Giant Left Atrium in an Infant with Anomalous Origin of Left Coronary Artery from Pulmonary Artery

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Indian J Radiol Imaging 2025;35:343–345.

Abstract

Anomalous origin of the left coronary artery (ALCAPA) from the pulmonary artery is a rare congenital anomaly with coronary steal and myocardial ischemia. The left ventricular dilatation leads to mitral regurgitation causing left atrial enlargement. We report a rare case of giant left atrium in an infant with ALCAPA-mediated secondary mitral regurgitation, which has been hitherto unreported.

Keywords

- ALCAPA
- congenital heart disease
- giant left atrium

Introduction

Anomalous origin of the left coronary artery (ALCAPA) from the pulmonary artery is a rare congenital anomaly with coronary steal and myocardial ischemia. The left ventricular dilatation leads to mitral regurgitation (MR) causing left atrial enlargement. We report a rare case of giant left atrium (LA) in an infant with ALCAPA-mediated secondary MR, which has been hitherto unreported.

Case Report

A 5-month-old female infant presented to the pediatric outpatient clinic due to poor weight gain and recurring lower respiratory tract infections. Clinical signs suggested features of left ventricular dysfunction. Cardiomegaly with splayed carina was observed on chest radiography suggesting left atrial enlargement. A transthoracic echocardiography showed dilated left ventricle (LV) with hyperechoic papillary muscles and severe MR. Subsequently retrospective electrocardiogram (ECG) gated computed tomography angiography (CTA) was performed, and images were reconstructed at 43% (best systole) and 74% (best diastole) of the cardiac cycle. CTA done for anatomic delineation revealed (►**Fig. 1**) an ALCAPA, a giant LA (measuring 51 mm × 49 mm) and dilated LV (measuring 45 mm at the mid-cavity level in end diastole). The enlarged LA was seen splaying the inferior vena cava and right atrium confluence with anterior displacement of the right atrium (►**Fig. 2**). No other cardiovascular anomalies were detected. No significant intercoronary collaterals were seen.

Discussion

ALCAPA is a rare entity first described by Brooks in 1885 wherein the left coronary artery (LCA) arises from the pulmonary artery with an incidence of 1 in 300,000 births with varied hemodynamic manifestations.[1] Embryologically, it may be attributed to abnormal septation of the conotruncus into the aorta and pulmonary artery, or abnormal persistence of pulmonary arterial buds together with involution of aortic buds.[2] Other notable rare anomalous coronary artery origin entities from pulmonary arteries include anomalous origin of

article published online
October 24, 2024

DOI https://doi.org/10.1055/s-0044-1791747.
ISSN 0971-3026.

Thieme Medical and Scientific Publishers Pvt. Ltd., A-12, 2nd Floor, Sector 2, Noida-201301 UP, India

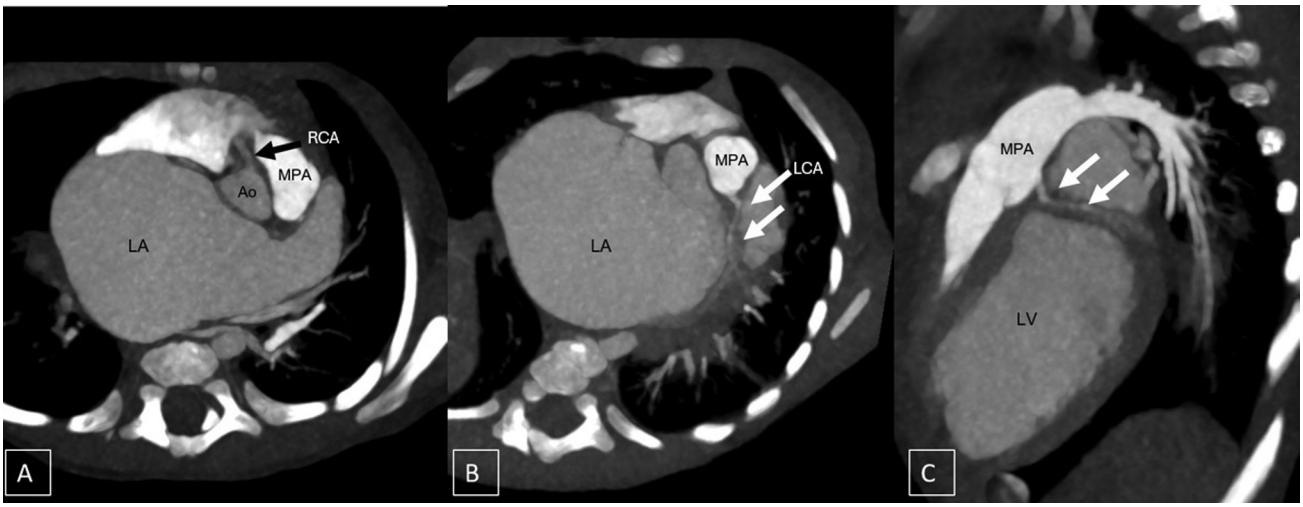

Fig. 1 (**A–C**) Computed tomography (CT) angiographic images show normal origin of right coronary artery (RCA; *black arrow*) with aorta (Ao) and anomalous origin of the left coronary artery (LCA; *white arrows*) from the main pulmonary artery (MPA). LA, left atrium; LV, left ventricle.

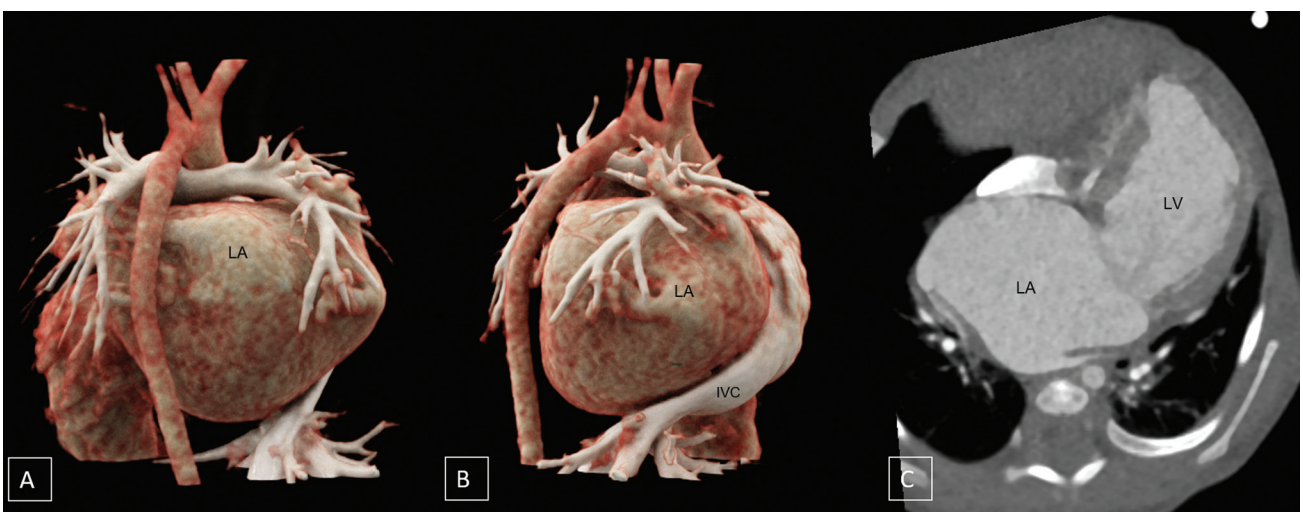

Fig. 2 (**A**, **B**) Cinematic rendered computed tomography (CT) images show giant left atrium (LA) with anterior displacement and splaying of the inferior vena cava (IVC) and right atrium (RA). (**C**) CT angiographic (CTA) image depicts the giant LA. LV, left ventricle.

the right coronary artery from the pulmonary artery (ARCAPA), anomalous origin of left circumflex artery from pulmonary artery (ACXAPA), and combined right coronary artery and LCA origin from the pulmonary artery. ALCAPA patients present with heart failure in early infantile period at around 3 to 4 months of age secondary to decrease in pulmonary arterial pressure and if untreated, around 90% patients die in first year of life. In fetal development, blood flow in the anomalous LCA primarily moves from the pulmonary artery to the myocardium. However, after the initial neonatal stage, the blood flow direction reverses, leading to left to right shunt. This shift occurs because pulmonary resistance decreases naturally, and collateral vessels develop gradually from the right coronary artery. Consequently, the LCA acts as a conduit, ultimately leading to gradual myocardial ischemia and dysfunction in the LV.[3] Ischemic insult to the LV leads to secondary MR due to papillary muscle ischemia and dilatation of the LV. Enlargement of the LA in cases of ALCAPA is attributed to MR and also to intrinsic properties of the LA wall.[4,5] Giant LA associated with ALCAPA has been previously reported once in a 5-year-old boy.[6] However, to the best of our knowledge, it has not been described in an infantile age group. Other differential diagnosis of giant LA in infantile age group is idiopathic congenital giant LA, rheumatic heart disease, left to right shunt lesions, and chronic atrial fibrillation. Our case is unique as it describes early complications of ALCAPA in an infant. Definite treatment for this condition is surgical intervention to establish a two-coronary system. In surgical approaches, direct reimplantation of the coronary artery into the aorta, also known as coronary button transfer, is used to correct ALCAPA. Other techniques include transpulmonary baffling, or the Takeuchi procedure, which helps in rerouting blood flow to the coronary arteries. The subclavian–left coronary anastomosis, which involves connecting the subclavian artery to the LCA to improve blood flow, can also be performed. Additionally, coronary artery bypass grafting can be performed using a saphenous vein graft or the left internal mammary artery, often with the ligation of the anomalous LCA to restore proper circulation. Cardiac

transplantation remains an option for patients who experience severe left ventricular dysfunction and persistent heart failure, providing a last-resort solution for those not responding to other treatments.[7] Our case underwent mitral valve repair with concomitant coronary button transfer with reimplantation of the LCA to the aorta.

Magnetic resonance imaging (MRI) is a robust modality in evaluating patients with ALCAPA with the added advantage of no exposure to ionizing radiation. MRI is particularly helpful in anatomic delineation, functional cardiac assessment, myocardial characterization, and flow assessment. Using CINE (cinematic) steady-state free precession (SSFP) sequences, left ventricular function, mitral insufficiency, or MR, regional wall motion abnormalities can be detected and quantified. First pass perfusion images can detect areas of perfusion deficit, and late gadolinium enhancement (LGE) images can detect areas of myocardial fibrosis secondary to infarct prompting the need for early surgical correction to subdue risk of malignant arrhythmias. Advanced flow quantification sequences using phase contrast imaging can be useful to quantity the degree of left to right shunt. In cases of ALCAPA, it is imperative to look for coronary vascular origins, presence or absence of inter coronary collaterals, any hypertrophied coronary artery segments, features of left ventricular dysfunction, and presence of myocardial fibrosis.[8] In the postoperative period, serial echocardiography is indicated for left ventricular functional assessment, and cross-sectional imaging such as CTA or MRI can be helpful in evaluating reverse left ventricular remodeling and regression of intracoronary collateral network.

This case underscores the importance of CTA in defining all the morphologic aspects of ALCAPA that may be useful in planning surgical repair.

Funding
None.

Conflict of Interest
None declared.

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