EUROMEDITERRANEAN BIOMEDICAL JOURNAL
2022,17 (23) 108-110
(*FORMERLY: CAPSULA EBURNEA*)

Case report

ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY: A CASE REPORT

Mirella Petrisano [1], Maria Scavone [2], Costantino Smaldone [3], Aurora Dinatale [2], Giambattista Gallicchio [2], Giorgio Madonna [2], Daniela Concolino [1], Antonio Sisto [2], Simona Pesce [2]

1. Pediatric Unit, Department of Science of Health, University Magna Graecia, Catanzaro, Italy
2. Neonatal Intensive Care Unit, San Carlo Hospital, Potenza, Italy
3. Cardiovascular Department, San Carlo Hospital, Potenza, Italy

ARTICLE INFO

Article history:
Received 20 Jun 2022
Accepted 25 Aug 2022
Published 15 Sep 2022

Keywords:
ALCAPA, congenital coronary artery anomaly, heart failure

ABSTRACT

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital disease in which the left coronary artery arises from the main pulmonary artery resulting in myocardial dysfunction. Myocardial ischemia in infancy results in nonspecific clinical symptoms, and therefore imaging performs a crucial role in the diagnostic process. Echocardiograms may reveal the anomaly, even if there are indirect radiological or ECG signs. Prompt surgical intervention to reestablish a two–coronary artery circulation allows gradual myocardial recovery nevertheless, the mortality rate from this condition is dramatically high if it is not recognized early. We present the case of a newborn referred to the neonatal intensive care unit on the 3rd day of life because of sudden skin marbling and tachycardia. The baby underwent several diagnostic tests which detected ALCAPA. He received surgical correction of the condition and started extracorporeal membrane oxygenation (ECMO), despite this he become hemodynamically unstable and expired.

1. Introduction

ALCAPA is a life-threating condition in which left coronary artery arises from the main pulmonary artery (Figure 1). It was first described by Brooks in 1886 and is also known as the Bland-White-Garland syndrome.[1] It occurs in one in 300,000 live births and accounts for 0.25-0.5% of newborns with congenital heart disease, leading in a mortality rate of up to 90% during the first year of life.[2] This cardiac defect progressively leads to severe ventricular hypoperfusion and dysfunction. The standard of care for this syndrome is a prompt surgical correction at the time of diagnosis with the aim of restoring a two-coronary flow and gradually recover myocardial perfusion.[3]

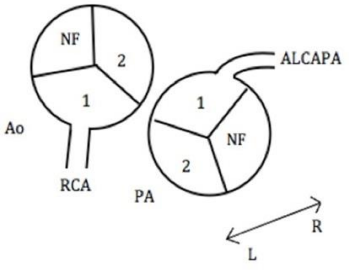

Figure 1. Sinus numeration in anomalous left coronary artery. Ao= Aorta, PA= pulmonar artery, NF= Non facing sinus, 1,2 =sinuses of the pulmonar valve RCA= right coronary artery, L= left, R= right, ALCAPA= Anomalous Origin of the Left Coronary Artery From the Pulmonary Artery. (Modified from Dodge-Khatami A, et al. Ann Thorac Surg. 2002 Sep; 74(3):946-55)

* *Corresponding author*: Mirella Petrisano, *mirella.petrisano@gmail.com*
DOI: 10.3269/1970-5492.2022.17.23
 ISSN: 2279-7165 - Available on-line at www.embj.org

2. Case Report

F.A., a full-term male infant born via caesarean section, was admitted to the neonatal intensive care unit on the 3rd day of life because of sudden skin marbling and tachycardia with subsequent onset of cyanosis, polypnea and oliguria. Antenatal ultrasonographic studies did not reveal any pathologies. Apgar scores were 9-10-10 at 1, 5, and 10 minutes, respectively. The birth weight was 3860 g (adequate for gestational age). At admission initial blood gas analysis showed: pH 6.9, pCO_2 72 mmHg, lactates 13.7 mmol/L, HCO_3^- incalculable, BE incalculable, then he required endotracheal intubation. Laboratory examination revealed the following: white blood cells 13.03×10^9/L, neutrophils 9.81×10^9/L, hemoglobin 14.4 g/dL, platelets 256×10^9/L; liver panel showed mildly elevated aspartate transaminase at 136 IU/L, no abnormality shown on renal function tests or electrolytes, CRP was 5.3 mg/dL. Blood exams revealed elevated cardiac troponin at 3846 ng/mL, CK-MB 58.9 IU/L. The ECG showed normal sinus rhythm and echocardiogram revealed depressed biventricular function and dilated right ventricle but it was not possible to investigate the origin of the left main coronary artery. He was given IV inotropic support to improve cardiac contractility. Suspecting myocarditis, virological exams were performed which were negative, as well as blood cultures. After receiving informed consent from the parents, metabolic investigations were performed on the patient but neither did these tests reveal any possible etiology.

A chest radiograph was performed, not revealing pathological findings. Finally, subsequent repeated echocardiograms revealed anomalous origin of left coronary artery from the pulmonary artery (Figure 2), so he was transferred to the cardiac intensive care unit for surgical correction. He underwent surgery and was placed on extracorporeal membrane oxygenation (ECMO). Postoperatively, the baby became progressively hemodynamically unstable and finally expired.

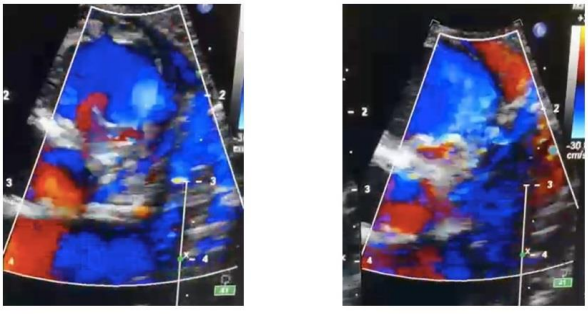

Figure 2. Parasternal long axis view. The figures show the retrograde flow from the left coronary artery (LCA) into the pulmonary artery.

3. Discussion

ALCAPA is a rare but clinically significant form of ischemic congenital heart disease representing one of the most common causes of myocardial infarction in children.

Embryologically, ALCAPA may result from abnormal separation of the conotruncus into the aorta and pulmonary artery (PA), or persistence of the pulmonary buds in conjunction with involution of the aortic buds so that the left coronary artery (LCA) and left heart receive blood from the PA.[4] Edwards first described the pathophysiology of the syndrome: during fetal life, high pulmonary vascular resistance and pulmonary artery pressure facilitates antegrade flow from the pulmonary trunk into the anomalous LCA.[5] The clinical presentation of the syndrome starts after the transition from fetal to neonatal life when pulmonary vascular resistance decreases leading to lower pulmonary artery pressure compared to left coronary artery pressure. Consequently, there is less antegrade blood flow to the LCA resulting in low left myocardial ventricular perfusion because of its dependence on intercoronary collaterals or right coronary artery. The coronary artery steal and the consequent myocardial ischemia leads to severe left ventricular dilation and dysfunction, mitral annular dilation with a worsening mitral insufficiency. Patients are classified on the basis of their survival patterns into infantile or adult type. In particular, according to a review of described cases, this disease presents clinically in four different ways: in infancy with angina-like symptoms or as cardiomyopathy, later as mitral insufficiency, continuous murmur, or by sudden death.[6] Smith et al. proposed a nomenclature relating to the sinus of origin of the anomalous coronary artery, using the standard designation 1 for the sinus of origin of the right coronary artery and 2 for the sinus from which a normal left main coronary artery would originate. The main diagnostic tool is echocardiography, showing an enlarged right coronary artery, a dilated left ventricle with global hypokinesia; color-flow Doppler reveals a left to right shunt from the anomalous LCA into the PA.[7] There should be indirect signs that suggest cardiac anomalies such as ALCAPA, represented by respiratory distress that does not change with appropriate therapy, chardiomegaly at chest X-ray and ischemic changes on electrocardiography. The main differential diagnosis is dilated cardiomyopathy, which usually shows as progressive heart failure, cardiomeghaly and cardiac ischaemia. Johnsrude et al. defined electrocardiographic criteria to diagnosticate ALCAPA and distinguish this disease from myocarditis or dilated cardiomyopathy.[8] In their study, they found the following signs to be present in ECGs of all ALCAPA patients: a Q wave depth of more than 3 mm, a Q wave width greater than 30 ms, a QR pattern in at least one of leads I, aVL, V5–V7, the absence of Q waves in leads II, III, and aVF. Our patient revealed depressed biventricular function and dilated right ventricle on ECG, but no pathological Q wave was found.

Although ALCAPA is usually an isolated entity, it could be associated with other congenital anomalies, such as atrial and ventricular septal defect, patent arterial duct, aortic arch abnormality, and conotruncal heart malformations.[9] In our report the newborn did not show any associated malformation.

It is estimated that nearly 90% of untreated patients die in the first year of life (infancy type), primarily due to myocardial ischaemia and heart failure, while only few patients with adequate collateral circulation between the right coronary artery (RCA) to the LCA may survive beyond infancy (adult type). Therefore, a prompt diagnosis and timely surgical intervention are mandatory to ensure patient survival.

Several surgical corrections have been described in order to re-establish a two-coronary system in ALCAPA with a progressively lower operative mortality rate and an improvement in myocardial function and reperfusion.[10]

4. Conclusions

Clinicians should consider ALCAPA during the work up of global myocardial dysfunction, especially if no other cause is identified. Being able to recognize this condition early is important in order to perform an immediate surgical correction with the aim of restoring a two-coronary system circulation in order to reduce mortality. Further studies are needed to assess the best medical management before and after surgery, in order to allow more favorable results.

References

1. Brooks HS. Two Cases of an Abnormal Coronary Artery of the Heart Arising from the Pulmonary Artery: With some Remarks upon the Effect of this Anomaly in producing Cirsoid Dilatation of the Vessels. J Anat Physiol. 1885 Oct; 20(Pt 1):26-9.
2. Keith JD. The anomalous origin of the left coronary artery from the pulmonary artery. Br Heart J 1959; 21:149–61.
3. Karimi M, Kirshbom PM. Anomalous origins of coronary arteries from the pulmonary artery: a comprehensive review of literature and surgical options. World J Pediatr Congenit Heart Surg. 2015; 6:526–540.
4. Boutsikou M, Shore D, Li W, Rubens M, Pijuan A, Gatzoulis MA, Babu-Narayan SV. Anomalous left coronary artery from the pulmonary artery (ALCAPA) diagnosed in adulthood: Varied clinical presentation, therapeutic approach and outcome. Int J Cardiol. 2018 Jun 15; 261:49-53.
5. Edwards JE. Anomalous coronary arteries with special reference to arteriovenous-like communications. Circulation. 1958 Jun; 17(6):1001-6.
6. Cambronero-Cortinas E, Moratalla-Haro P, González-García AE, Oliver-Ruiz JM. Case report of asymptomatic very late presentation of ALCAPA syndrome: review of the literature since pathophysiology until treatment. Eur Heart J Case Rep. 2020 Sep 19; 4(5):1-5.
7. Patel SG, Frommelt MA, Frommelt PC, Kutty S, Cramer JW. Echocardiographic diagnosis, surgical treatment, and outcomes of anomalous left coronary artery from the pulmonary artery. J Am Soc Echocardiogr. 2017; 30:896–903.
8. Johnsrude CL, Perry JC, Cecchin F, Smith EO, Fraley K,m Friedman RA, Towbin JA. Differentiating anomalous left main coronary artery originating from the pulmonary artery in infants from myocarditis and dilated cardiomyopathy by electrocardiogram. Am J Cardiol 1995; 75:71–4.
9. Cowles RA, Berdon WE. Bland-White-Garland syndrome of anomalous left coronary artery arising from the pulmonary artery (ALCAPA): a historical review. Pediatr Radiol. 2007 Sep; 37(9):890-5.
10. Zhang C, Zhang H, Yan J, Hua Z, Song Y, Sun H, Li S. Mid-Term Outcome for Anomalous Origin of the Left Coronary Artery From the Pulmonary Artery. Heart Lung Circ. 2020 May; 29(5):766-771.