CASE REPORT

# ADULT ALCAPA: CASE REPORT WITH RADIOLOGICAL PERSPECTIVE

Abdullah Bin Imaad, Imaad ur Rehman, Hassan Saleem, Ather Mehmood, Saria Ahmed Rajpoot

Department of Radiology, Islamabad Diagnostic Centre, Islamabad, Pakistan.

PJR July - September 2022; 32(3): 160-164

## ABSTRACT

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a very rare and life-threatening congenital anomaly. In the past ALCAPA was diagnosed by angiography or autopsy; however, the development of computed tomography cardiac angiography (CTCA) has allowed noninvasive evaluation of the coronary anatomy by direct visualization of the origin of the left coronary artery (LCA) from the pulmonary artery and is now considered the preferred modality in the diagnosis of ALCAPA.[1] We report a case of adult type ALCAPA who presented with history of exertional chest pain and elevated blood pressure with no significant symptoms prior to presentation. Diagnosis was confirmed by CTCA.

**Keywords:** ALCAPA, Anomalous origin of the left coronary artery from the pulmonary artery, congenital anomaly, computed tomography cardiac angiography, CTCA.

## Introduction

Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a very rare congenital abnormality where the left main coronary artery emerges from the pulmonary artery, instead of the coronary sinus of the ascending aorta and accounts for only 0.25-0.5% of all congenital cardiac anomalies. ALCAPA is divided into two types; adult and infant. Infants experience myocardial infarction and congestive heart failure, and approximately 90% die within the 1st year of life. Rarely, ALCAPA syndrome manifests in adults. Lethal complications such as malignant arrhythmia and sudden death can arise in adult type of ALCAPA.[2] Imaging is the current preferred procedure for diagnosis of ALCAPA as clinically it poses a challenge due to its ill-defined clinical presentation and variable laboratory findings. The definitive diagnosis of ALCAPA is reached by computed tomography cardiac angiography (CTCA). We report a case of adult type ALCAPA who was diagnosed accurately with the help of CTCA.

**Correspondence :** Dr. Abdullah Bin Imaad
Department of Radiology,
Islamabad Diagnostic Centre,
Islamabad, Pakistan.
Email: imaadur@gmail.com

Submitted 10 August 2022, Accepted 27 August 2022

## Case Presentation

A 32-year-old lady presented with history of sudden onset of exertional chest pain and elevated blood pressure for one week. She faced difficulty in completing her daily activities due to reduced effort tolerance. She had past medical history of asthma and past surgical history of appendectomy twelve years back. Her childhood history was unremarkable.

After initial work up, conventional coronary angiography was performed which showed right coronary artery (RCA) arising from right coronary sinus. Left coronary artery (LCA) was not opacified from left coronary sinus despite multiple attempts. RCA appeared dilated and tortuous with multiple collaterals retrogradely filling LCA. Origin of LCA could not be ascertained on angiography. (Fig.1). Suspicion of ALCAPA was raised and further workup by CTCA was advised.

CTCA was performed using a 128 slice CT scanner (Toshiba Aquilion). Scan parameters were as follows: slice collimation, 200 mm; rotation time, 0.35 sec; tube

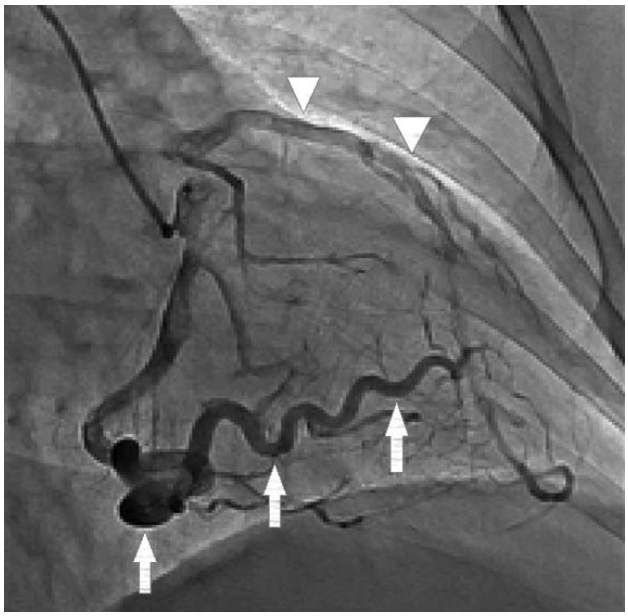

**Figure 1: Adult ALCAPA Angiographic findings.** Right coronary artery is dilated and tortuous (Arrows) with a large number of collaterals feeding the left coronary system with visualization of LCA (Arrow heads).

voltage, 40 kV; tube current, 500 mA; and pitch, 0.35. The scan parameters were modified to decrease the patient dose. The heart rate was kept around 60 bpm during the scan. The patient received a b-blocker prior to the CT examination for heart rate regulation. A pre-scan was taken at the level of the aortic root and a region of interest (ROI) was placed on the ascending aorta. CT angiography was triggered automatically by the arrival of the main contrast bolus (automatic bolus tracking) in ascending aorta. As soon as the density level in the ascending aorta reached the predefined threshold of 180 Hounsfield units (HU), the scan started. We injected 70 ml nonionic contrast medium (ultravist) at a flow rate of 5 ml/s in the right antecubital vein. This was followed by a 45 ml saline chaser bolus at a flow rate of 4 ml/s to wash out contrast from the right ventricle.

During the scan, the ECG was recorded simultaneously. The continuous data acquisition with retrospective gating allows slice reconstruction at different time points in the cardiac cycle. The reconstruction intervals for both right and left coronary arteries with the fewest motion artifacts were determined and used for further analysis. For reconstruction of axial images, we used helical thickness of 0.5 mm.

CTCA showed an anomalous origin of LCA from the pulmonary trunk (Fig.2). The relatively small sized LCA bifurcates into left anterior descending (LAD) and left circumflex arteries (LCX). Tortuous and dilated right coronary artery is seen arising from right coronary

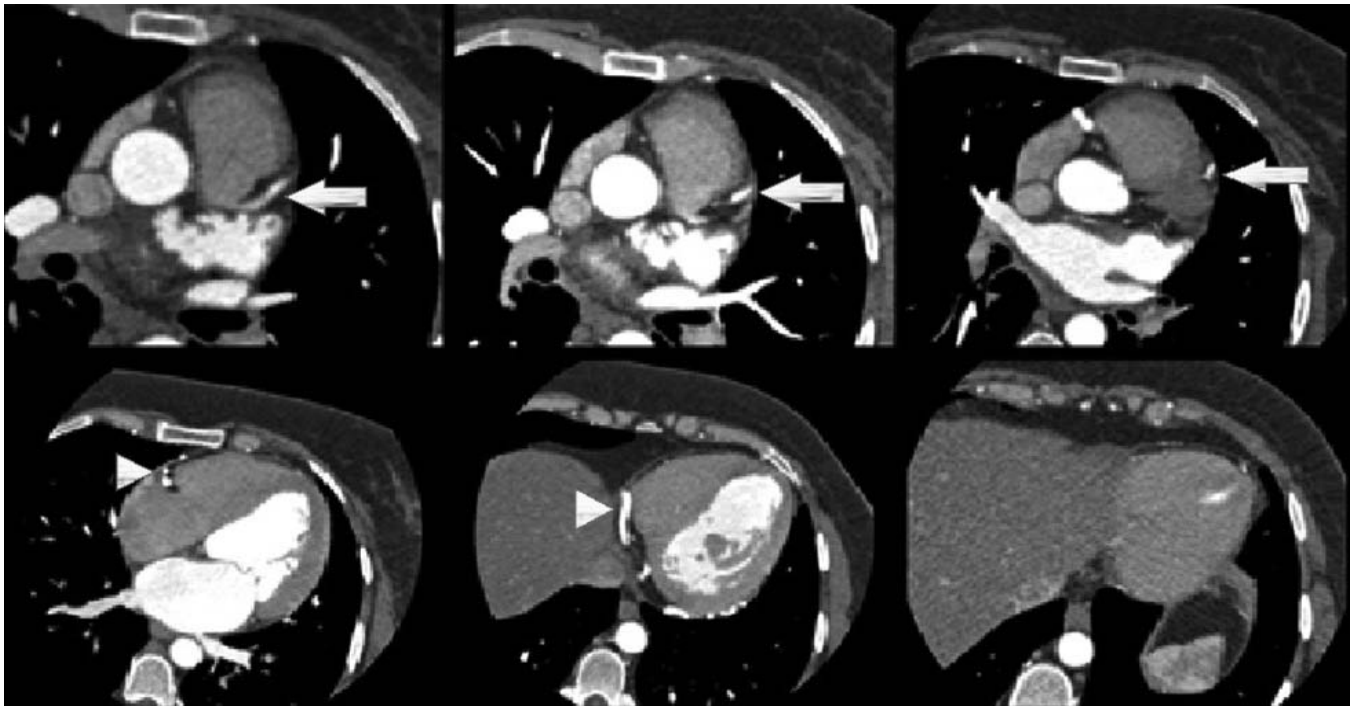

**Figure 2: Adult ALCAPA CTCA findings.** Left coronary artery is emerging from pulmonary trunk (Arrows). Right coronary artery is dilated with collaterals (Arrow heads)

sinus of aorta giving branches including acute marginal branch (AM) and Posterior descending artery (PDA) (Fig.3). Right-to-left collateralization with multiple collaterals seen arising from RCA (Fig. 4). No significant atherosclerosis or arterial stenosis were noted. Findings were compatible with adult ALCAPA.

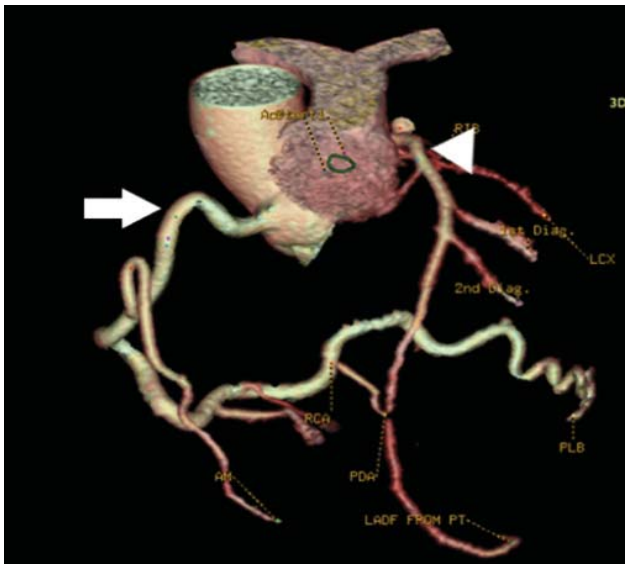


**Figure 3: CT angiography volume rendered reformatted image showing vessels only.** Relatively small sized Left coronary artery is emerging from pulmonary artery trunk (Arrow head) with its branches namely LAD and LCX. Right coronary artery is dilated and tortuous arising from right coronary sinus (Arrow) with its branches namely AM and PDA.

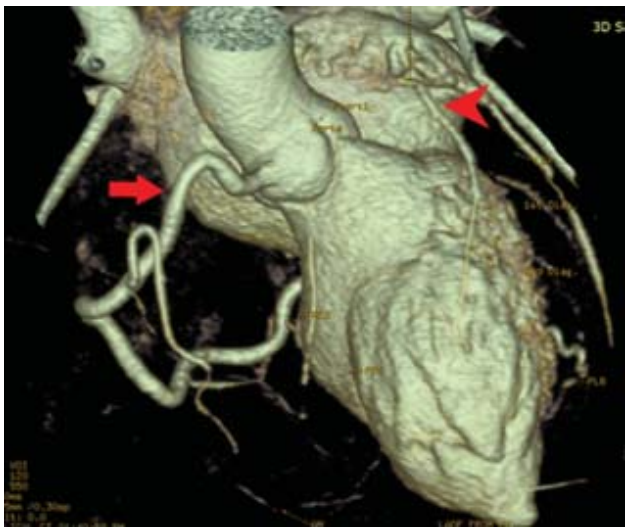

**Figure 4: CT angiography volume rendered image.** Right coronary artery is dilated arising from right coronary sinus (Arrow) with collateral branches. Relatively small sized Left coronary artery is emerging from pulmonary artery trunk (Arrow head).

She remained well and asymptomatic after imaging. No further cardiovascular imaging was planned. Surgical options and prognosis of disease, with and without surgery, was explained to the patient. Her decision is awaited regarding surgery while she is being managed conservatively on medications.

## Discussion

ALCAPA syndrome is a very rare congenital anomaly where the left coronary artery arises from the pulmonary artery instead of the aortic sinus. This leads to a left-to-right shunt, resulting in impaired coronary arterial supply. First case of ALCAPA with complete clinical description combined with autopsy findings was described by Bland, White and Garland in 1933. Hence, ALCAPA is also called Bland-White-Garland syndrome. This anomaly accounts for 1 in every 300,000 births.[3] There are two types of ALCAPA, the infant type and adult type, each of which has different manifestations and outcomes. Approximately 90% die within the first year of life in infant type due to myocardial infarction and congestive heart failure. In adult type, patients may be asymptomatic for years, as was our case, however if this anomaly is not corrected, patients may develop progressive cardiac dysfunction, malignant arrhythmia, myocardial infarction and sudden death. This has an estimated incidence rate up to 90 percent at the mean age of 35 years.[4]

Conventional coronary angiography used to be the gold standard to detect coronary anomalies until the development of the CTCA and magnetic resonance imaging (MRI). Nowadays, different imaging techniques have changed the daily practice to diagnose coronary anomalies. Conventional angiography is invasive and associated with a low but well-known risk of complications. Also, the precise course and the exact anatomy of the anomalous vessel may be difficult to delineate due to its complex geometry and origin of LCA cannot be directly visualized as seen in our case (Fig.1).

Technological advancements have substantially increased the detection of this abnormality in adults. CTCA and MRI are the current preferred modalities for those with suspected ALCAPA, as well as for pre-operative planning and post-operative follow-up. CTCA and MR imaging are valuable noninvasive modalities

that identify and define anomalous coronary arteries and their course with a very high accuracy. Hence the identification of this condition has increased and is resulting in major changes in the management and survival of ALCAPA patients.[5]

CTCA offers excellent contiguous resolution and volume rendering techniques to visualize the coronary arteries. CTCA is a non-invasive imaging technique which is fast and offers excellent spatial resolution, which is required to assess small vessels such as the coronary arteries. The short investigation time, relative non-invasiveness of the procedure, simple preparation and minimal aftercare make CTCA advantageous over conventional coronary angiography. The main disadvantages of CTCA are its relatively high radiation dose and its inability to assess flow.[5] The hallmark radiographic features in CTCA for ALCAPA include direct visualization of the origin of left coronary artery from the posterior aspect of PA, dilated and tortuous RCA, and dilated intercoronary collateral arteries along the external surface of the heart. All these features were seen in our case (Fig.2, 3 and 4).

MRI has the advantage of physiological and functional analysis in cardiac aberrations. In ALCAPA it can be used to assess the systolic function of the left ventricle, visualization of the communication and the retrograde flow from the left coronary artery to the PA and myocardial viability. MR imaging does not use ionizing radiation. But the main disadvantages of MR imaging in comparison with CT are its relatively long examination times and its low spatial resolution.[6]

The treatment of choice for this anomaly is the Takeuchi procedure, i.e. direct anastomosis of the left coronary artery from the pulmonary artery directly to the aorta to restore a dual-coronary-artery system which includes left coronary artery ligation with or without coronary artery bypass transplant. Another procedure that has been described recently with high success rate is occlusion of the ALCAPA using ventricular septal defect occluder and detachable patent ductus arteriosus coil.[7] Reaching the correct diagnosis of ALCAPA provides the patient a surgical treatment option with a potential promise of good prognosis.

In our case, the surgical options were explained to the patient. Her decision is awaited regarding surgery while she is being managed conservatively on medications.

## Conclusion

ALCAPA is a rare and life-threatening condition. Early diagnosis is very important because surgical treatment with optimal timing generally results in good prognosis. CTCA provides correct diagnosis with remarkable anatomical detail non-invasively with short investigation time and minimal aftercare. Therefore, it can be considered as the modality of choice in the diagnostic work up of ALCAPA.

**Conflict of interest:** None.

**Ethical Approval:** Approval of case report was taken from Ethical committee.

## References

1. Tan YE, Chia KK, Abdul Karim NK. Diagnosis of adult ALCAPA with computed tomography coronary artery. Egyptian Journal of Radiology and Nuclear Medicine. 2021; **52(290)**.

2. Hanna A, Torigian DA, Ferrari VA et-al. Anomalous origin of the left coronary artery from the pulmonary artery in adulthood on CT and MRI. AJR Am J Roentgenol. 2005; **185(2):** 326-9.

3. Hauser M. Congenital anomalies of the coronary arteries. Heart. 2005; **91:** 1240-5.

4. Zacharias M, Chandok D, Tighe D. A late presentation of an anomalous left coronary artery originating from the pulmonary artery (ALCAPA): a case study and review of the literature. J Cardiol Cases. 2015; **20:** 234-39

5. Doris MK, Newby DE. How should CT coronary angiography be integrated into the management of patients with chest pain and how does this affect outcomes? Eur Heart J Eur Heart J Quality Care Clin Outcomes. 2016; **2(2):** 72-80.

6. Yau JM, Singh R, Halpern EJ, Fischman D. Anomalous origin of the left coronary artery from the pulmonary artery in adults: a comprehensive review of 151 adult cases and a new diagnosis in a 53-year-old woman. Clin Cardiol. 2011; **19:** 198-201.

7. Sagar P, Sivakumar K. Transcatheter occlusion of an anomalous origin of left coronary artery from pulmonary artery in an adult as an alternative to surgery. Cardiol Young. 2021; **19(2):** 444-8.