Duplicity of the circumflex artery, a coronary anomaly: Case report

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ABSTRACT

Congenital anomaly of the coronary arteries (CACA) is a rare situation in the population, being accidentally found during coronary angiography due to suspicion of other pathologies. The individual may remain asymptomatic for a long period of time, making diagnosis difficult. Among the coronary irregularities is the circumflex coronary artery (Cx), which has been little reported in the literature. Thus, the present study aims to report a case of coronary anomaly, characterized by the duplicity of the Cx artery with anomalous origin in the right coronary artery, in a previously asymptomatic individual. The patient is a 57-year-old female patient with hypertension, diabetes, smoker and hypothyroidism. The patient sought medical attention due to chest pain of severe intensity, in tightness, associated with dyspnea on moderate exertion, which improved with rest. Coronary angiography showed left ventricular (LV) hypertrophy with preserved contractility and coronary anomaly, highlighting the presence of another Cx artery with anomalous origin in the right coronary artery, without the presence of a malignant pathway. After hospital discharge and outpatient return, the patient continues to be maintained with home drug therapy using enalapril, carvedilol, hydrochlorothiazide, simvastatin, ASA and levothyroxine. Although coronary anomalies are pathologies with a low population incidence, they can be potentially lethal, especially in young people, corresponding to the second leading cause of sudden death related to the cardiovascular system. Upon clinical suspicion, early diagnosis and treatment are of paramount importance.

Keywords: Coronary Anomaly, Double Circumflex Artery.

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INTRODUCTION

The coronary system, in its normal anatomy, is formed by the main coronary artery (originating from the left coronary sinus) and the right coronary sinus. The main coronary artery is divided into the anterior descending artery (DA) and the circumflex artery (Cx); the DA artery follows a posterior path to the pulmonary trunk, in the anterior interventricular sulcus; the Cx artery follows the posterior atrioventricular sulcus. From the right coronary sinus, the right coronary artery originates, which follows the path of the anterior atrioventricular sulcus. Thus, the occurrence of alterations in the suprascript anatomy are characterized as coronary anomalies9.

Congenital coronary artery anomaly (CACA) is an alteration resulting from disturbances during fetal development, causing anatomical variations regarding the origin, trajectory, termination or intrinsic anatomy of the coronary arteries8.

In the early stages of fetal development, within the human embryonic myocardium, vascular sinusoids develop that disappear when the myocardium becomes compact. At this time, they give rise to a network of veins, arteries and capillaries that communicate with other vessels present in the mediastinum around the 32nd day of gestation. After the formation of the aorta, from the division of the trunk arteriosus, around the seventh week of gestation, the primitive coronary vessels appear. These vessels develop and unite with the endothelial buds originating at the base of the trunk arteriosus, forming the definitive coronary artery system. Thus, factors such as anomalous involution, position of the endothelial buds, or septation of the trunk arteriosus can cause anomalies in the origin of the coronary arteries7.

ACCA is a rare situation in the population, being accidentally found during coronary angiography due to suspicion of certain pathologies3. Thus, the individual may remain asymptomatic for a long period of time, making the diagnosis difficult9. No data were found in the literature that reported differences in incidence in relation to gender or ethnicity7.

Chest pain, dyspnea or syncope are symptoms that occur in 18% to 30% of individuals with coronary anomalies7. Myocardial ischemia results from changes in perfusion caused by the movement of vessels with an anomalous trajectory during the cardiac cycle, which can cause acute myocardial infarction (AMI)[5].

The pathophysiology of the symptoms may be related to the proximal oblique course in the anomalous coronary artery, causing its ostium to have a slit shape that can collapse with the aortic expansion promoted in systole, resulting in impairment of the blood flow supplied to the myocardium. Thus, during physical exercise, the increase in systemic blood pressure and pulmonary territory can cause compression of the interarterial coronary segment, dynamic obstructions and, consequently, ischemia and arrhythmias2.

Another mechanism related to myocardial ischemia and sudden death is related to the occurrence of spasms in the anomalous coronary artery resulting from endothelial injury4. Studies indicate that ACAC corresponds to 17% of the causes of sudden death7.

In cases that portray a benign pathological trajectory, no greater risks to the individual with disease are demonstrated. However, there are situations with potential severity, which may trigger events such as myocardial ischemia, acute myocardial infarction, and sudden death, and the predictive factors of severity correspond to the origin and proximal course of the anomalous coronary artery9. The interarterial tract is considered malignant, since it is related to a higher risk of sudden death6. The Cx artery with an anomalous origin in the right coronary system is a coronary anomaly that has been rarely reported in the literature3.

RATIONALE

Studies performed by coronary angiography or autopsy indicate that the incidence of ACCA ranges from 0.3% to 1.5%[9]. However, it is estimated that coronary anomalies occur in less than 1% of the general population1. In Europe and the United States, the presence of ACAC is the second most frequent cause of sudden death related to the cardiovascular system in athletes, accounting for 12.2% to 17.2% of cases. Thus, athletes with coronary anomalies have a 79-fold increased risk of sudden death compared to non-athletes6.

Thus, as it is a rare pathology with lethal potential, especially related to premature morbidity and mortality in young adults9,6, this report contributes to the elucidation of the subject, since it is of paramount importance to establish early diagnosis and treatment. In addition, this study can serve as a basis for other health research.

GENERAL OBJECTIVE

To report a case of coronary anomaly characterized by duplicity of the circumflex artery with anomalous origin in the right coronary artery in a previously asymptomatic individual.

CASE DESCRIPTION

A 57-year-old female patient, hypertensive, diabetic, smoker, and hypothyroidism, sought medical attention due to severe chest pain, in tightness, associated with dyspnea on moderate exertion, which improved with rest.

Myocardial scintigraphy revealed transient hypoperfusion of the basal segment of the anterior wall of the left ventricle (LV), indirect signs of hypertrophic cardiomyopathy, and myocardial area at risk corresponding to 3% of the total myocardium. Coronary angiography revealed LV hypertrophy

with preserved contractility and coronary anomaly, highlighting the presence of another Cx artery with anomalous origin in the right coronary artery, without the presence of a malignant pathway.

After being hospitalized for 4 days, evolving hemodynamically stable and without intercurrences, the patient was discharged for home treatment using levothyroxine, enalapril, bisoprolol, simvastatin and metformin.

Ten days after hospital discharge, the patient returns for a follow-up appointment. She reported fatigue and angina on minimal exertion, and was classified in NYHA (*New York Heart Association) functional group III.* New laboratory tests and transthoracic echocardiogram were requested, which showed grade 1 LV diastolic dysfunction (relaxation dysfunction), LV concentric hypertrophy, mild mitral and tricuspid regurgitation, slight left atrial enlargement, and mild ascending aortic ectasia. The patient continues to be maintained with home drug therapy with the use of enalapril, carvedilol, hydrochlorothiazide, simvastatin, ASA and levothyroxine.

DISCUSSION

In 1933, the occurrence of anomalous origin of the Cx artery in the right coronary artery was described for the first time. In 2008, other authors reported a case of Cx artery duplicity. Subsequently, other cases of the presence of two Cx arteries were described in the literature3. This study reports a coronary abnormality evidenced by the presence of two Cx arteries, one of which has an anomalous origin in the right coronary artery.

The majority of ACAC patients are asymptomatic, which is a factor that hinders early diagnosis6,7. However, in the presence of symptoms, atypical chest pain, dyspnea, exercise-related presyncope or syncope, arrhythmias, and left ventricular dysfunction are often observed6. In the present case, the patient developed chest pain and dyspnea after a long period of asymptomatology.

In the study by Cosansu et al3, they reported a case of a patient with two Cx arteries, one of which originates in the left main trunk, and the other originates from the proximal part of the right coronary artery, showing great similarity with our study.

Regarding diagnosis, coronary angiography is considered the ideal test to diagnose coronary anomalies5,9. However, non-invasive imaging studies have shown better definitions of coronary anomalies related to the origin and course of these arteries. Thus, computed tomography of the coronary arteries demonstrates significant accuracy regarding the origin and proximal course of the ACCA. Transesophageal echocardiography is a high-sensitivity test to detect the anomalous origin of coronary arteries and to delineate the proximal course and blood flow pattern in these arteries9.

As for treatment, there are three types to be considered: 1- drug treatment and patient observation; 2- angioplasty with endoprosthesis placement; 3- Surgical treatment, and this procedure is recommended for most cases of anomalous origin of the left coronary artery6 and for patients with

significant symptoms4. In children, the anomalous origin of the left coronary artery in the pulmonary artery is indicated for surgical treatment9.

According to the *American College of Cardiology and the American Heart Association*, surgical revascularization is recommended for individuals who present: anomalous origin of the left main coronary artery with interarterial pathway; anomalous origin of the right coronary artery with interarterial tract associated with myocardial ischemia; and presence of myocardial ischemia in the territory of the anomalous coronary artery, with no other evident cause6.

The presence of ACAC as an anomalous origin of the left coronary artery in the right sinus or other coronary anomalies that show symptoms caused by ischemia, high-risk anatomy or major changes in perfusion, indicate surgical treatment, and it is important to advise the restriction of exercises until surgery. Postoperative follow-up is based on tests such as electrocardiogram and echocardiogram7.

Despite the presence of significant diffuse stenosis in the anomalous Cx artery reported by Cosansu et al3, interventions in this artery were not considered, since the symptoms presented by the patient were relieved after the procedure in dominant Cx. Another study presented an asymptomatic patient with Cx anomaly, and conservative treatment was chosen4. In our study, there was no need for surgical intervention to correct the patient's coronary anomaly, receiving only drug treatment to control previous comorbidities.

CONCLUSION

Coronary anomalies are pathologies with a low population incidence. However, they can be potentially lethal, especially in young people, and are the second leading cause of sudden death related to the cardiovascular system.

ACCA presents as asymptomatic and benign or malignant pathologies, expressing symptoms such as chest pain, dyspnea, syncope, and ischemic events, which may result in AMI.

Upon clinical suspicion, early diagnosis and treatment are of paramount importance. Computed tomography of the coronary arteries is the imaging test of choice. However, coronary angiography is more commonly performed.

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