Single Coronary Artery in Transplanted Heart: An Unusual Association

Thaís Rossana Cruz de Souza,[1] Arthur Luiz Norte Gomes Pereira Araruna,[1] Cláudia do Nascimento Silva Fernandes,[4] Roberto Moreno Mendonça,[2,5] Epitácio de Oliveira Belém Neto,[3] Robson Macedo[2]

Universidade Potiguar;[1] Instituto de Radiologia de Natal;[2] CLINICOR;[3] Hospital do Coração de Natal;[4] Centro de Ciências da Saúde/UFRN,[5] Natal, RN – Brazil

Introduction

Coronary anomalies (CA) are rare and may be classified according to the origin, pathway and destination. Course anomalies are further classified into five subtypes: interarterial, subpulmonary, pre-pulmonary, retroaortic or retrocardiac.[1] Anomalies with an interarterial pathway are characterized by a pathway between the aorta and pulmonary arterial trunk, commonly described as a "malignant pathway" due to an increased risk of sudden death,[2] although the prevalence and its absolute risk are still unknown.[1]

AC can be found in 0.3 – 5.6% of the population and are associated with morbidity and mortality in young adults.[2] The retroaortic pathway anomaly is the most common subtype, with prevalence estimated at 0.28%. As for interarterial anomalies, the anomalous origin of the anterior descending artery (ADA) associated with a malignant pathway is rare. Its prevalence is estimated to be 0.03%, while the anomalous origin of the right coronary artery (RCA) associated with malignant pathways is 0.23%.[1] ADA and circumflex artery (Cx) with independent origins are not often found. ADA with anomalous origin is usually associated with congenital heart diseases. Cx is the one that most commonly presents an anomalous origin, occurring in 0.32 – 0.67% of the population. The single coronary artery (SCA) is extremely rare, accounting for 2% to 4% of all CA, being found in 0.0024% to 0.066% of coronary angiographies. In this condition, only one coronary artery originates in the aorta, being responsible for supplying blood to the whole heart.[3] Currently, computed tomography angiography (CTA) or magnetic resonance imaging (MRI) are considered the gold standard for demonstrating coronary anatomy.[2]

The objective of this study is to describe a rare case of malignant CA diagnosed in a patient after cardiac transplantation, besides reviewing the main aspects related to diagnosis and management, whose approach and epidemiology remain uncertain.

Case Report

JMSL, a 44-year-old woman with Chagas' cardiomyopathy with severe ventricular dysfunction (ejection fraction: 16%). She underwent orthotopic cardiac transplantation, progressing steadily under use of immunosuppressants. She underwent endomyocardial biopsy, with no evidence of organ rejection. After three years, she started to have angina, of moderate intensity, irradiating the back, causing it to feel tight, triggered by exertion and improving at rest. Echocardiogram demonstrated asynchrony of septal contraction with no signs of rejection. Transluminal coronary angioplasty (TCA) revealed ADA and Cx originating in the right aortic sinus and interarterial and retroaortic pathways, respectively, without obstructive lesions (Lipton R-III-C classification) (Figures 1, 2, 3 and 4). The patient was conservatively managed with diltiazem, mycophenolate and cyclosporin, progressing with good clinical response.

Discussion

CA in patients with transplanted hearts are incidental and extremely rare. When found, its consequences must be must understood, seeking objective evidence of myocardial ischemia for therapeutic definition.[4]

Transluminal coronary angioplasty and MRI are considered Class I for CA diagnosis.[5] This choice depends on multiple factors, such as local experience and availability, as well as the intrinsic advantages and disadvantages of the method. In many centers, TCA is preferable due to the speed, high spatial resolution and lower cost. Although it uses iodinated contrast and involves exposure to radiation, dose reduction strategies and technical advances have resulted in increased safety.[1] Its anatomical and coronary configuration and other cardiac structures make it a promising diagnostic method for this application. MRI, on the other hand, provides an image of the coronary artery without the need for radiation or iodinated contrast. However, it has a lower spatial resolution, longer scan times and higher cost.[1] It provides a better delineation between lumen and vascular wall, allowing more precise parietal measurement. In addition to providing excellent anatomical information, MRI can also provide functional information by means of flow analysis techniques, for example. It also assesses the presence of inflammatory changes in the arterial walls.[6]

The classification proposed by Lipton for SCA divides these CAs into "R", when they originate from the right coronary sinus, and "L," from the left. These groups are subdivided into types I, II and III. In type I, a single vessel follows the normal pathway of a coronary artery and, through collaterals, it irrigates the contralateral territory. In type II, a coronary artery arises from the proximal portion of the contralateral coronary, of normal origin, crossing the base of the heart, until it reaches its normal distribution. In type III, the ADA and Cx are born separately in the proximal part of a normal right coronary artery (R-III). There are five anatomical subtypes classified according to their relationship with aorta and pulmonary artery: 'anterior', 'between', 'septal', 'posterior' or 'combined'.[3] The case described in this article falls in the R-III-C classification.

Keywords

Chagas Cardiomyopathy; Coronary Artery Disease/physiopathology; Heart Transplantation/complications; Angina Pectoris; Echocardiography.

Mailing Address: Thaís Rossana Cruz de Souza •
Rua Lafayete Lamartine, 1880. Postal Code 59064-510, Candelária, Natal, RN – Brazil
E-mail: thaisrossana1@gmail.com
Manuscript received June 3, 2017; revised October 12, 2017; accepted December 14, 2017

DOI: 10.5935/2318-8219.20180013

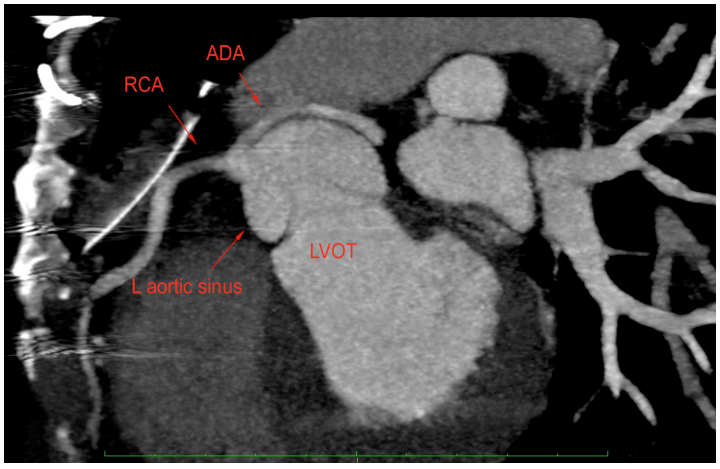

Figure 1 – *Maximum intensity projection (MIP) in oblique section revealing the LCA and the ADA originating from the single coronary trunk that emerges from the right aortic sinus.*

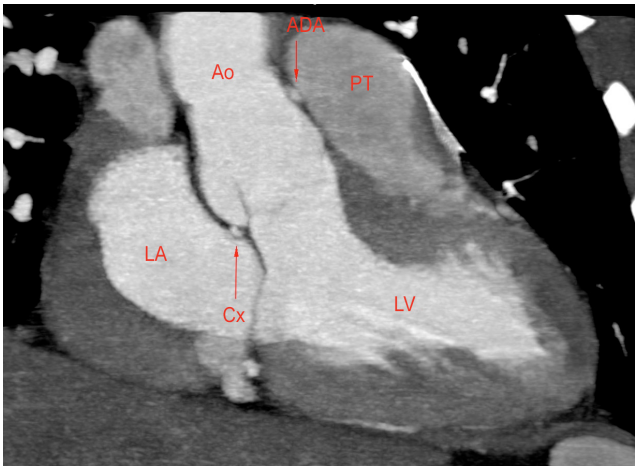

Figure 2 – *"Three-chamber" section in MIP revealing ADA interaterial location and Cx retroaortic location.*

Anomalous left coronary artery arising from the right aortic sinus with pathway between the aorta and the pulmonary trunk (malignant pathway) presents a risk of fatal cardiac arrhythmia.[7] Since CA is rare in the general population, and cardiac donors are often young, less likely to have coronary heart disease, coronary angiography or TCA are not routine in pre-transplant evaluation. However, surgeons try to catch exuberant atherosclerosis through palpation and inspect the heart in order to identify CA upon collection.[4] The natural history of CA, as well as the lack of a reliable method to predict sudden death, justify the rejection of a donor heart with this anomaly. However, the surgical correction of the anomalous pathway or bypass at the time of transplantation makes the acceptance of a donor with anomalous origin of the left coronary artery a relevant alternative. The long waiting time for an organ has recently led to a review of what can be considered an acceptable donor.

In the general population, management recommendations for patients with CA are based on age and presence of symptoms, such as syncope or chest pain. In symptomatic patients, surgical management is recommended. Patients with cardiac grafts who are diagnosed with CA during follow-up should be managed similarly to the non-transplanted population.[7]

According to the recommendations of the American College of Cardiology and American Heart Association (ACC/AHA) published in 2011, coronary artery bypass grafting is indicated (class I) for: 1) anomalous origin of left coronary trunk with interarterial pathway; 2) anomalous origin of RCA with interarterial pathway associated with evidence of myocardial ischemia; 3) evidence of myocardial ischemia in the territory of the anomalous coronary artery without another evident causal factor.[8]

The aforementioned patient started to have symptoms of angina 3 years after transplantation and its ischemic

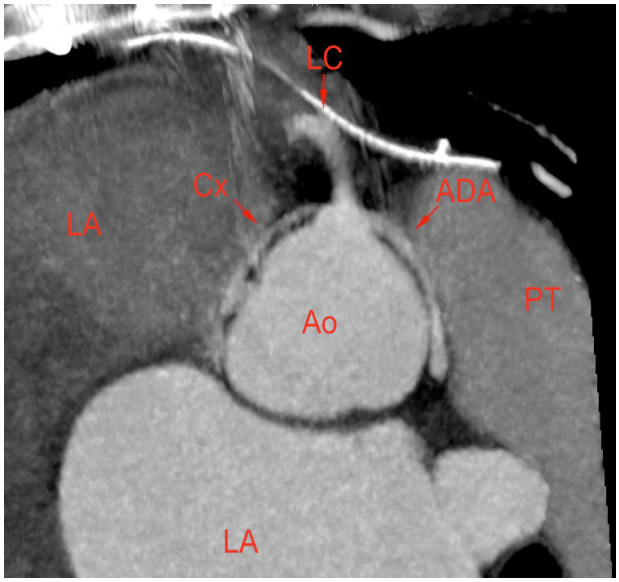

Figure 3 – *Axial section of aortic root in IPM revealing the single coronary trunk emerging from the right aortic sinus, immediately trifurcating in ADA, RCA and Cx, as well as the interarterial course of the ADA proximal segment and Cx retroaortic orientation.*

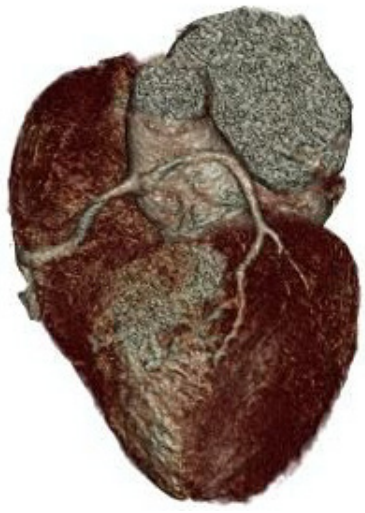

Figure 4 – *Volumetric reconstruction of the origin of the right aortic sinus single coronary trunk with immediate trifurcation after its origin.*

nature was not confirmed by complementary method. Once the transplanted patient is denervated, typical anginal symptoms may not be present and the first manifestation of the disease may be sudden death. However, it is believed that the partial reinnervation of the transplanted heart begins in 1 year in a gradual and regionally heterogeneous process.[9,10] In this case, the onset of anginal symptoms after more than 1 year generates uncertainty as to its origin and relationship with ischemia.

The main late postoperative complications to be considered include graft vascular disease (GVD), responsible for most of the deaths after the first year of follow-up, and early diagnosis is also limited by denervation, obscuring anginal symptoms. With this, most of the protocols recommend, after the first year, an annual angiographic study to detect the disease.

Because it is a noninvasive and detailed evaluation of the coronary anatomy, TCA can be considered for screening, diagnosis, stratification and follow-up of GVD.[9]

Due to the rarity of the case, there are no studies comparing outcomes of surgical correction of CA in patients transplanted under clinical treatment. Surgical treatment could minimize the deleterious effects caused by CA, but due to the scarcity of data in the literature, long-term management of transplanted patients with CA should be individualized, considering the risks, benefits and particularities of each case.

Authors' contributions

Data acquisition: Araruna ALNGP, Souza TRC, Fernandes CNS, Macedo R, Mendonça RM, Belém Neto EO; Data analysis and interpretation: Araruna ALNGP, Souza TRC, Fernandes CNS, Macedo R, Mendonça RM, Belém Neto EO; Manuscript drafting: Araruna ALNGP, Souza TRC, Fernandes CNS, Macedo R, Mendonça RM, Belém Neto EO.

Potential Conflicts of Interest

There are no relevant conflicts of interest.

Sources of Funding

This study had no external funding sources.

Academic Association

This study is not associated with any graduate program.

References

1. Cheezum KM, Liberthson R, Shah R, Vellines TC, O'Gara PT, Landzberg MJ, et al. Anomalous aortic origin of a coronary artery from the inappropriate sinus of valsalva. J Am Coll Cardiol. 2017;69(12):1592-608. Doi:10.1016/j.jacc.2017.01.031
2. Neves PO, Andrade J, Monção H. Coronary anomalies: what the radiologist should know. Radiol Bras. 2015;48(4):233–41. Doi:10.1590/0100-3984.2014.0004
3. Guérios EE, Andrade PMP, Melnick G, Barbosa Neto DS. Anomalia coronária única. Rev Bras Cardiol Invasiva. 2010;18(2):226-30. Doi: 10.1590/S2179-83972010000200019
4. Abudiab MM, Hakim FA, Fortuin DF. Anomalous coronary artery in a transplanted heart: a rare incidental diagnosis. J Saudi Heart Assoc. 2016;28(1):46-8. Doi:10.1016/j.jsha.2015.06.001
5. Sara L, Szarf G, Tachibana A, Shiozaki AA, Villa AV, Oliveira AC, et al. II Diretriz de ressonância magnética e tomografia computadorizada cardiovascular da Sociedade Brasileira de Cardiologia e do Colégio Brasileiro de Radiologia. Arq. Bras. Cardiol. 2014;103(6 supl 3):1-86. Doi:10.5935/abc.2014S006
6. Filho RM, Mendonça RM. Ressonância magnética e doenças não ateroscleróticas da aorta. In: Moreira MCV, Montenegro ST, Paola AAV. Livro texto da Sociedade Brasileira de Cadiologia. 2ª ed. Barueri(SP): Manole;2015.p.696-9.
7. Vasseur G, Anderson MB, Freudenberger S. Anomalous coronary artery in a transplanted heart: a technical modification. J Heart Lung Transplant. 2004;23(8):1008-9. Doi: 10.1016/S1053-2498(03)00234-1
8. Hillis LD, Smith PK, Anderson JL,Bitti JA, Bridges CR, Byrne JG, et al. 2011 ACCF/AHA guideline for coronary artery bypass graft surgery: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines: developed in collaboration with the American Association for Thoracic Surgery, Society of Cardiovascular Anes- thesiologists, and Society of Thoracic Surgeons. J Am Coll Cardiol. 2011;58:e123–210. Doi:10.1016/j.jacc.2011.08.009
9. Bacal F, Souza-Neto JD, Fiorelli AI, Mejia J, Marcondes-Braga FG, Mangini S, et al. II Diretriz Brasileira de Transplante Cardíaco. Arq Bras Cardiol.2009;94(1 supl.1):e16-e73. http://dx.doi.org/10.1590/S0066-782X2009002000001
10. Tallo FS, Guimarães HP, Carmona MJ, Bianco ACM, Lopes RD, Teles JMM. Manual de perioperatório de cirurgia cardíaca da AMIB. São Paulo: Atheneu; 2012.