Residência RP Pediátrica
Publicação Oficial da Sociedade Brasileira de Pediatria

Submitted on: 06/18/2017
Approved on: 10/16/2017

CASE REPORT

Anomalous origin of the left coronary artery from the pulmonary artery: A case report

Angélica Luciana Nau[1], Alan Fernandes Strapasson[2], Guilherme da Silva Martins[2], Cristiane Nogueira Binotto[3]

Keywords:
Heart Defects, Congenital,
Infant,
Coronary Disease.

Abstract

Introduction: Anomalous origin of the left coronary artery (ALCAPA) is a rare cardiac defect, which causes myocardial ischemia and cardiomyopathy. Case report: male, 7 months, failure to thrive, pallor, sweating, irritability, and systolic murmur in the mitral area. Echocardiogram revealed anomalous origin of the left coronary artery from the pulmonary artery, and cardiac catheterization corroborated the diagnosis. The child was submitted to surgical correction, with good postoperative evolution. **Discussion:** ALCAPA has difficult diagnosis. It usually manifests before 2 months of age, but the symptoms can be misinterpreted and the disease underdiagnosed. The electrocardiogram may show changes in the ST segment or Q wave, suggestive of acute myocardial ischemia. Diagnosis can be made by two-dimensional echocardiogram, with direct visualization of the abnormal origin of the left coronary artery, associated with the hemodynamic study. Treatment is surgical, with redeployment of the coronary artery in the aorta, and in general, the evolution is satisfactory.

[1] Resident Physician in Pediatrics.
[2] Medical Student.
[3] PhD in Child and Adolescent Health; Pediatric Cardiologist and Echocardiographer.

Correspondence to:
Angélica Luciana Nau.
Hospital Pequeno Príncipe. Desembargador Motta, 1070 - Água Verde, Curitiba, Paraná, Brasil. - CEP 80250-060.

INTRODUCTION

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), also known as Bland–White–Garland syndrome[1], is a rare cardiac defect observed in 0.25%–0.5% of patients with congenital heart disease. This condition causes myocardial ischemia, and it may be progressive and result in ischemic cardiomyopathy. Congestive heart failure generally occurs within the first 2 months of life[1,2]. It may occur in isolation or be associated with other defects, such as patent ductus arteriosus, atrioventricular septal defect, tetralogy of Fallot, and coarctation of the aorta[1].

An abnormal coronary artery has a relatively low myocardial perfusion pressure and oxygen content owing to its origin from the pulmonary artery. Following birth, with the transition from intrauterine to extrauterine circulation, the collateral vessels withdraw blood from the myocardium into the pulmonary artery, leading to left–right shunt, ischemia, myocardial dysfunction, and not rarely, neonatal death[2].

This report aimed to describe the case and outcome of a male infant diagnosed with ALCAPA in reference to the literature on the subject.

CASE REPORT

A 7-month-old male presented with a history of impaired growth since the age of 5 months. During a routine consultation with a pediatrician, irregular heart rhythm was observed, which raised the suspicion of heart disease. The parents reported pallor, excessive sweating, and irritability, while denying other symptoms. The pathological history of the patient did not present relevant information; the parents denied comorbidities, previous surgeries, use of medications, or hospitalization. The patient had an up-to-date vaccination schedule; food history and psychoneuromotor development were appropriate for his age. Maternal obstetric history was as follows: $G_2P_1A_1$ and a miscarriage in the second month of the previous gestation. The mother had attended seven prenatal consultations and undergone seven ultrasounds and one morphology ultrasound. The patient was born at term through cesarean section due to pelvic presentation; his weight was appropriate for his gestational age (Apgar 10/10). The mother and child were discharged after 48 h without complications. Family history did not disclose any information relevant to the case.

Initial physical examination revealed weight of 6,900 g (below the 5th percentile for age), HR of 134 bpm, RR of 26 bpm, temperature of 37.4°C, oxygen saturation in ambient air of 96%, good general condition, without pallor, acyanotic, hydrated, and afebrile. Blood pressure was not measured at the first examination. On cardiac auscultation, heart sounds were found to be rhythmic and normal, without accessory sounds and with a systolic murmur of 1+/6+ in the mitral area. The liver was palpable at a distance of 2 cm from the right costal margin. Further, limb edema was not observed, and other physical aspects were normal.

Chest radiography revealed an overall increase in the cardiac area in the left cavity and normal lung transparency. Electrocardiography showed sinus rhythm with left ventricular hypertrophy, diffuse changes in ventricular repolarization, and deep Q wave in D1 and aVL. Further, echocardiography indicated ALCAPA (dilated right coronary artery), dilated cardiomyopathy, moderate left ventricular systolic dysfunction, enlarged left chambers, and significant mitral insufficiency (Figure 1). Laboratory tests revealed hypochromic and microcytic anemia, leukocytosis (18,839 cells/µL; differential count without particularities), and an erythrocyte sedimentation rate of 18 mm/1 h. CK-MB presented a normal value (18.00 U/L). Other tests, including platelet count; liver and renal function; and C-reactive protein, liver enzyme, and electrolyte levels, were within the normal limits.

After echocardiographic diagnosis, treatment with 0.03 mg/kg/day carvedilol, 0.78 mg/kg/day captopril, and 1 mg/kg/day furosemide was initiated; a hemodynamic study was requested, which indicated ALCAPA (Figure 2). On the 4th day of hospitalization, 0.01 mg/kg/day digoxin administration was initiated, and the patient who was having a bronchospasm crisis was stabilized for surgery. On the 13th day of hospitalization, the left coronary artery was reimplanted into the aorta with an autologous pericardial patch owing to a short pulmonary trunk and mitral valve repair. He was referred to the Cardiac Intensive Care Unit for postoperative stabilization. He presented a normal postoperative electrocardiogram, which showed mild residual mitral regurgitation, enlarged left chambers, and mild left ventricular systolic dysfunction (ejection fraction: 28%). The patient showed satisfactory progression and was extubated after 5 days. After postoperative 9 days, the patient was transferred to the general ward. He was prescribed digoxin, furosemide, enalapril, and spironolactone, and he evolved with progressive improvement of cardiac function. The patient was discharged after postoperative 14 days.

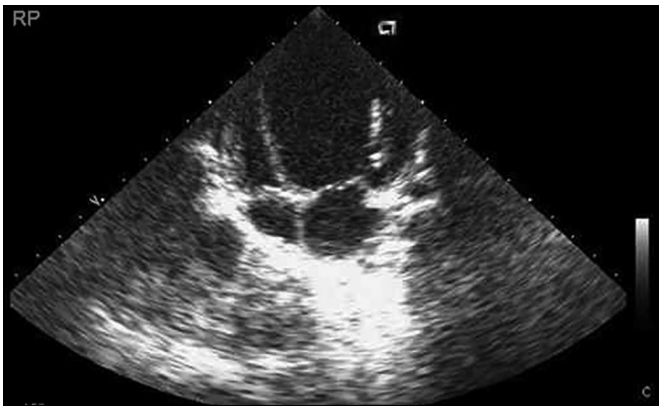


Figura 1. Cardiac catheterization shows the left coronary artery originating from the pulmonary artery.

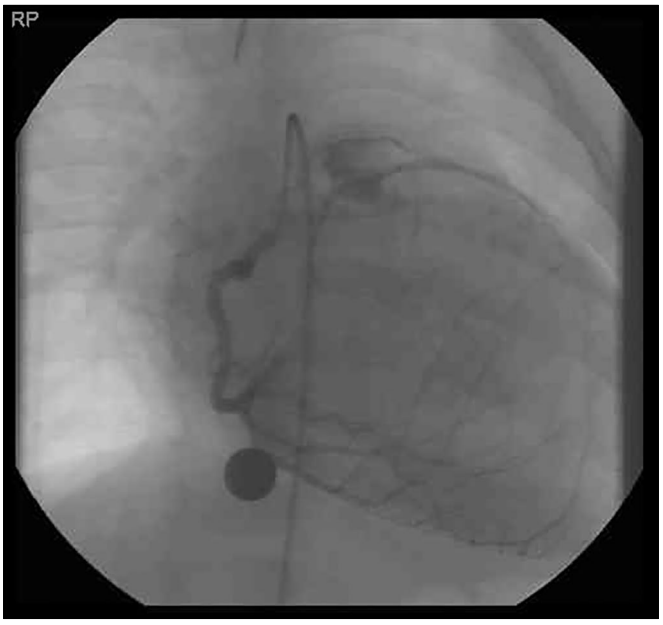


Figura 2. Two-dimensional echocardiogram showing left ventricular dilated cardiomyopathy and increased echogenicity of the left papillary muscles.

DISCUSSION

ALCAPA is difficult to diagnose and should be suspected in children with dilated cardiomyopathy[3]. Approximately 85% of cases manifest by up to 2 months of age, but the symptoms can be misinterpreted and the disease may be underdiagnosed[4]. Patients usually show signs of shock, and physical examination reveals gallop rhythm or mitral regurgitation murmurs due to papillary muscle dysfunction caused by myocardial ischemia. A continuous murmur suggesting patent ductus arteriosus flowing from the aorta to the pulmonary artery (left–right shunt) may be indicative of flow through the collateral arteries[2].

The clinical manifestations presented by the current patient such as sweating, pallor, irritability, intense crying, and low weight gain are consistent with those described in the literature and correlate with left–right shunt and consequent heart failure due to ventricular dysfunction, mitral regurgitation, myocardial infarction, and malignant dysrhythmias[1,2,5-7]. It is important to note that some patients present systolic murmur due to mitral insufficiency caused by poor valve perfusion. As in the present case, this finding may be the clinical sign of suspicion and reason for referral to a cardiologist[7].

Complementary diagnostic tests include chest radiography, electrocardiography, echocardiography, and cardiac catheterization, with the gold standard test being coronary angiography[2]. The findings of cardiomegaly and electrocardiographic tracing suggesting acute myocardial infarction with changes in the ST segment or Q wave in D1, aVL, V5, and V6 are suggestive of ALCAPA[1,8]. In children, diagnosis can usually be made by two-dimensional echocardiography with direct visualization of the abnormal origin of the left coronary artery and retrograde flow to the pulmonary artery in addition to mitral regurgitation and increased echogenicity of the papillary muscles of the mitral valve[4]. Echocardiography is also an essential test in the evaluation of ejection fraction, especially in postoperative evolution[9]. Other possible tests included angiotomography and magnetic angioresonance of coronary arteries; in practice, a hemodynamic study is preferred for assessment of patients[10]. Levels of myocardial enzymes, such as CK-MB or troponin, may increase, but they have no diagnostic role[1,10].

This condition requires surgical treatment that restores the flow of oxygenated blood to the myocardium. The procedure of choice comprises coronary artery reimplantation into the aorta[9]. Mitral valve repair can be performed on the same surgical occasion; this technique is recommended because these patients present valve dysfunction[2]. Other options include coronary artery tunneling to the aorta (Takeuchi repair) or aortocoronary bypass with the internal mammary artery or saphenous vein[10,11]. Surgical mortality is estimated to be less than 5%–10%. After reimplantation of the anomalous left coronary artery, the prognosis is often excellent[10].

In the preoperative period, patients are stabilized with diuretics and occasionally with inotropic and anti-hypertensive agents in addition to routine procedures, such as infusion of blood components, according to the clinical need[5.] Postoperative management should be performed in the intensive care unit; it usually requires administration of inotropic agents, afterload reducing agents, and diuretics as well as mechanical ventilation[5,10]. In the intermediate postoperative period, ejection fraction improvement may occur, and some mild residual mitral insufficiency may remain[9]. Patient follow-up should include periodic evaluation of ventricular function and possible obstruction of the coronary reimplantation[12].

The clinical picture of congestive heart failure in the first 1–2 months of life with signs of tachycardia, diaphoresis, refusal to eat, and low weight gain warrant cardiological investigation. Due to failure in early clinical recognition of this condition, the present patient's diagnosis was late, leading to a delay in the search for a referral service. Despite the heart failure condition, the patient was treated in accordance with the techniques and recommendations described in the literature, resulting in a satisfactory outcome.

REFERENCES

1. Mancini MC. Anomalous left coronary artery from the pulmonary artery. Medscape [Internet journals]. 2017 Jan [accessed on 22 March 2017]. Available at: http://emedicine.medscape.com/article/893290-overview.
2. Koenig PR, Hijazi ZM, Graham TP, Triedman JK SG. Congenital and pediatric coronary artery abnormalities. enig PR, Hijazi ZM, Graham TP, Triedman JK. In: UpToDate, Post TW (Ed), UpToDate, Waltham, MA. 2010.
3. Gribaa R, Slim M, Ben Salem H, Neffati E, Boughzela E. Anomalous origin of the left coronary artery from the pulmonary artery presenting as dilated cardiomyopathy: a case report. J Med Case Rep 2014;8:170.
4. Kristensen T, Kofoed KF, Helqvist S, Helvind M, Søndergaard L. Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) presenting with ventricular fibrillation in an adult: a case report. J Cardiothorac Surg 2008;3:33.

5. Younus Z, Iftikhar R, Iftikhar A. Anomalous origin of left coronary artery from pulmonary artery (ALCAPA). J Coll Physicians Surg Pakistan 2013;23(10):743–4.
6. Modi H, Ariyachaipanich A, Dia M. Anomalous origin of right coronary artery from pulmonary artery and severe mitral regurgitation due to myxomatous mitral valve disease: a case report and literature review. J Invasive Cardiol 2010;22(4):49–55.
7. Rodriguez-Gonzalez M, Tirado AM, Hosseinpour R, de Soto JS. Anomalous origin of the left coronary artery from the pulmonary artery: diagnoses and surgical results in 12 pediatric patients. Tex Heart Inst J 2015;42(4):350–6.
8. Bakiler AR, Eliaçik K, Köse S AY. Anomalous origin of the left coronary artery from the pulmonary artery presenting as dilated cardiomyopathy. Arch Turk Soc Cardiol 2013;41(5):448–50.
9. Muzaffar T, Ahmad Ganie F, Gpoal Swamy S, Wani N-U-D. The surgical outcome of anomalous origin of the left coronary artery from the pulmonary artery. Int Cardiovasc Res J 2014;8(2):57–60.
10. Mancini MC. Surgical approach to anomalous left coronary artery from the pulmonary artery. Medscape. 2015 May [accessed on 05 April 2017]. Available at: http://emedicine.medscape.com/article/905229-overview.
11. Lenzi AW, Solarewicz L, Ferreira WS, Sallum F, Miyague NI. Análise da técnica de Takeuchi usada para o tratamento da origem anômala da artéria coronária esquerda do tronco pulmonar. [An analysis of the Takeuchi technique used to treat the anomalous origin of the left coronary artery from the pulmonary trunk] Arq Bras Cardiol 2008;90(3):185–90.
12. Yuan S-M. Anomalous origin of coronary artery: taxonomy and clinical implication. Braz J Cardiovasc Surg 2014;29(4):622–9.