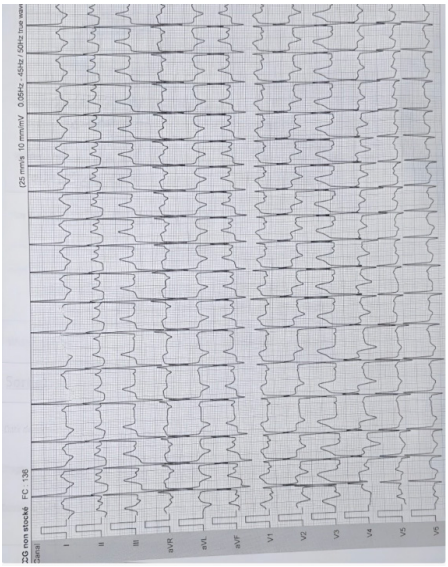

Figure 2: Electrocardiogram Showing Left Ventricular Hypertrophy and Anterolateral Ischemic Changes in a One-Year-Old Infant with ALCAPA.

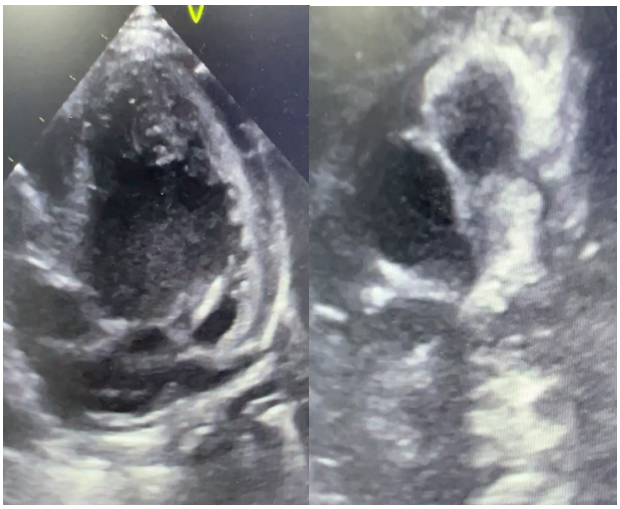

Figure 3: Transthoracic echocardiography demonstrating left ventricular dilatation, hyperechogenic mitral papillary muscle and anomalous origin of the left coronary artery arising from the pulmonary artery.

The clinical presentation in infancy typically reflects ischemic cardiomyopathy. Symptoms may include irritability, feeding difficulty, diaphoresis, tachypnoea and growth failure. Mitral regurgitation frequently develops secondary to papillary muscle dysfunction. Without intervention, mortality in the first year of life has historically exceeded 80-90 percent.

Survival beyond infancy depends on the extent of collateral circulation arising from the right coronary artery. In such cases, the RCA becomes enlarged and tortuous, as observed in our patient, while collateral vessels attempt to maintain myocardial perfusion[5,6]. Nevertheless, even in survivors, the risk of malignant arrhythmias, infarction and sudden death persists.

Multislice CT coronary angiography has emerged as a cornerstone imaging modality in this context. Its high spatial resolution allows precise delineation of coronary origins, collateral pathways and associated anomalies within a single non-invasive acquisition. Compared with conventional angiography, CT offers rapid imaging with excellent anatomical detail, which is particularly advantageous in paediatric populations.

Juxtaposition of the atrial appendages represents an additional congenital morphological variant. It is most commonly associated with transposition of the great arteries and other conotruncal defects. Its coexistence with ALCAPA is unusual but surgically relevant, as it may influence operative orientation and exposure.

Definitive management of ALCAPA is surgical. The preferred technique involves reimplantation of the anomalous left coronary artery into the aorta, thereby restoring a dual coronary system. Early surgical correction is associated with substantial recovery of ventricular function and favourable long-term prognosis[7].

Conclusions

Anomalous origin of the left coronary artery from the pulmonary artery is a rare but life-threatening congenital condition that requires a high index of suspicion, particularly in infants presenting with unexplained ventricular dysfunction or heart failure. Advanced imaging modalities, especially CT coronary angiography, enable accurate anatomical diagnosis and facilitate surgical planning. Early recognition and timely surgical repair remain the principal determinants of survival and myocardial recovery.

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