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Case Report

Anomalous origin of left coronary artery from pulmonary artery (ALCAPA): A case report ☆,☆☆

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ABSTRACT

Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) also known as Bland-White-Garland Syndrome is a rare anomaly of coronary arteries comprising of 0.25%-0.5% of all congenital heart defects with a prevalence of 1 in every 300,000 live births. Its clinical significance lies in the possibility of resultant coronary steal phenomenon with a left-to-right shunt causing aberrant left ventricular perfusion which may ultimately lead to myocardial ischemia and infarction in children having the abnormality. ALCAPA may manifest as an isolated defect but in 5% of cases it may be associated with other cardiac anomalies such as atrial septal defect, ventricular septal defect, and aortic coarctation. We present a case of 7 years female with ALCAPA with collaterals between RCA and LCA and additional findings of juxtaposition of left atrial appendage. Juxtaposition of atrial appendage is associated with some major congenital heart diseases, transposition of great vessels being the common one. In our case, however, juxtaposition of left atrial appendage is associated with ALCAPA. Surgery is the definite treatment modality for ALCAPA available till date. Early diagnosis of ALCAPA with the help of multislice CT angiography is always good for the patient to prevent the possible grave consequences.

Introduction

Anomalies of coronary arteries can be in their origin, course or termination. Among different types of anomalous origin, anomalous origin of left coronary artery from pulmonary artery (ALCAPA) also known as Bland-White-Garland Syndrome is a rare type comprising of 0.25%-0.5% of all congenital heart defects with a prevalence of 1 in every 300,000 live births [1]. The clinical significance of this anatomic anomaly is that it may result in coronary steal phenomenon with a left-to-right shunt causing aberrant left ventricular perfusion which may ultimately lead to myocardial ischemia and infarction in children having the abnormality. Some studies have reported that up to 90% of patients with ALCAPA syndrome die during the first year of life if they are not treated [2]. The severity of myocardial ischemia and the severity of cardiovascular sequelae are determined by the development of collateral vessels between the right and left coronary arteries (RCA and LCA, re-

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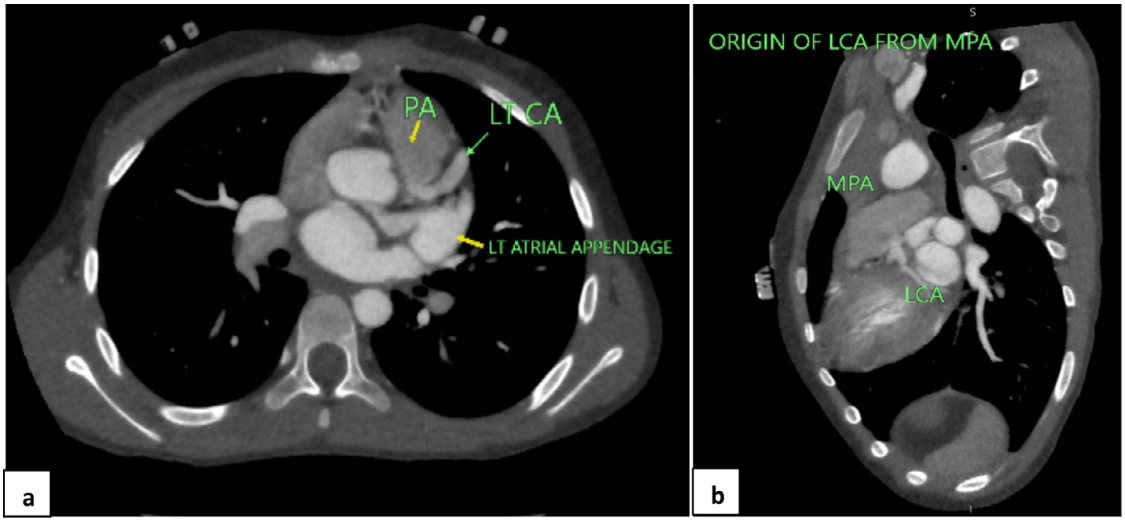

Fig. 1 – "a" and "b" are the axial and sagittal angiographic images of chest showing anomalous origin of left coronary artery from pulmonary trunk.

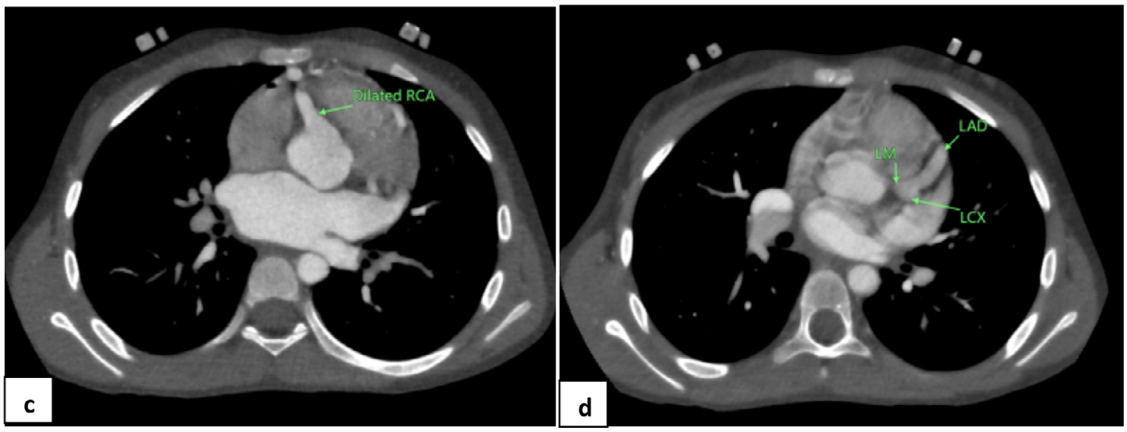

Fig. 2 – "c" and "d" are the axial angiographic images of chest showing enormously dilated, tortuous and dominant right coronary artery (RCA) emerging from the right aortic sinus on "c" and the left anterior descending artery (LAD) and left circumflex artery (LCX) branching from LMA on "d."

spectively). ALCAPA may manifest as an isolated defect but in 5% of cases it may be associated with other cardiac anomalies such as atrial septal defect, ventricular septal defect, and aortic coarctation [1]. We present a case of 7 years female with ALCAPA with collaterals between RCA and LCA and additional findings of juxtaposition of left atrial appendage.

Clinical history

A 7-year-old girl was referred to our hospital from a nearby hospital where she presented in the emergency department with complaints of chest discomfort and shortness of breath while playing outdoors. On work-up, she was found to have para-sternal systolic murmur on auscultation with cardiomegaly and signs of pulmonary congestion on chest x-ray. On echo, dilatation of left ventricle, mild mitral and tricuspid insufficiency and decreased ejection fraction was appreciated for which she was referred to our center for further evaluation of cardiac and coronary anomalies. A 640 slice CT scanner was used to perform coronary CT angiogram following standard imaging protocols.

Imaging findings

Anomalous origin of left coronary artery from pulmonary artery was noted (Fig. 1a and b). Right coronary artery was dominant vessel arising from right coronary sinus and was enormously dilated and tortuous in its course (Fig. 2c). The left anterior descending artery (LAD) and left circumflex

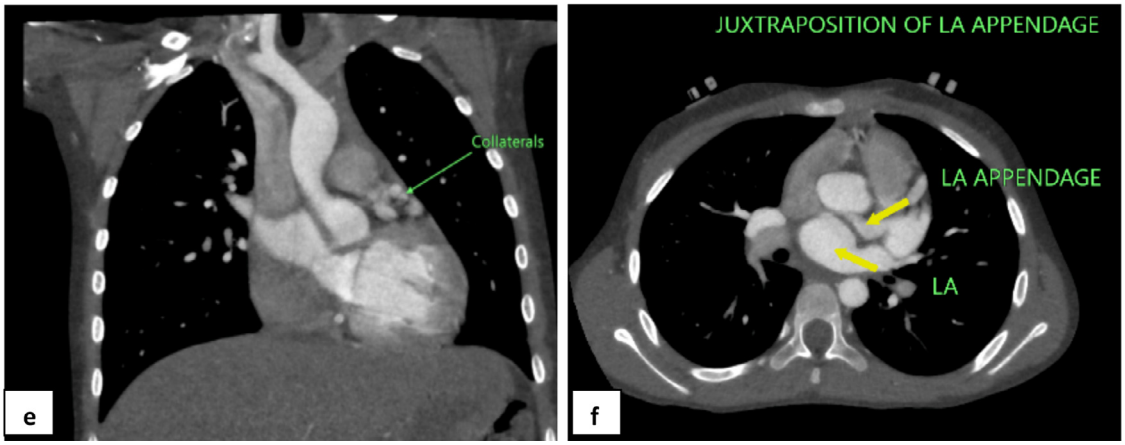

Fig. 3 – "e" is a coronal angiographic image of chest showing large number of collateral vessels feeding the left coronary system, which showed enlarged and tortuous vessels. Figure "f" is and axial angiographic image showing left atrial appendage lying in between ascending aorta and left atrium suggesting its juxtaposition.

artery (LCX) were originating from left main artery (LMA) (Fig. 2d). A number of collateral vessels were noted feeding the left coronary system (Fig. 3e). Left atrial appendage was seen to lie in between ascending aorta and left atrium suggesting its juxtaposition (Fig. 3f).

Discussion

Origin of the left coronary artery from the pulmonary artery, or Bland-White-Garland Syndrome, is a rare frequently lethal anomaly seen in children and adults [1,3]. Left-to-right shunting from the higher pressure left coronary arterial system to the lower pressure pulmonary arterial system leading to circulatory insufficiency, myocardial infarction, or life-threatening cardiac dysrhythmias is the cause of death in infants in most of the cases [1,4]. Development of abundant intercoronary collateral arteries may facilitate survival beyond infancy [5]. Multislice CT angiography is an important noninvasive imaging technology with outstanding spatial resolution for diagnosing abnormal coronary arteries and their course. Juxtaposition of atrial appendage is associated with some major congenital heart diseases, transposition of great vessels being the common one [6]. In our case, however, juxtaposition of left atrial appendage is associated with ALCAPA. Surgery is the definite treatment modality for ALCAPA available till date [7]. Early diagnosis of ALCAPA is always good for the patient to prevent the possible grave consequences.

Conclusion

ALCAPA is an uncommon and potentially fatal illness. Early detection is critical since surgical treatment delivered at the right time usually results in a positive outcome. Multislice CT angiography provides anatomical information and reliable diagnosis in a noninvasive way with minimal investigation time. As a result, it can be considered the modality of choice in the diagnosis of ALCAPA and other congenital heart diseases with no need of other invasive diagnostic procedures and radiation exposure. Also it is always important to look for other co-existing congenital conditions of heart in patients with ALCAPA.

Patient consent

Date of consent taken: April 27, 2022
Taken by: The author
Taken from: Father of the child
Informed written consent has been taken from the parent of the child by myself, the principal author and can be reproduced as and when required.

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