Case Report

ALCAPA Syndrome in a Young Woman

Síndrome de ALCAPA em uma Mulher Jovem

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Abstract

An 18-year-old woman presented with a one-year history of syncope, angina, and palpitations. The critical clue was a dilated right coronary artery on transthoracic echocardiography. Computed tomography findings resulted in the diagnosis of anomalous origin of the left coronary artery from the pulmonary artery syndrome.

Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) syndrome, also known as Bland-Altman-Garland Syndrome, was first clinically described by these authors.[1] As a rare congenital heart condition, ALCAPA is rarely found in adults since few affected individuals survive childhood without surgical repair. Here, we report a case of this rare congenital anomaly that presented as a one-year history of syncope, angina, and palpitations. The critical clue to the final diagnosis was a dilated right coronary artery on transthoracic echocardiography (TTE).

Case description

An 18-year-old woman had a one-year history of syncope at rest accompanied by angina and palpitations on exertion. Worsening dyspnea on exertion was evident. She had no coronary risk factors or family history of premature coronary artery disease or congenital heart condition. Physical examination results were unremarkable. Electrocardiography (ECG) findings included sinus rhythm, 67 bpm, embryonic r wave in V1–V3, and a biphasic T wave in the lateral wall. Holter monitoring showed no arrhythmia, and no episodes of cardiac arrhythmia on cardiac monitoring were detected during her hospital stay. Chest radiography findings were normal. Tests for serial cardiac enzymes were negative. TTE demonstrated mildly dilated left cardiac chambers with wall motion abnormalities (WMAs) of the proximal two-thirds of the anterior wall and anterior septum and a normal left ventricular ejection fraction. An unusual dilation of the right coronary artery (RCA) was observed (Figure 1). Cardiac magnetic resonance (CMR) documented hypokinesia of the basal and mid segments of the anterior wall and the anterior septum with subendocardial late gadolinium enhancement. No other associated cardiac anomalies were noted. These findings strongly suggested an anomalous origin of the left coronary artery (LCA). Multidetector computed tomography (MDCT) coronary angiography subsequently established the diagnostic hallmark of ALCAPA syndrome (Video 1). MDCT coronary angiography images revealed an LCA branching off from the main pulmonary artery (Figure 2, arrowhead). The RCA arose from the aorta (Figure 2, arrow) with collateral circulation to the LCA. A coronary angiogram performed before surgery revealed that all coronary arteries and their branches were ectatic with tortuous courses. An extremely large and tortuous right RCA arising from the right coronary cusp and extending collaterals to the left coronary system was evident (Figure 3). Steal phenomenon was evident, with retrograde flow from the LCA into the main pulmonary artery. The patient underwent surgical correction with aortic reimplantation of her left main coronary artery. Her postoperative course was uneventful. The patient is now followed regularly.

Discussion

The estimated incidence of ALCAPA is 1/300 000 live births (0.24–0.46% of all congenital cardiac anomalies).[2] Since its late presentation is usually described,[3,4] this may be a significant underestimation of its true incidence. ALCAPA syndrome is associated with early infant mortality and sudden adult death. Its clinical expression results from evolving morphological–functional alterations in the pulmonary circulation that occur after birth. After birth, pulmonary artery saturation and pressure gradually decrease, and flow from the pulmonary artery to the LCA stops. Retrograde flow occurs from the RCA collaterals to the pulmonary artery. By this time, left ventricular perfusion fully depends upon collaterals to the LCA developing from the RCA. Coronary steal syndrome develops, leading to hypoxic damage to the left ventricular myocardium. If left untreated, up to 90% of affected children die within the first year of life.[2] An estimated 10–15% of affected children reach adulthood.[5] Their survival depends on the extent of acquired collateral circulation. Those with a well-established collateral circulation between the right and left coronary vessels have the adult type of disease, while those without collateral vessels have the infant type. The manifestations and outcomes differ between the two types.[2] The adult type is characterized by collateral circulation between the RCA and the LCA via a shunt

Keywords

Adult, ALCAPA Syndrome, Bland-White-Garland Syndrome, Echocardiography.

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Manuscript received 7/23/2020; revised 11/12/2020; accepted 11/26/2020

DOI: 10.47593/2675-312X/20213401eabc129

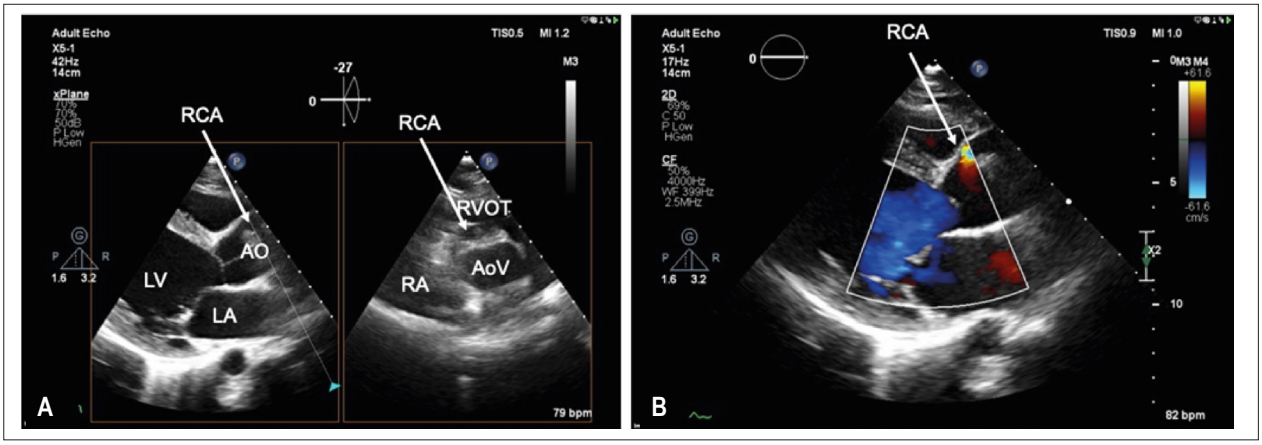
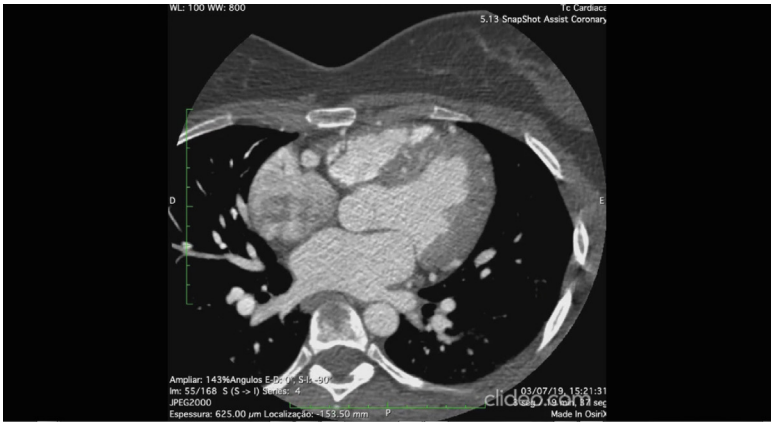


Figure 1 – *Transthoracic echocardiography examination. Panel A: (Right) parasternal long-axis view with dilated RCA. (Left) perpendicular cut along the RCA on PLAX. Parasternal short-axis view at the level of the ascending aorta with the right coronary artery on top. Panel B: Parasternal long-axis view of diastolic flow of visibly dilated RCA. AO, ascending aorta; AoV, aortic valve; LA, left atrium; LV, left ventricle; RA, right atrium; RCA, right coronary artery; RVOT, right ventricular outflow tract.*



Video 1 – *Multidetector computed tomography coronary angiography video demonstrating the abnormalities described in Figure 1.*

compensation mechanism. Symptoms and ischemia ensue whenever collateral circulation decompensation occurs. Most cases in adulthood demonstrate evidence of some degree of irreversible impairment of cardiac function.[6]

The one-year history of the syncope, angina, and palpitations triad was the initial red flag. This waiting period could be dangerous because cardiac arrest due to ventricular fibrillation can be the major clinical presentation of ALCAPA in adults.[7] The ECG findings may provide the first clue to the diagnosis. The ECG of a baby with ALCAPA syndrome usually shows typical signs of an anterolateral myocardial infarction with abnormal Q waves and transient ST changes in leads I, aVL, V5, and V6.[8] Our patient had an embryonic r wave in leads V1–V3 and biphasic T waves in the lateral wall. The critical clue to the final diagnosis was the TTE examination finding of unusual dilation of the RCA in conjunction with mildly dilated left cardiac chambers and WMAs on the LCA territory. The strong clinical- and echocardiography-based suspicions led to MDCT coronary angiography and CMR examinations. MDCT coronary angiography can reveal anomalous coronary arteries, as direct visualization of the LCA arising from the main pulmonary artery is the diagnostic hallmark of the syndrome. CMR elucidates the consequences of chronic myocardial ischemia, with hypokinesia of the LCA territory and subendocardial infarction. Indeed, imaging is pivotal to the ALCAPA diagnosis. CT coronary angiography provides an accurate and detailed description of the origin and course of the coronary arteries. CMR is used to make a more functional assessment and establish subendocardial ischemic changes or infarction (replacement-type fibrosis).[9]

Surgical treatment is suggested in patients with ALCAPA syndrome, even if asymptomatic. The objective of surgical repair is to restore normal coronary circulation and improve left ventricular myocardial perfusion.[2] Restoration of the coronary system prevents further ischemia and arrhythmias of acute ischemic origin. However, the anatomical substrate for ventricular arrhythmias in patients with old myocardial infarction will not be altered by the repair. Since our patient had myocardial scars from

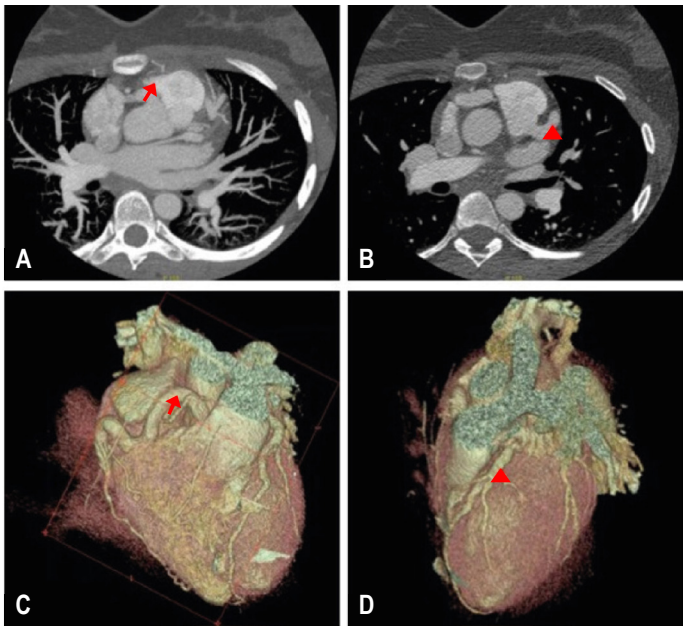


Figure 2 – *Multidetector computed tomography coronary angiography. Panels A and C: The RCA arose from the aorta (arrow) with collateral circulation to the LCA. Panels B and D: The LCA is branching off from the main pulmonary artery (arrowhead). AO, ascending aorta; ALCAPA, anomalous origin of the left coronary artery from the pulmonary artery; PA, pulmonary artery; RCT, right coronary artery.*

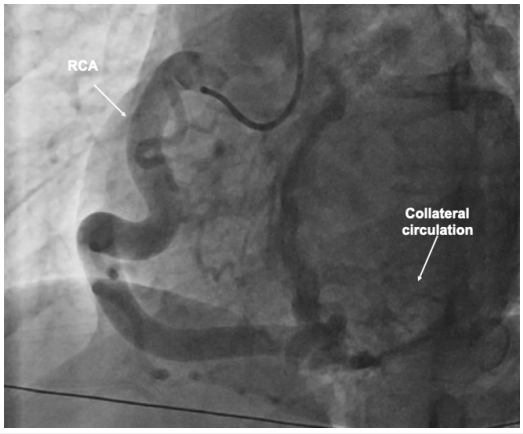


Figure 3 – *Invasive coronary angiography. The right coronary artery (RCA) with visible collateral circulation to the left coronary artery (LCA).*

chronic ischemia, long-term clinical and electrocardiographic (ECG and Holter ECG) monitoring continues.[7]

Conclusion

In this case report, the critical clue was the pathologic ECG findings of the ischemic heart and mildly dilated left cardiac chambers with unusual RCA dilation. CMR further demonstrated myocardial scarring, while MDCT established the diagnostic hallmark. It is critical to remember that the patient's complaints may be validated by the ECG findings and careful TEE.

Author contributions

Research creation and design: Campos D, Puga L, Guardado J, Saleiro C, Lopes J, Teixeira R, Gonçalves L. Análise e interpretação dos dados: Campos D, Puga L, Guardado J, Saleiro C, Lopes J, Teixeira R, Gonçalves L. Manuscript writing: Campos D, Puga L, Guardado J, Saleiro C, Lopes J. Critical revision of the manuscript for important intellectual content: Teixeira R, Gonçalves L.

Conflict of interest

The authors have declared that they have no conflict of interest.

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