Case Report

Cardiac Arrest in Pacient with ALCAPA Syndrome. Case Report

Parada Cardiorrespiratória em Paciente com Síndrome de ALCAPA. Relato de Caso

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Introduction

Alcapa Syndrome, also known as Bland-Altman-Garland Syndrome, is a rare coronary anomaly in which the left coronary artery anomalously originates from the pulmonary artery. Two types of Alcapa syndrome are described: the infant type and the adult type. The infant type is the most symptomatic one and 90% of patients die in the first months of life, while the adult type is characterized by the formation of a collateral system between the right coronary artery (RCA) and the left coronary artery (LCA), with a shunt volume compensation mechanism at varying levels. The adult type tends to be asymptomatic, with collateral circulation decompensation leading to ischemia. The following is the report of a case of Alcapa syndrome in a 28-year-old female patient who presented an episode of cardiopulmonary arrest and remained at the intensive care unit for 33 days. [1-3]

Case report

Female patient, 28 years old, admitted to the emergency room after cardiopulmonary arrest (CPA), where she was resuscitated for 22 minutes and referred to the intensive care unit (ICU). The patient was sedated, on invasive mechanical ventilation, and presented severe hemodynamic instability dependent on vasoactive drugs (VAD). Pulmonary examination: vesicular murmur reduced at the bases, on mechanical ventilation with respiratory rate of 16 rpm, PEEP 5 cmH2O, FiO2 40%. Cardiovascular examination: heart rate of 76 bpm, VAD-dependent 85 mmHg MAP, sinus rhythm, systolic murmur in mitral focus (2+). Abdominal examination: distended tympanic abdomen with reduced hydroaereal noises. Lower limbs: no edema, bilaterally palpable pulses. History of mitral valve prolapse on metoprolol 25 mg daily. Laboratory tests revealed hematocrit 41.50%, hemoglobin 13.80 g/dL, leukocytes 11,000/mm^3, platelets 125,000/mm^3, creatinine 1.98 mg/dL, potassium 4.0 mEq/L, sodium 141 mEq/L, BNP 1080.3 pg/mL, troponin I 1,564 ng/mL, creatine phosphokinase MB fraction 109.01 U/L, normal and partial arterial blood gas with no particularities.

Keywords

Coronary Disease; Cardiology; Congenital Abnormalities.

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On electrocardiogram, the patient presented sinus rhythm, signs suggestive of left ventricular hypertrophy and ventricular repolarization disorders in the upper lateral wall. As for imaging scans, chest radiography showed infiltration in the pulmonary bases; later, right perihilar opacification and bilateral pulmonary congestion. On transthoracic echocardiogram, the patient presented discrete left ventricular dysfunction (54% ejection fraction by the Teichholz method), no papillary muscle hyperrefringence, and presence of pericardial effusion in the inferior, posterior and lateral left ventricular walls, and mitral valve prolapse with moderate to severe regurgitation. Chest tomography showed a mixed pattern of acute respiratory distress syndrome (ARDS) and cardiac magnetic resonance imaging showed no abnormalities.

The patient remained at the ICU on mechanical ventilation with high PEEP and FiO2, and was subsequently tracheostomized. She presented repetitive episodes of nosocomial pneumonia, evolving to ARDS. The main events on evolution were acute lung edema and septic shock. She was prescribed various antibiotic regimens and used vasoactive drugs while in hospital.

After a few days, the patient presented hemodynamic and PO2/FiO2 ratio improvement and starting weaning of vasoactive drugs and mechanical ventilation. After four days on spontaneous ventilation, implantable cardiodesfibrillator was implanted as a primary prevention of sudden death. Thirty-three days after admission, she was discharged from the ICU. Coronary angiotomography was performed as an operative evaluation for the correction of mitral regurgitation, previously diagnosed on echocardiography. Incidental diagnosis of anomalous origin of coronary artery was delivered. Months later, she was referred for surgical procedure at another service, where she died during the surgery.

Discussion

We report the case of a patient with no previous diagnosis of Alcapa Syndrome, admitted to the Intensive Care Unit after cardiopulmonary arrest, presenting multiple complications in the evolution, being discharged after thirty-three days in hospital. The patient died during the surgery to correct coronary anomaly after a few months of diagnosis.

The Alcapa Syndrome is a rare coronary anomaly, with an incidence of 1 in every 300,000 live births. It is described as anomalous origin of the LCA from the pulmonary artery (PA), and is considered a pathway origin anomaly. In general, the Alcapa syndrome occurs in isolation, but there are rare cases of occurring in conjunction with other congenital pathologies. Two types of Alcapa syndrome are described: the infant type and the adult type. The infant type is more common and symptomatic, with the presence of chest pain (signaled by neonate irritation),

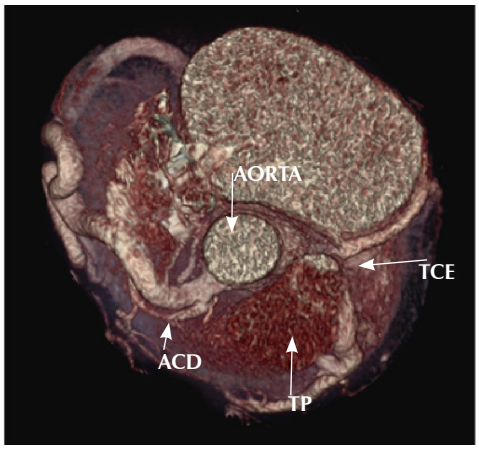

Figure 1 – *3D coronary computed tomography angiography reconstruction showing the pulmonary trunk (PT) originating the left main coronary artery (LMCA). On the side, the aorta originating the right coronary artery (RCA).*

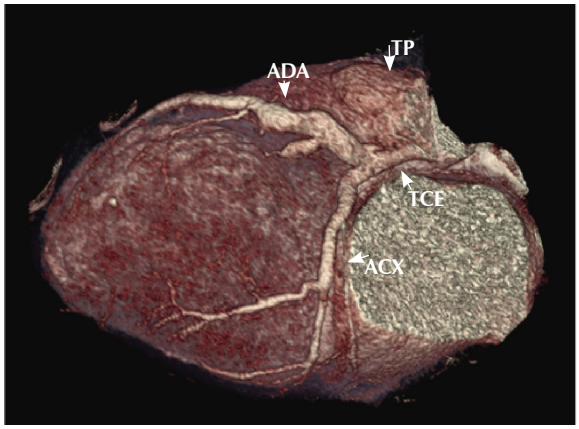

Figure 2 – *3D coronary computed tomography angiography reconstructions originating the LMCA, from which the anterior descending artery (ADA) and the circumflex artery (CxA) come out.*

pallor, sweating and dyspnea, with a high mortality rate. The adult type presents a clinical picture that may vary from asymptomatic (due to collateral compensation between RCA and LCA) and symptomatic (due to failure of collateral compensation). [1, 3- 6]

Imaging tests of choice are multislice computed tomography (MSCT) and cardiac nuclear magnetic resonance imaging (MRI). MSCT shows direct morphology and anatomical variations, which could also be identified by MRI or angiography, but less accurately. MRI is used to make a more functional assessment and can identify the inverse flow in LCA. Another extremely important test, especially in the infant type, is the two-dimensional echocardiogram, which presents well-established criteria for diagnosing the Alcapa syndrome, namely: identification of dilated RCA, retrograde Doppler flow of the LCA to the pulmonary artery and collateral flow from the septal flow. Coronary angiography can also be used and requires three criteria to confirm the Alcapa syndrome: (1) LCA retrograde flow; (2) LCA originating from the pulmonary artery trunk; and (3) absence of LCA originating in the aorta. Functional tests, such as myocardial scintigraphy and Holter monitoring, should be performed annually on any adult with the ALCAPA syndrome, even in the absence of symptoms.[2, 4, 5, 7]

Therapeutic approach is divided into non-surgical and surgical. The non-surgical approach includes the use of beta-blockers to reduce ischemia and reduction of the patient's physical exertion to reduce myocardial work. However, this is not a consensual treatment. The most commonly used surgical approach is to repair the anatomy by redeploying the LCA in the ascending aorta. Heart transplantation is recommended for patients with severe cardiac dysfunction, contraindicating the commonly used surgical technique.[5,8]

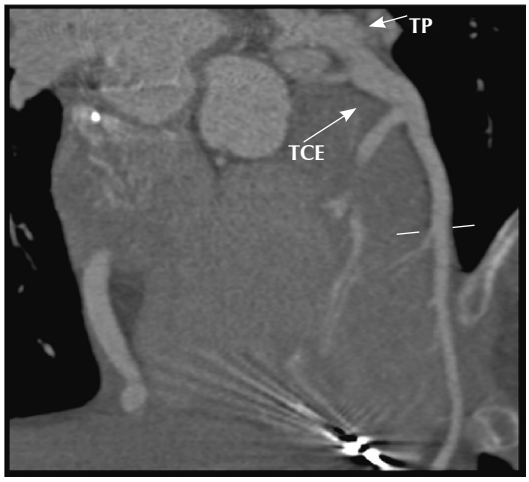

Figure 3 – *3D computed tomography angiography reconstructions showing the PT originating the LMCA.*

In the case described here, the clinical presentation was asymptomatic until adulthood, in which the patient presented a sudden CRA. Diagnosis was based on coronary angiography, performed as a preoperative examination to correct mitral regurgitation, previously seen on echocardiography. Initial treatment was with beta-blockers, prescribed due to mitral valve prolapse. Surgical treatment was proposed after diagnosis of Alcapa Syndrome.

Recently, there has been an improvement in the prognosis of patients with ALCAPA due to the early diagnosis provided by imaging tests and improvement of surgical techniques. The success of surgical procedures varies depending on the myocardial condition at the time of diagnosis and patient's clinical repercussion. The later the diagnosis, the greater the chances of myocardial damage caused by ischemia and the greater the chances of ventricular dysfunction and mitral regurgitation. These findings may have a significant impact on the prognosis of these patients.[7]

We report the case of an adult female patient with Alcapa syndrome, diagnosed late after CRA, with recovery and later death during corrective surgery.

Conflict of interest

The authors declare that there is no conflict of interest regarding this manuscript.

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