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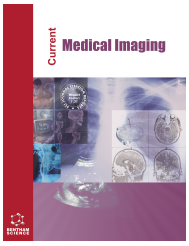

CASE REPORT

Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery Diagnosed in Adulthood

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Abstract:

Introduction:

An anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare heart malformation, with 90% of patients dying during the first year of life. If the right coronary artery compensation and multiple collateral circulation are sufficiently established, the patient's myocardial ischemia symptoms are mild and appear later, which is called the adult type ALCAPA.

Case Description:

A 42-year-old woman presented to our hospital with one-month history of the aggravation of active shortness of breath which gradually progressed to nocturnal paroxysmal shortness of breath and cough. Admission physical examination suggested mild edema of both lower limbs. Transthoracic echocardiography (TTE) showed that a small vessel shadow was abnormally connected to the pulmonary artery (PA), and moderate pulmonary artery hypertension. Coronary computed tomography angiography (CTA) showed an anomalous origin of the left main coronary artery (LMCA) dividing into the left anterior descending (LAD) and left circumflex (LCX) artery from the PA, with no clear connection to the left coronary sinus. The right coronary artery (RCA) was significantly dilated and originated from the normal Valsalva sinus. It was accompanied by multiple collateral circulations, most of which traveled anterior to the right ventricular free wall and anterior interventricular sulcus, and some emanated from the posterior descending branch of the posterior interventricular sulcus and walked toward the posterolateral wall of the left ventricle.

Conclusion:

Coronary computed tomography angiography (CTA) can be used to visualize the abnormal origin and distribution of the coronary artery's course and may be the first choice in the diagnosis of ALCAPA.

Keywords: An anomalous left coronary artery from the pulmonary artery, Computed tomography angiography, Left coronary artery, Pulmonary artery, Coronary, Adulthood.

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1. INTRODUCTION

An anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital coronary anomaly. If left untreated, approximately 90% of ALCAPA patients will die at the age of 1-2 years [1 - 3]. It is extremely rare for this abnormality to be diagnosed in adulthood [4, 5]. If the collateral circulation is sufficiently established, the patient's myocardial ischemia symptoms are mild and appear later, which is called the adult type. Only a few adult-type ALCAPA cases have been reported in the literature. Here, we report a case of ALCAPA diagnosed in adulthood. This manuscript is written following the CARE checklist.

2. CASE DESCRIPTION

A 42-year-old woman presented to our hospital with one-month history of the aggravation of active shortness of breath which gradually progressed to nocturnal paroxysmal shortness of breath and cough. Her symptoms eased after sitting or lying on the right side. Admission physical examination suggested

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mild edema of both lower limbs. Her blood pressure was 162/96 mmHg. Admission laboratory examination showed N-terminal pro-brain natriuretic peptide (NT-proBNP) was 3125 pg/mL. ECG showed sinus tachycardia (106 b.p.m), with an incomplete right bundle branch block and a left anterior fascicular block. Transthoracic echocardiography (TTE) (Fig. **1**) showed an enlarged left atrium and left ventricle. Mild mitral and tricuspid regurgitation was also observed. Left ventricular ejection fraction (EF) was 60%, and no wall motion abnormalities were observed. The beginning of the left coronary artery was poorly visualized, with a small vessel shadow abnormally connected to the pulmonary artery (PA). Also, moderate pulmonary hypertension was demonstrated. Color Doppler flow imaging (CDFI) showed retrograde flow into the inner wall of the PA and then to the pulmonary valve. Coronary computed tomography angiography (CTA) showed an anomalous origin of the left main coronary artery (LMCA) dividing into the left anterior descending (LAD) and left circumflex (LCX) artery from the PA, with no clear connection to the left coronary sinus, and accompanied by abundant collaterals (Fig. **2A** and **B**). The LAD extended downward in the anterior interventricular sulcus to the apex, supplying the anterior wall, the apical portion, and most of the ventricular septum. The LCX traveled posteriorly in the left atrioventricular sulcus, supplying the left ventricular wall. The right coronary artery (RCA) was significantly dilated, originating from the normal Valsalva sinus. It was accompanied by multiple collateral circulations, most of which traveled anterior to the right ventricular free wall and anterior interventricular sulcus, and some emanated from the posterior descending branch of the posterior interventricular sulcus and walked towards the posterolateral wall of the left ventricle. The patient had heart failure with preserved EF treated with furosemide and spironolactone, sacubitril valsartan sodium, and metoprolol. The patient's pulmonary artery pressure was reduced to mild pulmonary hypertension by using sildenafil citrate. After anti-heart failure treatment, the patient's NT-proBNP decreased to 112 pg/mL, the symptoms were effectively relieved, and the condition was stabilized and well controlled, coupled with the patient's wishes not to implement surgical treatment in our hospital. Two months later, the patient underwent surgery in an external hospital due to aggravating symptoms. However, she did not know the detailed surgery process. During the subsequent two years' follow-up, the patient has recovered well.

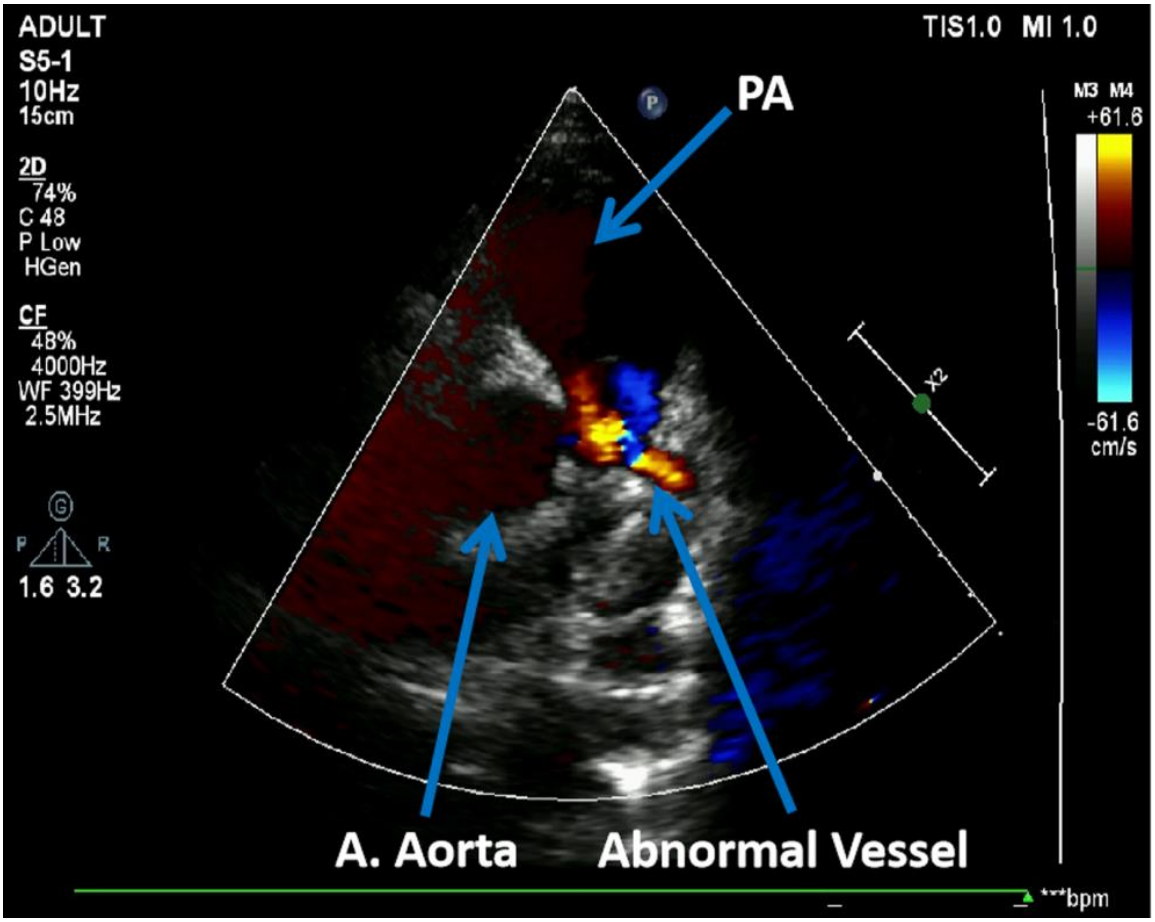

Fig. (1). Transthoracic echocardiography (TTE) shows an abnormal small vessel shadow connecting to the pulmonary artery (PA), while color Doppler flow imaging (CDFI) shows retrograde flow into the PA.

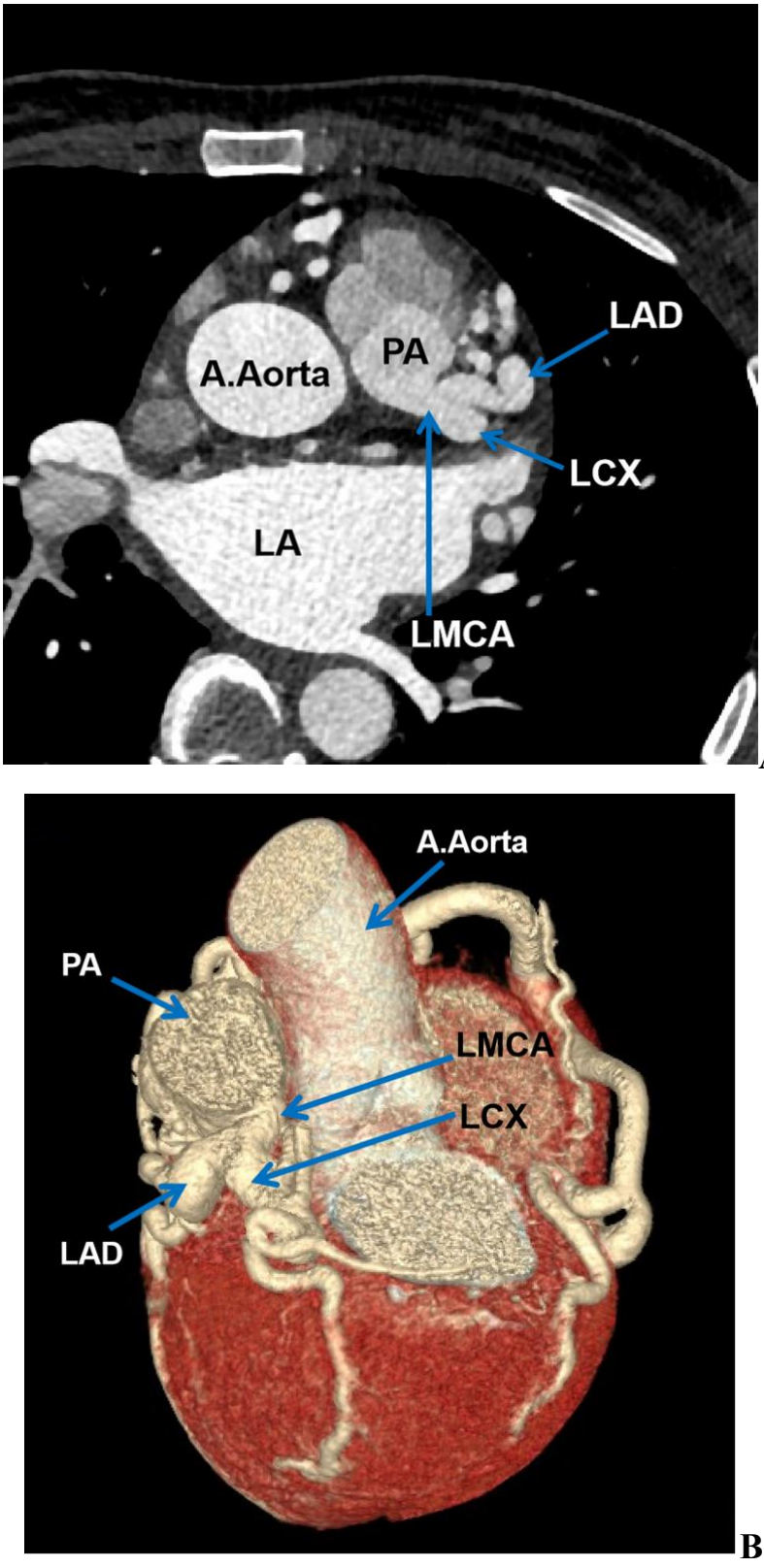

Fig. (2). Images from coronary CT angiography in a 42-year-old woman who presented with shortness of breath. Axial contrast-enhanced CT (**A**) and Cinematic rendering (**B**) images show an anomalous origin of the left anterior descending and left circumflex arteries through the left main coronary artery from the pulmonary artery (blue arrows). A. Aorta = ascending aorta; LA = left atrium; LAD = left anterior descending artery; LCX = left circumflex artery; LMCA= left main coronary artery; PA = pulmonary artery.

3. DISCUSSION

ALCAPA is an incidental finding and is an extremely rare heart malformation, with approximately 90% of patients dying during the first year of life [1 - 3]. Most patients become symptomatic in infancy or early childhood, which is called the infantile type. If the collateral circulation is adequately established and the blood from the RCA reaches the LCA through the collateral circulation and returns to the PA, the patient's myocardial ischemia symptoms are mild and appear later, which is called the adult type. The RCA supplies blood to the LCA through a large number of collateral circuits, so the patient's myocardial ischemic symptoms are somewhat relieved. The patient in our study was the adult type. Both right and left coronary arteries may originate anomalously from the pulmonary artery, while LAD is more common, with an incidence of 0.008% [6, 7].

The prognosis for adult-type ALCAPA is reported to be poor, especially after the age of 40 years [8 - 11]. According to the literature, the fundamental treatment modality is surgical correction of the anomalous coronary artery [8 - 10]. However, in adults with ALCAPA, patients without surgery can survive if the condition results in mild chronic ischemia.

In the present case, we observed essentially identical CT values in the lumen of the abnormal origin of the LCA with that of adjacent PA, while the CT values distal to the PA were lower. This could also indirectly indicate altered hemodynamics, suggesting a reverse flow of blood from the high-resistance coronary system into the low-resistance pulmonary artery. This was confirmed by color Doppler ultrasound.

The diagnosis of ALCAPA needs to be differentiated from left coronary-pulmonary artery fistula (CPAF), which is the normal origin of the affected coronary artery and the abnormal connection is located distal to the origin of the coronary artery. The abnormal origin of LCA cannot be accurately demonstrated on ultrasound. However, coronary CTA allows for a comprehensive assessment. Coronary CTA can be used to visualize the abnormal origin and distribution of the coronary artery's course and can even trace the area of the collateral circulation supplying the artery. It is considered the first-line imaging modality for the assessment of anomalous coronary anatomy [12 - 15].

CONCLUSION

In conclusion, our report describes a rare case of adult-type ALCAPA. Congenital anatomical anomalies revealed by coronary CTA should not be neglected. The earlier they are detected, the more timely that surgical correction and prevention of potential complications can be performed. This will result in a better outcome for ALCAPA patients. Coronary CTA can be the first choice in the diagnosis of ALCAPA.

LIST OF ABBREVIATIONS

ALCAPA	=	Anomalous Left Coronary Artery From the Pulmonary Artery
CDFI	=	Color Doppler Flow Imaging
CPAF	=	Coronary-Pulmonary Artery Fistula
CTA	=	Computed Tomography Angiography
LAD	=	Left Anterior Descending
LCX	=	Left Circumflex
PA	=	Pulmonary Artery
RCA	=	Right Coronary Artery
TTE	=	Transthoracic Echocardiography

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Not applicable

HUMAN AND ANIMAL RIGHTS

Not applicable

CONSENT FOR PUBLICATION

The patient's consent has been obtained.

STANDARDS OF REPORTING

CARE guidelines were followed.

AVAILABILITY OF DATA AND MATERIALS

The data and supportive information are available from the corresponding authors, [W.M] on request.

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CONFLICT OF INTEREST

The authors declared that they have no potential conflicts of interest to disclose.

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Declared none.

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