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ANGINA PECTORIS AS A MANIFESTATION OF ALCAPA SYNDROME IN A 20-YEAR-OLD FEMALE: A CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Anomalous left coronary artery from the pulmonary artery (ALCAPA) is considered a rare congenital heart disease where the take-off of the left coronary artery abnormally originates from the pulmonary artery instead of left aortic sinus. It is associated with a high mortality rate in the first year of life and sudden death in adults if left untreated. We report an adult form of ALCAPA syndrome in a 20-year-old female who presented with anginal pain for the previous few months. Unfortunately, the patient was hesitant to have surgery at the time.

KEYWORDS

ALCAPA, echocardiography, angina pectoris

LEARNING POINTS

- The abnormal origin of the left coronary artery from the pulmonary artery (ALCAPA) is rare and potentially fatal coronary congenital disease, accounting for 0.5% of all congenital heart diseases; it is associated with poor outcomes if left untreated.
- ALCAPA is classified into infantile and adult forms. The prevalence of adult individuals with ALCAPA syndrome has significantly increased as a result of recent developments in non-invasive cardiac imaging.
- The prevalence of sudden mortality in childhood and the early stages of adulthood makes surgery the preferred treatment, and coronary reimplantation surgery is considered the surgical procedure of choice.

INTRODUCTION

ALCAPA is a rare coronary anomaly, occurring in 1 in 300,000 live births, and is classified into infantile and adult forms depending on age, type of symptoms and the development of intercoronary collaterals at presentation. If sufficient coronary collaterals between the right and left coronary arteries are present, then the clinical presentation of this abnormality may be delayed until adulthood or even late adulthood. Transthoracic echocardiography in our case raised suspicion of a coronary anomaly, and non-invasive imaging confirmed the diagnosis.

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CASE DESCRIPTION

A 20-year-old female with no significant past medical history presented with episodes of typical anginal pain for a few months' duration. She has no family history of significant heart disease. She denied using recreational drugs, does not smoke and has no known allergies. Her clinical exam was notable for loud heart sounds and a persistent loud murmur that peaked at the left parasternal border. Her vital signs were normal, and the rest of the examination was unremarkable. Her body surface area was 1.47 m^2. Her electrocardiogram showed sinus rhythm with non-specific T-wave abnormality in lateral leads *(Fig. 1)*. Transthoracic echocardiography revealed a dilated right coronary artery (RCA) of 8 mm with a mildly dilated left ventricle (LV) and preserved systolic function *(Fig. 2A, clip 1)*. On the short axis view the colour Doppler showed a turbulent flow within the interventricular septum, which raised the possibility of intercoronary collateral between right and left coronary arteries *(Fig. 2B, clip 2)*. On the parasternal right ventricle outflow view, a take-off of anomalous left coronary artery was noted from the posteromedial aspect of the main pulmonary artery (MPA) *(Fig. 3C, clip 3)*. A colour Doppler showed a flow passage from the left coronary artery (LCA) into the pulmonary artery indicating a left–to–right shunt *(Fig. 4D, clip 4)*. The echocardiographic picture was highly suggestive of ALCAPA syndrome. The patient underwent a multislice computed tomographic angiography (CTA) *(Fig. 3)*. The CTA with three-dimensional reconstruction showed a dilated RCA that originates from the right aortic sinus and anomalous drainage of the LCA to the posteromedial aspect of the MPA *(Fig. 4A)*. The three-dimensional reconstructed images clearly exposed the extensive intercoronary collaterals

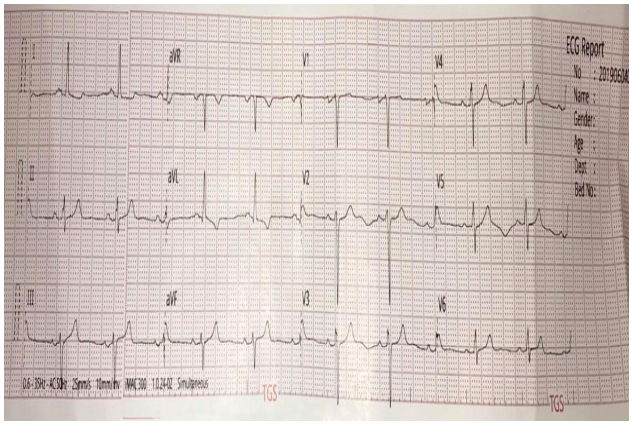

Figure 1. ECG on presentation

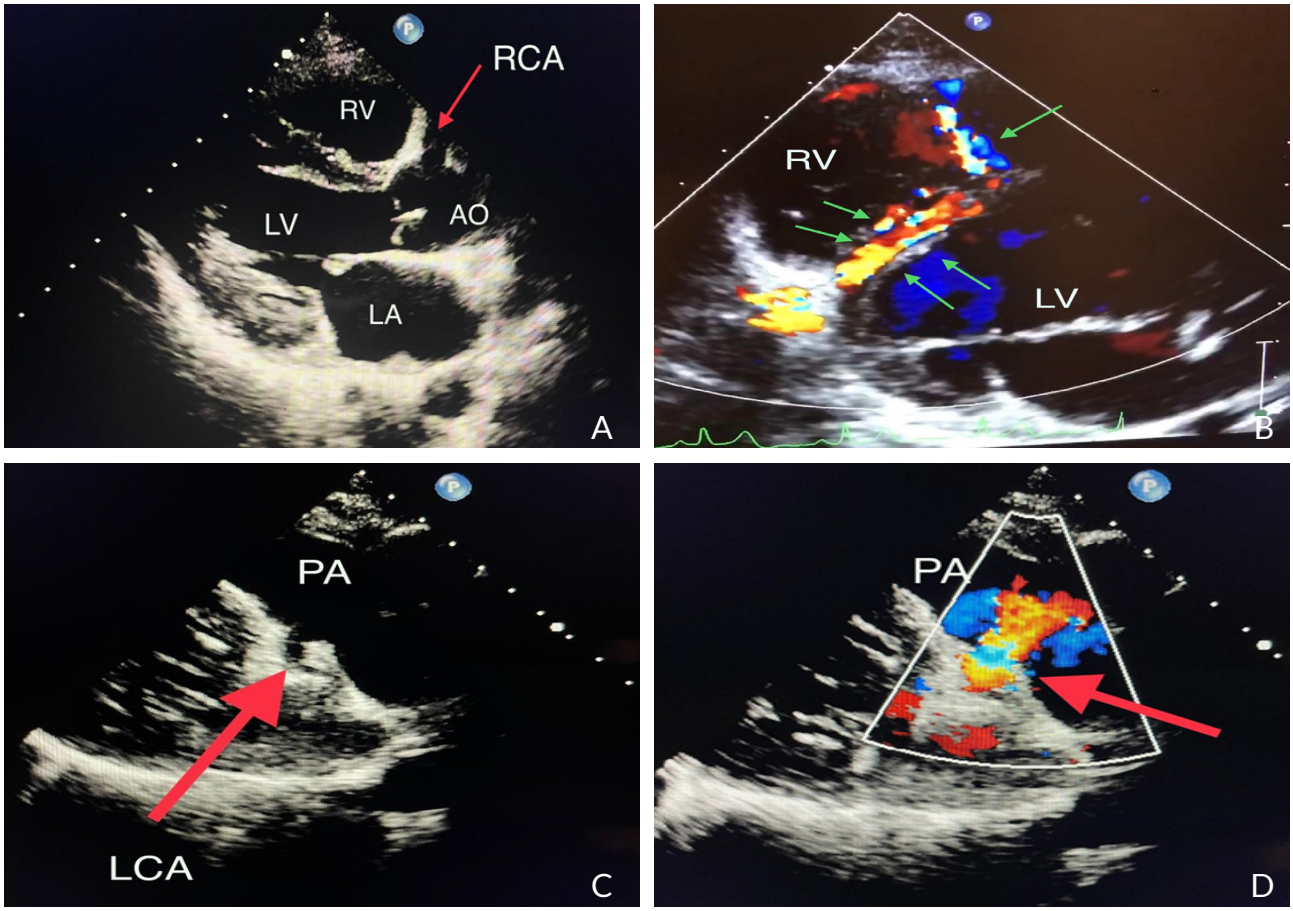

Figure 2. (A) The parasternal long-axis view. The dilated right coronary artery (RCA) is clearly visible. (B) The parasternal short-axis view. The green arrows demonstrate the increased flow in the intraventricular collateral vessels from the RCA to the LCA. (C) The parasternal right ventricle outflow view shows the take-off of the anomalous left coronary artery (red arrow) from the pulmonary artery. (D) Retrograde blood flow to the pulmonary artery from LCA (red arrow). AO: aorta, LA: left atrium, LV: left ventricle, RV: right ventricle, PA: pulmonary artery, LCA: left coronary artery.

Clip 1: https://youtu.be/18wAsx8Ab9I *Clip 2: https://youtu.be/AfUAmPnoRCg*

Clip 3: https://youtu.be/DIVfNVBOh_Y *Clip 4: https://youtu.be/uUEPZm-1xP8*

between the left and right coronary systems *(Fig. 4B, C and D)*. The ALCAPA syndrome was confirmed and the case was discussed with the surgical team who recommended surgical treatment. Although the woman and her family were advised that surgical therapy was the best option, they expressed reluctance regarding the scheduling of the operation.

DISCUSSION

According to Fernandes et al., congenital coronary artery diseases have been classified into three groups including abnormalities related to the origin of the coronary artery, terminal abnormalities and abnormalities in the distribution of the coronary artery[1]. Anomalous origin of the LCA from the

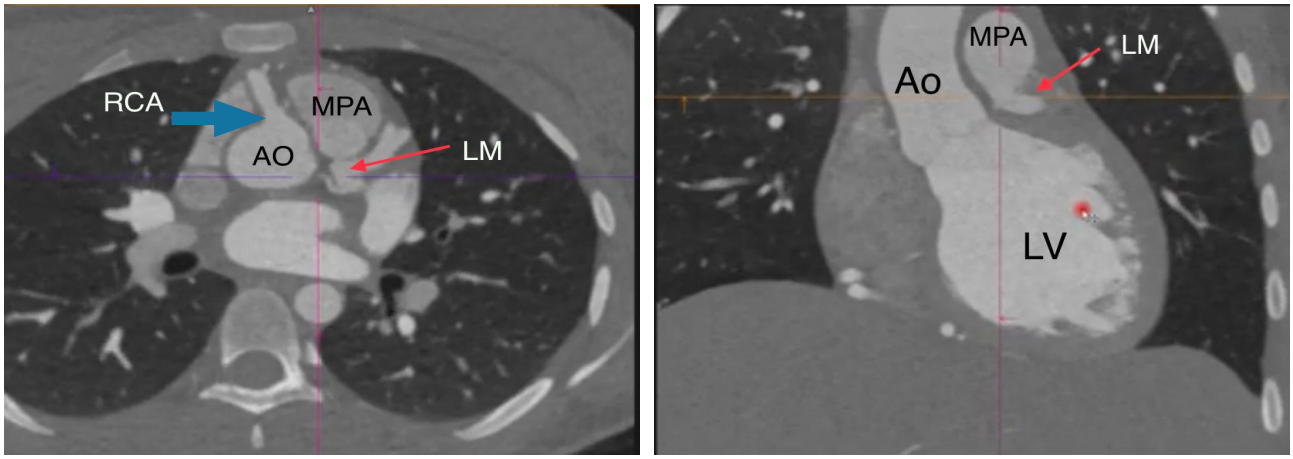

Figure 3. Coronary computed tomography: two-dimensional views of the proximal right coronary artery from the aorta (thick blue arrow) and anomalous left main coronary artery flowing into the pulmonary artery (thin red arrow). LM: left main coronary artery, AO: aorta, LV: left ventricle, MPA: main pulmonary artery, RCA: right coronary artery

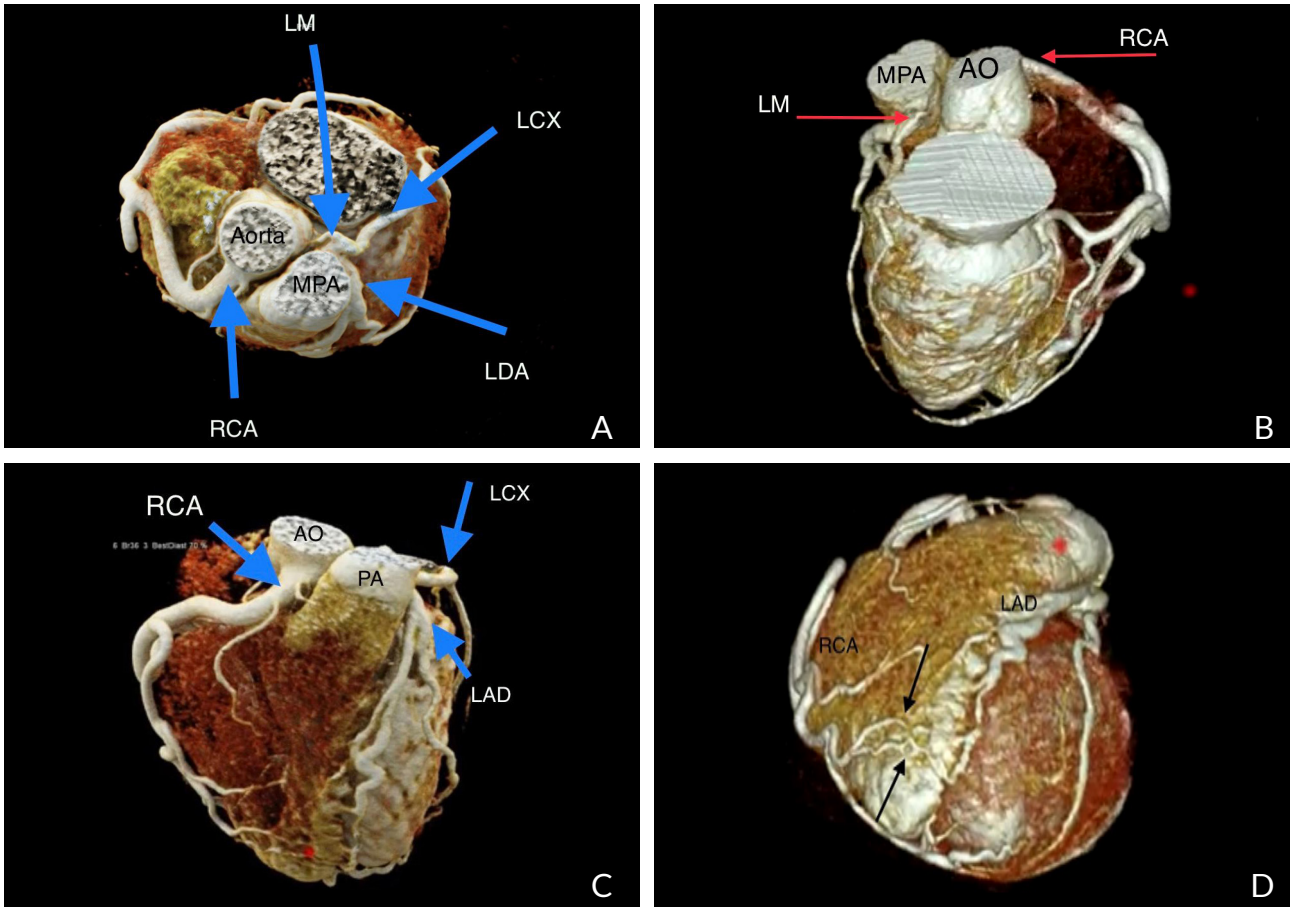

Figure 4. Multislice computed tomographic coronary angiography with 3-dimensional reconstruction images. (A) Cross-section volumetric rendering demonstrates a dilated right coronary artery (RCA) from the right aortic sinus and anomalous drainage of the left main coronary artery (LM) to the posteromedial aspect of the main pulmonary artery (MPA). (B) A posterior volumetric rendering shows RCA from the aorta and LM from MPA with collaterals between the left and right coronary systems. (C) An anterior volumetric rendering illustrates the origin of dilated RCA from the aorta and both branches of the left main coronary artery (LM), left descending artery (LDA) and left circumflex artery (LCx). (D) Volumetric rendering of the apex of the heart demonstrates a tortuous RCA with extensive apical collateralisation (black arrows) to the wrap-around left anterior descending artery (LAD)

pulmonary artery (ALCAPA) is an example of coronary origin abnormality, whereas coronary artery fistula is a coronary artery terminal abnormality. With an estimate of 1 in 300,000 live births, ALCAPA is considered a rare and potentially fatal congenital coronary artery disease accounting for 0.5% of all congenital heart diseases[2]. In 1933, a group of authors first characterised the clinical and pathological characteristics of a newborn with ALCAPA syndrome and as a result, the pathology was given the term Bland-White-Garland syndrome[3]. ALCAPA may manifest alone or in conjunction with other cardiovascular abnormalities. The proposed mechanism of embryological genesis of ALCAPA disease is either aberrant aorticopulmonary septation, or the persistence of a pulmonary endothelial bud and its attachment to the growing LCA[2]. Around 15% of ALCAPA cases may live to adulthood without surgery, while the bulk of instances were found in infants who died from acute heart failure in the first year of life[2]; however, the exact prevalence is unknown in the adult population. The modern advances in non-invasive cardiac imaging caused a significant rise in the number of adult cases diagnosed with ALCAPA syndrome[4]. The classical classification of ALCAPA syndrome is based on the age of onset and type of symptoms at presentation, and can be divided into infantile and non-infantile (or adult) types. The infantile type is characterised by little or no coronary collateral flow and clinically manifests as symptoms of heart failure within the first 12 months of life, and possible myocardial ischaemia. In the neonatal period, the anterograde flow in the anomalous artery coming from the pulmonary trunk usually persists due to the elevated pulmonary pressure that equals the systemic pressure. Nonetheless, the gradual drop in the pulmonary vascular resistance with decreased pulmonary pressure will lead to a reduction in anterograde flow and subsequent reverse flow into the pulmonary artery, and result in a coronary steal phenomenon. Both left-to-right shunt and myocardial ischaemia will determine the clinical picture as well as the intercoronary collateral circulation. The clinical picture of this type in addition to the infancy onset consists of LV enlargement, heart failure with decreased ejection fraction, aneurysm formation at the apex of the LV, mitral valve insufficiency and pathological Q waves in lateral leads on the electrocardiogram[5]. Other symptoms of infantile type entail circumoral pallor, poor weight gain, persistent tachypnoea and tachycardia[6]. Without surgical intervention, the prognosis for the infantile form of ALCAPA syndrome is typically poor and is linked to high mortality. Up to 90% of these infants die within the first year of life if left untreated.

The establishment of coronary collaterals and retrograde perfusion of the LV through the RCA are the main hallmarks of the non-infantile, or adult form. Myocardial ischaemia or sudden death is regarded as the most common presentation of the adult type. The amount of myocardial ischaemia will be determined by the acquired intercoronary collaterals. Nevertheless, the collateral flow may not be enough to supply the subendocardial region, which may cause a chronic ischaemic region that will be a potential substrate for malignant arrhythmias and increase the risk of sudden death. Based on the timing and severity of symptoms, Lotman et al. suggested dividing the adult type into two subtypes: the symptomatic adult type and the late adult type. The late adult type began in the seventh decade of life and had minimal to no limiting symptoms[7]. In the same review by Lotman et al., a case of late adult onset of ALCAPA syndrome in a 76-year-old female with minimal symptoms and good coronary collateral circulation was reported during elective coronary angiography, where a medical treatment strategy was chosen[7]. In adult patients, the risk of sudden death appears to decline after the age of 50 despite less frequent surgical correction in this population[4]. From April 1984 to July 2012, Xiao et al. reported 23 individuals with ALCAPA syndrome at Beijing Anzhen Hospital in China, with 16 instances being identified as infantile type (onset age under 12 months) and 7 cases as adult type (onset age beyond 12 months)[8]. Most adult instances presented with chest pain and heart murmurs, which can be mistakenly identified as coronary artery disease, myocarditis or patent ductus arteriosus. Echocardiography and angiography were used to diagnose ALCAPA syndrome and 21 patients underwent cardiac surgery, 6 of whom died postoperatively[8]. A total of 151 published case reports of adult-type ALCAPA syndrome were reviewed in a comprehensive literature review from 1908 to 2008 by Yau et al. The oldest adult-type ALCAPA patient in this research was 83 years old, and the average age of adult-type patients was 41 years. While ventricular arrhythmias, syncope or sudden death were observed in 17% of these patients, angina, dyspnoea or palpitations were the presenting symptoms in 66% of them, and 14% of the patients were asymptomatic. Around 12% of deaths had a diagnosis at the autopsy[9]. Most of these individuals underwent some type of surgical treatment throughout their therapeutic care. Yau et al. concluded that the availability of non-invasive modern diagnostic modalities led to an elevation of the identification of this pathology in the older population[9].

The primary diagnostic modalities for ALCAPA syndrome are 2D transthoracic echocardiography and colour Doppler flow imaging, by visualising the origin, course and flow direction of the LCA and RCA in addition to the abnormal flow/shunt into the MPA. Other indices can be also measured on echocardiography including LV size and systolic function, the presence of mitral regurgitation and the size of the RCA. Yu et al. conducted a review to assess the diagnostic value of echocardiography in detecting the various types of ALCAPA syndrome; 24 out of 30 patients were diagnosed with ALCAPA by echocardiography[10]. One of the unique characteristics of echocardiographic findings of ALCAPA in adults is the increased systolic coronary flow in a pulsed wave Doppler, which indicates the presence of collateral circulation and a coronary left-to-right shunt[11]. Other echocardiographic findings include abnormal vessel

inserting into the pulmonary artery, continuous shunt into the pulmonary artery and intercoronary collateral signals within the ventricular septum[12]. The main limitation of echocardiography is the poor images due to multiple factors. For ALCAPA syndrome, the diagnostic accuracy of echocardiography ranged between 46.0 and 80.0%[13]. Other imaging modalities that provide direct visualisation of coronary artery anatomy with 3D reconstruction are CTA and cardiac magnetic resonance (CMR). Multiplanar reformatted and 3D reconstructed computerised tomography (CT) images can confirm the suspected coronary anomaly and currently, CTA is considered the diagnostic imaging modality of choice[14].

CT and CMR findings of ALCAPA in adults contain direct visualisation of the origin of the LCA from the posterior aspect of the pulmonary artery, a dilated and tortuous RCA and visualisation of dilated intercoronary collateral arteries along the external surface interventricular septum[15]. Based on CT findings, the surgical treatment can be planned and performed[16]. An additional advantage of CMR is the assessment of mitral valve function, and delayed gadolinium enhancement may help determine myocardial viability[17].

As reported in the literature, the increased incidence of sudden death during childhood and early adulthood advances surgery as the treatment of choice. During the last few decades, a number of procedures were suggested as treatment options. Currently, for ALCAPA treatment four different operative procedures have been recommended. The procedure of choice that provides a definitive two-coronary artery anatomy with good short- and long-term results is reimplantation surgery[18]. Other procedures include the simple ligation of ALCAPA, coronary artery bypass grafting and channel repair (Takeuchi surgery)[19].

CONCLUSION

ALCAPA syndrome is a rare and potentially fatal congenital disease of the coronary artery. The prevalence of adult individuals with ALCAPA syndrome has significantly increased as a result of recent developments in non-invasive cardiac imaging.

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