Online publication ahead of print, AJMHS Vol 65, 2024

Anomalous Origin of the Left Coronary Artery from Pulmonary Artery in an Asymptomatic Athletic Adolescent: A Case Report

Mimoza Maldi[1*], Cristina Carro[1], Simone Ghiselli[1], Altin Veshti[2], Stefano M. Marianeschi[1]

[1] Congenital Cardiac Surgery Unit, Grande Ospedale Metropolitano Niguarda, Milan, Italy
[2] Cardiac Surgery Unit, University Hospital Center "Mother Teresa", Tirana, Albania

Abstract

Background: Anomalous origin of the left coronary artery from the main pulmonary artery is a rare congenital anomaly associated with early infant mortality and adult sudden cardiac death. As it is mainly diagnosed in the first year of life, adulthood presentation is rare and associated to left ventricular dysfunction, significant mitral valve insufficiency and myocardial ischemia. Surgical correction is recommended as soon as diagnosis is made.

Case report: In this report we describe a case of an anomalous origin of the left coronary artery from the main pulmonary artery in an asymptomatic 14-year-old athletic patient diagnosed at our institution by coronary computed tomography. The patient underwent successful surgical repair of the coronary anomaly by a direct coronary reimplantation to the aorta soon after the diagnosis. At the time of this report, the patient is well and he is performing his athletic activity.

Conclusion: In both infants and adults, the establishment of a dual coronary system is the ideal surgical treatment for the anomalous origin of the coronary artery from pulmonary artery to provide progressive ventricular recovery and no furthermore damages.

Address for correspondence: Mimoza Maldi*, Congenital Cardiac Surgery Unit, Grande Ospedale Metropolitano Niguarda, Piazza Ospedale Maggiore 3, 20162 Milan, Italy. Email: moza.maldi@hotmail.com

INTRODUCTION

Anomalous origin of the left coronary artery from the main pulmonary artery (Alcapa syndrome) is a rare congenital anomaly associated with early infant mortality and adult sudden cardiac death. As it is mainly diagnosed in the first year of life, adulthood presentation is rare and associated to left ventricular dysfunction, significant mitral valve insufficiency and myocardial ischemia.

Surgical correction is recommended as soon as diagnosis is made. In both infants and adults, the establishment of a dual coronary system is the ideal surgical treatment leading to progressive ventricular recovery.

In this report we describe a case of Alcapa syndrome in an asymptomatic 14-year-old athletic patient diagnosed at our institution by coronary computed tomography. The patient underwent successful surgical repair of the coronary anomaly by a direct coronary reimplantation to the aorta soon after the diagnosis. At the time of this report, the patient is well and he is performing his athletic activity.

CASE REPORT

An athletic 14-year-old male underwent standard medical examination by sport physician. He had no cardiovascular risk factors and denied any cardiovascular symptoms. His physical examination was unremarkable.

Nevertheless, an exercise stress electrocardiogram revealed inferolateral ST depression in lead V6. Therefore, he performed a Holter electrocardiogram that detected rare ectopic ventricular heart-beats. The 2-dimensional echocardiography showed left ventricular moderate hypertrophy associated to a preserved function and a trivial mitral valve regurgitation. Cardiac magnetic resonance imaging (MRI) detected a spongy left ventricle (LV) with hypokinesis of the apical septal wall.

Finally, the patient was referred to our institution for overall diagnosis.

We performed a coronary computed tomography (CT) that detected an anomalous origin of the left coronary artery (LCA) from the posterior aspect of the main pulmonary artery (PA) (Figure 1). LCA was dilated (maximum transverse diameter of 5 mm) and it coursed toward interventricular groove branching into the left anterior descending (LAD) and circumflex artery (CX), both enlarged. A markedly dilated (maximum transverse diameter of 7 mm) and tortuous right coronary artery (RCA) with correct origin was also detected such as multiple and ectasic intercoronary collateral arteries.

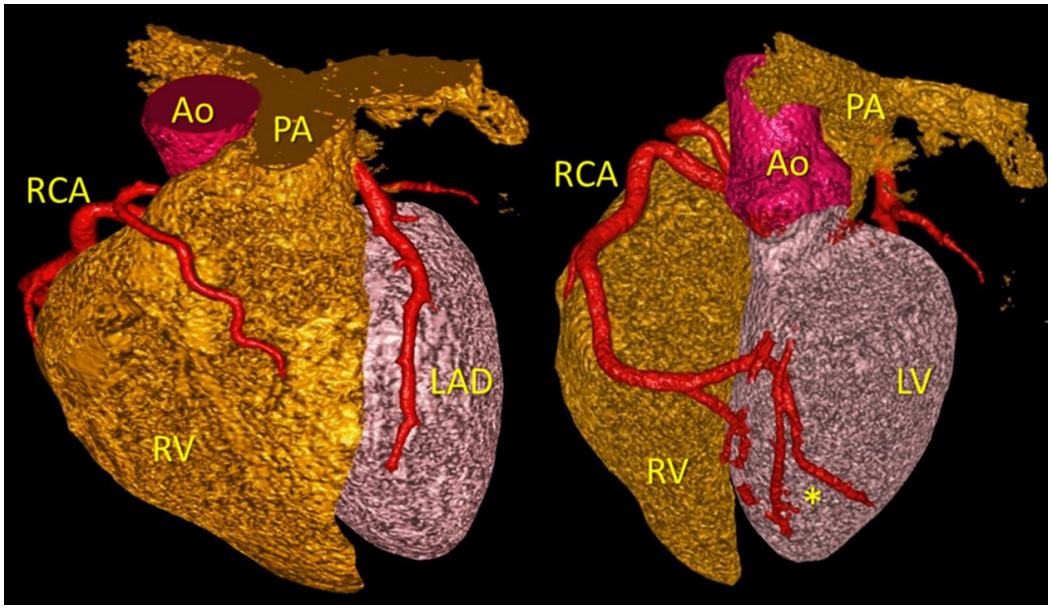

Figure 1. Preoperative coronary computed tomography: anomalous origin of the left coronary artery from the posterior aspect of the main pulmonary artery.
*(Ao: aorta; PA: pulmonary artery; RCA: right coronary artery; RV: right ventricle; LV: left ventricle; LAD: left arterial descending; * eterocoronary circulation).*

The patient underwent a coronary angiography that confirmed the diagnosis of Alcapa syndrome and demonstrated an important left-to-right shunt from the LCA into the pulmonary artery trunk, a retrograde filling through collaterals arising from an enlarged RCA to an ectasic LAD and CX arteries (Figure 2).

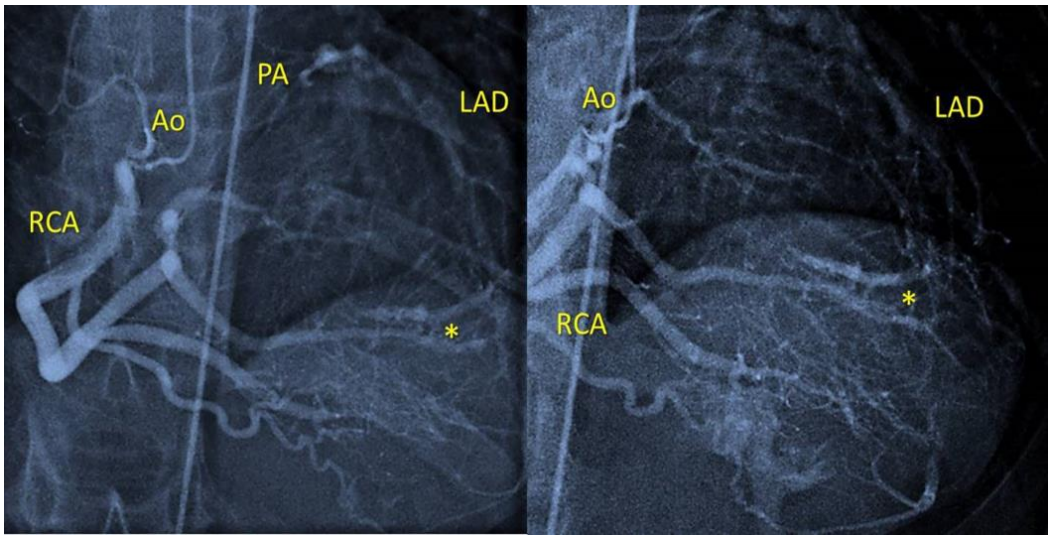

Figure 2. Preoperative coronary angiography: coronary angiogram shows the retrograde filling of the LAD through collaterals arising from the tortuous RCA; the LAD is connected directly to the PA
*(Ao: aorta; PA: pulmonary artery; RCA: right coronary artery; LAD: left arterial descending; * eterocoronary circulation).*

Consequently, the patient underwent surgical repair by a median sternotomy, cannulation of the aorta, two venae cavae separately and a vent in the left atrium. Before starting cardiopulmonary bypass, the snaring of pulmonary branches with tourniquets was realized. After aortic cross clamp, cold blood cardioplegia was injected in both aorta and pulmonary trunk. The PA was transacted and the anomalous artery arising from the posterior sinus of the PA was completely mobilized. A direct aortic re-implantation of the coronary bottom in the postero-lateral side of the aorta with a trapdoor incision was performed. Finally, the main pulmonary artery was reconstructed with a bovine pericardial patch.

The post-operatory course was unremarkable.

The echocardiogram realized in the early postoperative period and a coronary CT performed 3 months after surgery, detected a correct result of the surgical repair with a preserved left ventricular function.

At the time of this report, the patient is in good clinical conditions and went back to his intensive sport activity after a stress test showing a complete recovery of the LV.

DISCUSSION

Alcapa syndrome or Bland-White-Garland syndrome is a rare congenital defect accounting for 0.25-0.5% of congenital heart disease (1).

Eighty-five percent of all cases of Alcapa syndrome present within the first 2 months of life when pulmonary artery pressure drops. Usual presentation involves severe left ventricular failure and mitral regurgitation due to papillary muscle ischemia. Initial symptoms are feeding difficulties, diaphoresis, tachypnea and tachycardia. Without treatment, 90% of infants die within the first year of life (2).

Most rarely, Alcapa syndrome presents in adults and usually with important left ventricular dysfunction, severe mitral regurgitation or sign of myocardial ischemia. At the time of presentation, the majority of patients have symptoms of angina, dyspnea and palpitations, particularly on exertion; Alcapa syndrome may be an important cause of sudden cardiac death, particularly in young athletes.

Indeed, malignant arrhythmias could be triggered from an acute ischemic event during exercise, where coronary steal phenomena may cause inadequate myocardial perfusion.

A high incidence of sudden death at young age has been demonstrated and only 18% of these patients experienced symptoms prior to sudden death (3). Older patients, defined by age 18 years and older, experience less frequent life-threatening presentations and sudden death (4).

Moreover, symptoms may be misinterpreted or may even be absent.

Survival into adulthood and clinical course depend on coronary collaterals development from RCA to LCA; death ensues if collaterals are poorly developed.

Our patient was completely asymptomatic living active life, including important athletic activities, because of extensive and sufficient collateral blood supply from the RCA. However, this

supply often does not adequately perfuse the LV, especially in the subendocardial region; a chronic ventricular subendocardial ischemia can then develop, causing malignant ventricular arrhythmias.

In the last years, cardiac imaging investigations have increased the number of diagnosed cases of adolescents or adult patients with silent clinical history.

Traditionally, Alcapa syndrome was detected by angiography; nowadays, the development of non-invasive imaging allows the diagnosis and European Society of Cardiology recommends coronary CT angiography as gold standard for diagnosis of these abnormalities (5).

Frequently, diagnosis of Alcapa syndrome can also be realized by 2-dimensional echocardiography. However, since the echo window does not usually permit the LCA visualization with the classical Alcapa syndrome pattern, this diagnostic method is of difficult application.

Once the diagnosis is established, a prompt surgical repair is mandatory for all patients.

However, some authors suggest that in asymptomatic adults with only moderate chronic ischemia and limited necrosis, survival is possible without surgery. Kandzari reported a case of a 72-year-old woman with an uncorrected Alcapa syndrome because the risk-benefit ratio of surgery was not sure (6).

The aim of surgery is to restore a 2-coronary system circulation that has a lower morbidity and mortality versus a one-coronary system.

Several surgical treatment options have been proposed: coronary bottom transfer with direct aortic re-implantation, intrapulmonary baffling such as the Takeuchi technique or ligation of the LCA associated to a coronary artery bypass graft (7). Hao Luo et al. report a case of an ALCAPA in a 55-year-old female, for the correction of whom they used a Gore-Tex 6 mm artificial blood vessel (8). In our patient, the distance between the origin of the anomalous artery and the aorta associated to an adequate coronary dissection, allowed a direct coronary bottom re-implantation to the aorta.

Since prognosis depends on left ventricular function and presence of myocardial scar tissue, early surgical repair usually results in an excellent prognosis in infants, adolescents and adult patients.

Acknowledgements: None declared.

Conflict of Interest Statement: The author declares that have no conflict of interest.

REFERENCES

1. Tutar, E., A. Uysalel, N. Nacar, T. Kendirli, A. Aral, E. Ince, et al., Anomalous origin of the left coronary artery from the main pulmonary artery detected on echocardiographic screening study of newborns. Int J Cardiol 2004. 97(3):561-2.

2. Wesselhoeft, H., J.S. Fawcett, and A.L. Johnson, Anomalous origin of the left coronary artery from the pulmonary trunk. Its clinical spectrum, pathology, and pathophysiology, based on a review of 140 cases with seven further cases. Circulation 1968. 38(2): 403-25.

3. Jurishica, A.J., Anomalous left coronary artery; adult type. Am Heart J 1957. 54(3): 429-36.

4. Yau, J.M., R. Singh, E.J. Halpern, and D. Fischman, Anomalous origin of the left coronary artery from the pulmonary artery in adults: a comprehensive review of 151 adult cases and a new diagnosis in a 53-year-old woman. Clin Cardiol 2011. 34(4): 204-10.

5. Schroeder, S., S. Achenbach, F. Bengel, C. Burgstahler, F. Cademartiri, P. de Feyter, et al., Cardiac computed tomography: indications, applications, limitations, and training requirements: report of a Writing Group deployed by the Working Group Nuclear Cardiology and Cardiac CT of the European Society of Cardiology and the European Council of Nuclear Cardiology. Eur Heart J 2008. 29(4): 531-56.

6. Kandzari, D.E., J.K. Harrison, and V.S. Behar, An anomalous left coronary artery originating from the pulmonary artery in a 72-year-old woman: diagnosis by color flow myocardial blush and coronary arteriography. J Invasive Cardiol 2002. 14(2): 96-9.

7. Dodge-Khatami, A., C. Mavroudis, and C.L. Backer, Anomalous origin of the left coronary artery from the pulmonary artery: collective review of surgical therapy. Ann Thorac Surg 2002. 74(3): 946-55.

8. Luo, H., O.E. Kwaku, Y. Lai, and R. Yue, Adult-type anomalous origin of the left coronary artery from the pulmonary artery and right coronary-right atrial fistula: a case report. BMC Cardiovasc Disord 2024. 24(1): 31.