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CORONARY ARTERY ANOMALY: CASE REPORT

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Abstract: Introduction: Coronary artery anomalies (CAA) are congenital alterations in the origin, course and structure of the epicardial coronary arteries. The origin and proximal course of anomalous coronary arteries are the main predictors of severity. It constitutes the second most frequent cause of sudden death of cardiovascular origin in competitive athletes and can be a cause of heart failure and death in infants. In this article, a case of a patient with unstable angina for 6 months finally diagnosed with the disease will be described. **Case report:** A 35-year-old female patient with no comorbidities was admitted to the Emergency Room with unstable angina. She had been undergoing an outpatient investigation for chest pain for 6 months. She performed a submaximal and positive treadmill test for myocardial ischemia, as well as coronary CT angiography (CT angiography) that showed an anomalous origin of the left main coronary artery, originating in the right coronary sinus and with a malignant path between the aorta and the pulmonary artery. Oral therapy with cardioselective beta-blockers (bisoprolol) and trimetazidine was started. It evolved in a refractory manner to drug treatment, with worsening of the angina with increased frequency and intensity, appearing at rest and with associated dyspnea (ccs4). She sought the emergency room and was hospitalized for surgical approach. For better visualization of the coronary bed, a preoperative catheterization was performed. The procedure performed by cardiovascular surgery was the bridge of the right internal mammary artery to the marginal artery and the left internal mammary artery to the anterior descending artery. The patient evolved without complications in the postoperative period. **Discussion:** The diagnosis of AAC is a challenge, since patients are usually asymptomatic and physical examination does not reveal any changes. The *American Heart*

Association considers cardiac CT angiography an adequate method for diagnosing CAA. Once diagnosed, AAC must be surgically corrected promptly to prevent complications and sequelae typical of the natural history of the disease. **CONCLUSIONS:** Although rare, it is a lethal condition if not diagnosed and treated early. After clinical confirmation of the disease, surgical therapy presents a viable option and, in general, with a good prognosis. As it is a rare disease, further studies are still needed to determine an effective screening and optimal treatment for each patient.
Keywords: Coronary arteries, anomalies, Congenital heart diseases.

INTRODUCTION

Coronary artery anomalies (CAA) are congenital alterations in the origin, course and structure of the epicardial coronary arteries. The classification proposed by Angelini considers normal characteristics of the coronary arteries: the presence of 2 to 4 ostia, with a location in the right and left coronary sinuses, and a proximal orientation of the coronary artery of 45° to 90° to the aortic wall (Ao) ; the presence of only one common trunk (CT), located on the left; that the proximal segment presents a direct path, from the ostium to the territory it irrigates; the middle segment is subepicardial, with branches suitable for the dependent myocardium, and that this system ends in capillaries.

The true incidence of AAC in the general population remains unclear. By definition, AAC occur in less than 1% of the general population, with alterations with higher incidences being considered variants of normal. Currently, there are still no data that point to a difference in incidence between men and women or between different races.

The origin and proximal course of anomalous coronary arteries are the main predictors of severity. It constitutes the second most frequent cause of sudden death of cardiovascular origin in competitive athletes and can be a cause of heart failure and death in infants.

In embryology, vascular sinusoids develop within the embryonic myocardium. When it becomes compact, they disappear and a network of veins, arteries and capillaries appears that communicate with other mediastinal vessels, approximately at 32 days of gestation. The primitive coronary vessels appear around the seventh week of gestation, after the formation of the aorta from the division of the coronary artery. *truncus arteriosus*. As the coronary artery network develops, endothelial buds appear at the base of the *truncus arteriosus*, which later join with the coronary arterial network, which develops from the sinusoids, establish the definitive coronary arterial system. The anomalous involution, the position of the endothelial buds or the septation of the *truncus arteriosus* can lead to the development of anomalies at the origin of the coronary arteries.

In the pathophysiology, it is observed that during intrauterine life there is no functional change. The problem, however, occurs after birth, as there is a reduction in arteriolar resistance, pulmonary artery pressure and oxyhemoglobin saturation of venous blood. In this context, the ductus arteriosus and foramen ovale are closed, progressively decreasing the blood perfusion of the myocardium irrigated by the anomalous artery, also leading to ischemic changes. CAAs usually arise from an isolated defect, but can be associated, in 5% of cases, with other heart defects, such as ventricular septal defect, atrial septal defect, coarctation of the aorta, tetralogy of Fallot and other anomalies.

As for the clinical picture, it presents two forms: an infantile form and an adult form. The infantile form is characterized by poor intercoronary collateral circulation, which can

result in myocardial infarction, heart failure, or sudden death. In this form of presentation, symptoms occur in the first months of life, with the appearance of constant crying, intense pallor, sudden cessation of suction, weight loss and signs of heart failure. The adult form is characterized by a richness of intercoronary collateral circulation, of sufficient magnitude to allow survival into adulthood, with reported survival up to 72 years of age. Sometimes there are no appreciable clinical manifestations and the diagnosis is suspected by the presence of murmurs: continuous, by the passage of blood through the coronary-pulmonary fistula, or systolic, by mitral valve dysfunction.

In this article, a case of a patient with unstable angina for 6 months, finally diagnosed with the disease, will be described.

CASE DESCRIPTION

A 35-year-old female patient with no comorbidities was admitted to the Emergency Room with unstable angina.

She had been undergoing an outpatient investigation for chest pain for 6 months. A submaximal and positive exercise test for myocardial ischemia was performed, as well as coronary computed tomography (CT) angiography (Figure 1) which showed an anomalous origin of the left main coronary artery, originating in the right coronary sinus and with a malignant path between the aorta and the pulmonary artery.

Oral therapy with cardioselective beta-blockers (bisoprolol) and trimetazidine was started. It evolved in a refractory manner to drug treatment, with worsening of the angina with increased frequency and intensity, appearing at rest and with associated dyspnea (ccs4).

She sought the emergency room and was hospitalized for surgical approach. For better visualization of the coronary bed, a preoperative catheterization was performed (Figure 2). The procedure performed by cardiovascular surgery was the bridge of the right internal mammary artery to the marginal artery and the left internal mammary artery to the anterior descending artery. The patient evolved without complications in the postoperative period.

DISCUSSION

The diagnosis of AAC is a challenge, since patients are usually asymptomatic and physical examination reveals no changes. In the electrocardiogram, stress test or Holter, there are no specific electrocardiographic changes that make the diagnosis of AAC. The presence of alterations suggestive of ischemia and cardiac arrhythmias, in children or young people, may raise suspicion and guide towards other complementary means of diagnosis. Echocardiography is an attractive screening method, considering that it is a non-invasive method, is widely available, has a low cost and does not involve the use of ionizing radiation. However, in the diagnosis of AAC, studies demonstrate variable sensitivity.

Conventional coronary angiography was traditionally considered the gold standard in the diagnosis of AAC. However, this is an invasive test and involves the use of nephrotoxic contrast and ionizing radiation. On the other hand, non-invasive coronary angiography by CT Angio or Magnetic Resonance allows a better characterization of the origin of the coronary arteries when there is difficulty, or even impossibility, of selective catheterization of the coronary arteries by invasive coronary angiography. The *American Heart Association*, considers cardiac CT angiography an adequate method for diagnosing CAA, giving it a score of 9 (maximum classification attributable to a complementary diagnostic method for a given purpose).

Once diagnosed, AAC must be surgically corrected promptly to prevent complications

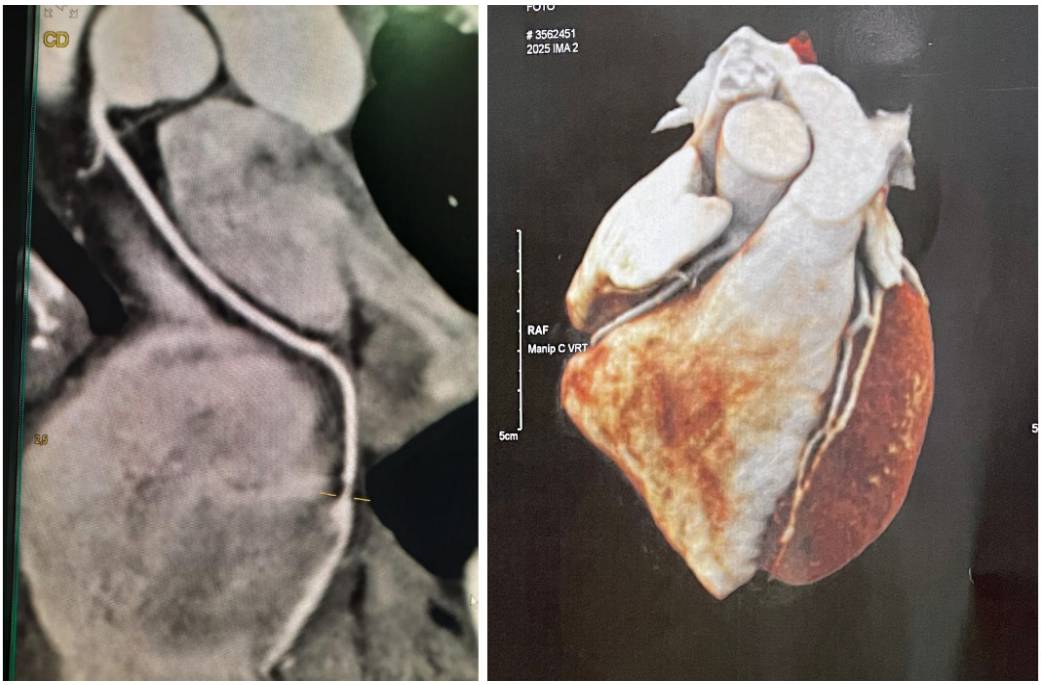

Figure 1. CT angiography demonstrating the left main coronary artery without luminal reduction, which arises from the right coronary sinus with a path between the aorta and the pulmonary artery towards the anterior face of the heart.

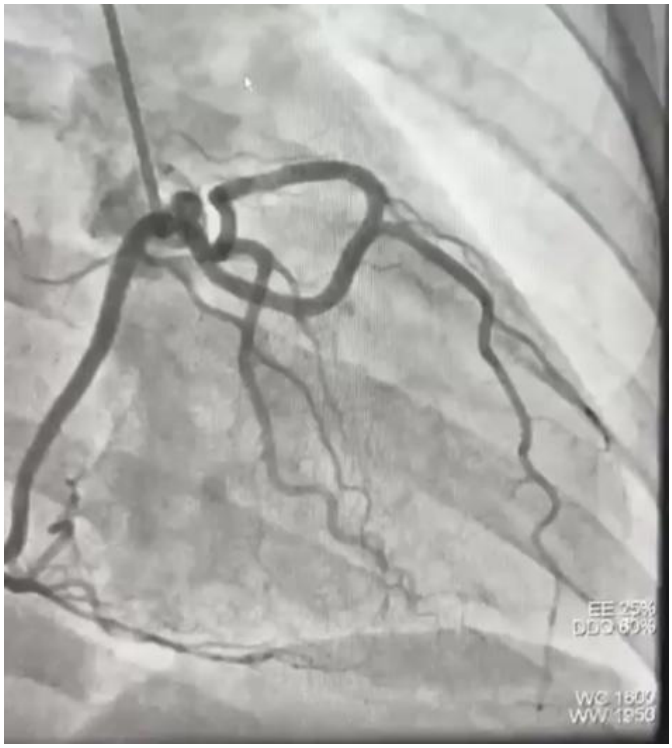

Figure 2. Coronary angiography performed with JR 5F Catheter and selective injection of contrast in the right coronary artery (RC). We observed opacification of the entire CD and also of the left coronary artery (LE), which has its origin in the right coronary sinus, next to the CD.

and sequelae typical of the natural history of the disease. The surgical technique of choice is reimplantation with translocation of the left coronary artery from the pulmonary trunk to the aorta, which is possible in most cases. In the infantile type of this syndrome, the anomalous coronary artery can be directly implanted into the aorta or an intrapulmonary conduit can be created from the left coronary ostium to the aorta (Takeuchi procedure). In adults, ligation of the left coronary artery to the pulmonary artery is usually performed, combined with placement of an internal mammary artery or saphenous vein graft, as performed in the case described.

CONCLUSIONS

Coronary artery anomalies are a relevant cause of sudden death of cardiovascular origin in adults and can be a cause of heart failure and death in infants. Although rare, it is a lethal condition if not diagnosed and treated early. After clinical confirmation of the disease, surgical therapy presents a viable option and, in general, with a good prognosis.

As it is a rare disease, further studies are still needed to determine an effective screening and an ideal treatment for each patient.

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