Case Report

Adult-type Bland-White-Garland syndrome

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ABSTRACT

The anomalous origin of the left coronary artery from the pulmonary artery, or Bland-White-Garland syndrome, is a rare congenital coronary anomaly that results in altered myocardial perfusion and left-right shunt. Ninety percent of patients with this syndrome die within the first year of life if the condition is left untreated. Diagnosis of this abnormality in adulthood is even rarer. We describe the cardiac catheterization and coronary computed tomography angiography findings of a recently diagnosed 27-year-old patient who underwent surgical correction.

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Síndrome de Bland-White-Garland do tipo adulto

RESUMO

A origem anômala da artéria coronária esquerda do tronco pulmonar, ou síndrome de Bland-White-Garland, é uma anomalia coronária congênita rara, que resulta em perfusão miocárdica alterada e shunt esquerdo-direito. Dentre os pacientes com esta síndrome, 90% morrem no primeiro ano de vida se não tratados. O diagnóstico desta anormalidade na idade adulta é ainda mais raro. Descrevemos os achados do cateterismo cardíaco e da angiotomografia coronária de uma paciente de 27 anos, recentemente diagnosticada e submetida à correção cirúrgica.

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Introduction

The anomalous origin of the left coronary artery from the pulmonary artery is congenital and rare and characterizes Bland-White-Garland (BWG) syndrome. This condition in embryos can result from atypical septation of the truncus arteriosus into the aorta and pulmonary artery or the persistence of lung buds, resulting in involution of the aortic buds, which subsequently form the coronary arteries. Its incidence is estimated at one in every 300,000 live births, accounting for 0.5% of all congenital heart diseases. Mortality is high (90%) in the first year of life, and only 10 to 15% of patients with this condition reach adulthood. There are two recognized forms: the infant and the adult type. The former is associated with poor collateral circulation, which can lead to myocardial infarction, heart failure and sudden death. The latter is characterized by extensive intercoronary collateral circulation, which allowed survival of up to 72 years of age as reported in the literature. The purpose of this study was to describe the case report of an adult patient with this rare syndrome.

Case report

Patient FNSM, a 27-year-old, white, female accounting professional, stated that she began to have symptoms of chest pain 5 years before diagnosis. The chest pain was constrictive, of mild intensity and related to major physical exertion accompanied by fatigue,
dyspnea and palpitations. She reported that she was always healthy but remembered feeling some retrosternal discomfort and fatigue when performing exhausting activities since childhood. She had no known risk factors for coronary artery disease.

Physical examination revealed a good general status, acyanotic state, uncharacteristic facies, eutrophic weight (body mass index - BMI - of 24.2 kg/m²), blood pressure of 120 x 80 mmHg and heart rate of 74 bpm. Peripheral pulses were present and were symmetrical and within the normal range. Cardiac auscultation was normal, and chest auscultation revealed normal respiratory sounds. The abdomen revealed no evidence of visceromegaly, and bowel sounds were normal. The lower limbs had no edema and exhibited adequate tissue perfusion.

Chest radiography revealed no abnormalities (cardiothoracic index < 0.5 and unchanged lung fields). The electrocardiogram and the Doppler echocardiogram showed no changes. ECG stress testing revealed a rectified ST segment depression of up to 3 mm.

Myocardial perfusion scintigraphy revealed transient hypoperfusion of a moderate to severe intensity in the anterior wall and the apex of the left ventricle and mild to moderate hypoperfusion in the anterolateral wall, with preserved systolic function of the left ventricle.

The patient underwent cardiac catheterization with cineangiography, left ventriculography and aortography to elucidate the diagnosis. The procedure was performed by radial access using the Judkins technique. No changes in intracardiac pressure curves were noted. The right coronary artery was dominant and exhibited an increased diameter without significant atheromatosis. Extensive collateral intercoronary circulation was observed to originate from the right coronary sinus, which was anastomosed with the left coronary artery. The left coronary artery was not selectively catheterized but viewed in a retrograde mode (Figure 1). Ventriculography revealed no changes.

A 64-slice coronary computed tomography (CT) angiography was performed to confirm the diagnosis. The procedure was performed with non-ionic iodinated contrast agent injection and dose modulation. A coronary calcium score of zero, coronary circulation with right dominance, and a left coronary artery originating from the pulmonary artery and measuring 7.3 mm in diameter with no atheromas or luminal narrowing were observed. The anterior descending artery bypassed the apex of the left ventricle and was diffusely ectatic, measuring up to 6.8 mm in diameter with no apparent atheromas or luminal narrowing. The circumflex artery, which is of great importance, lacked atheromas or luminal narrowing and exhibited ectasia in its proximal third, measuring up to 6 mm in diameter. The right coronary artery originated in the typical manner from the right coronary sinus and was diffusely ectatic, measuring up to 8 mm at its largest diameter. No atheromas or luminal narrowing was noted. A marked tortuosity was also observed in its middle portion (Figure 2). The dimensions and biventricular systolic functions of the heart chambers were preserved, and intracavitary thrombi and valvular calcification were lacking. The pericardium exhibited no signs of thickening. Communication was observed between the right and left coronary arteries, resulting in reverse flow of the latter. The reverse

Figure 1. Cineangiography demonstrating extensive intercoronary collateral circulation originating from the right coronary sinus, which was anastomosed with the left coronary artery. The left coronary artery was not selectively catheterized but viewed in a retrograde mode.

Figure 2. (A and B) Coronary CT angiography. The left main coronary artery (LMCA) originated from the pulmonary artery and measured 7.3 mm in diameter. The left anterior descending artery (LAD) surrounded the apex of the left ventricle and measured up to 6.8 mm in diameter. The left circumflex artery (LCx), which is of great importance, exhibited ectasia in its proximal third and measured up to 6 mm in diameter. The right coronary artery (RCA) originated from the right coronary sinus and measured up to 8 mm at its largest diameter.
flow initiated from the distal portions of the smaller branches of the left coronary artery to the proximal third of the anterior descending and circumflex artery followed by the left coronary artery and opening in the pulmonary artery.

After the diagnosis of anomalous origin of the left coronary artery from the pulmonary artery was made, initial clinical treatment with 5 mg nebivolol hydrochloride was indicated due to the stability of the patient’s symptoms, and the patient refrained moderate- and high-impact physical activities. Surgery was successful with redirection of coronary blood flow by means of longitudinal pulmonary arteriotomy and occlusion of the left coronary artery ostium with bovine pericardial patch and internal mammary artery graft placement. The patient recovered well and is asymptomatic.

Discussion

The anomalous origin of the coronary artery from the pulmonary artery (ALCAPA) was first described in 1908 in a 60-year-old patient. In 1933, Bland et al. reported this syndrome in a 3-month-old child. The present case report presents the most common form of ALCAPA, in which the left coronary artery originates from the pulmonary artery and the right coronary artery arises from the aorta, characterizing the BWG syndrome. In the prenatal period, pulmonary artery pressure is equal to the systemic pressure due to the nonrestrictive ductus arteriosus, and oxygen concentrations are similar due to the parallel circulation. However, after birth, as the circulation becomes serial, pulmonary arterial pressure and resistance decrease, as does pulmonary blood flow oxygen content. With its huge demand for oxygen, the left ventricle is perfused with low-pressure desaturated blood. This condition predisposes the patient to myocardial ischemia, especially during exertion. Further increases in myocardial oxygen consumption can lead to infarction of the anterolateral wall of the left ventricle, mitral valve papillary muscle dysfunction and variable degrees of mitral regurgitation.

Clinical manifestations, which stem mostly from the extent of myocardial ischemia, are directly proportional to the development of collateral circulation between the right and left coronary arteries. When the collaterals are adequate, symptoms may be absent or insignificant and allow growth to adulthood. The clinical picture is generally nonspecific without appreciable manifestations but may include fatigue, night dyspnea, syncope, arrhythmias and less frequent angina. Generally, the diagnostic hypothesis of ALCAPA may arise when there is a systolic murmur due to dysfunction of the mitral valve or a continuous murmur due to coronary pulmonary fistula blood flow. Some patients develop heart failure.

Oligosymptomatic or asymptomatic patients are rare in adulthood, i.e., those without ventricular dysfunction and mitral regurgitation. Generally, this patient presents with stenosis of the left coronary artery, which allows blood flow from the right coronary artery to permeate the left coronary arteriolar system before reaching the left coronary artery and draining at the pulmonary artery. This reason, collateral circulation may not be evident on angiography. Often, even stress tests, such as scintigraphy or ECG stress test, may be negative. This situation is exceptional and has a favorable outcome when treated clinically.

ECG may reveal ischemic changes, especially in the lateral wall, or sequelae of myocardial infarction. Scintigraphy, echocardiography and MRI may identify chronic ischemia signs in stress tests. Recently, the gold standard for diagnosis has become the multiple detector coronary CT angiography. Catheterization is of limited use for this purpose given its invasive characteristics. The advent of coronary CT angiography synchronized with the electrocardiogram has made it possible to trace the origin, course and termination of anomalies originating from the coronary arteries non-invasively and with greater efficiency.

The literature indicates that initial treatment should be surgical, which comprises the reestablishment of double coronary perfusion, either by anastomosis of the artery itself in the aorta or by myocardial grafts. Otherwise, evolution is rather unfavorable with high mortality, especially in childhood. In adults, the most common procedure is ligation of the left main coronary artery in the pulmonary artery combined with internal mammary artery or saphenous vein graft placement. Surgical prognosis is directly related to the myocardial condition at initial diagnosis together with the patient’s clinical response. The longer the time before diagnosis or surgical treatment is, the greater the heart damage due to ischemia, ventricular dysfunction and degree of mitral regurgitation.

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None.

Conflicts of interest

The authors declare no conflicts of interest.

References