Anomalous Origin of the Coronary Artery from an Inappropriate Coronary Sinus

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Abstract

Congenital anomaly of the coronary arteries is a potentially lethal disease, especially in young people. Currently, non-invasive cardiovascular imaging tests have shown better definition of the origin and course of the coronary arteries. Surgical treatment should be indicated in symptomatic patients, while the ideal management of asymptomatic patients remains controversial. We report a case of anomalous origin of the left coronary artery from the right coronary sinus with an interarterial course, in an asymptomatic patient treated with revascularization surgery.

Introduction

Congenital anomaly of the coronary arteries can be benign or potentially severe, causing myocardial ischemia, infarction, and sudden death. The incidence varies between 0.3% and 1.5% in necropsy studies and coronary cineangiography. The origin and proximal course of the anomalous coronary arteries are the main factors predicting severity.

This anomaly represents the second most frequent cause of sudden death of cardiovascular origin in competitive athletes. Its diagnosis is difficult, as the individual may be asymptomatic until before the lethal event. The surgical techniques used to treat this pathology have evolved; however, there is no consensus regarding the most appropriate treatment for each type of patient.

In the case of anomalous origin of the left coronary artery with an interarterial course, surgical revascularization is recommended, regardless of documented ischemia or symptoms.

The objective of this study is to report an unusual case of anomalous origin of the left coronary artery from the right coronary sinus with an interarterial course in an asymptomatic patient.

Keywords

Coronary Vessel Anomalies; Sudden Death; Myocardial Revascularization.

Case report

We report the case of a 47-year-old male patient, who walked and ran 3 times a week. The patient, who denied angina, dyspnea, syncope, or palpitations, sought his physician for routine exams. He denied systemic arterial hypertension, diabetes, or smoking. The physical examination was normal. Electrocardiogram was normal, and the patient had normal blood pressure. He underwent an exercise test that revealed electrocardiographic changes during exercise compatible with an ischemic response pattern, without complaints of angina, with a normal blood pressure response, without arrhythmia, and a good workload (10 METs).

Doppler echocardiography was normal. Given these exercise abnormalities, myocardial perfusion scintigraphy was requested to assess the presence of myocardial ischemia.

Myocardial perfusion scintigraphy was performed under the Bruce protocol reaching stage IV, lasting 10 minutes and 6 seconds, reaching 10 METs, and 97% of the age-predicted maximum heart rate. The blood pressure curve was normal; the patient did not report chest pain, and the electrocardiographic tracing during exercise revealed a descending ST depression of 2 mm in CM5 and 1 mm in CM5 during recovery (Figure 1).

The perfusion images, analyzed qualitatively and quantitatively using Wackers-Liu software (Yale University), were normal, and the circumferential profiles of the images showed normal distribution (Figure 2). Left ventricular ejection fraction was also normal, without segmental contractile dysfunction.

Due to the inability to clarify the electrocardiographic changes induced by exercise, coronary tomography angiography was requested (Figures 3, 5, 6, 7, 8, 9, 10 and 11). The coronary calcium score was zero. The dominance was right; the left coronary trunk was absent. The anterior descending artery was non-dominant, originating from the right coronary sinus in an independent ostium; following a course between the aorta and the pulmonary artery trunk toward the anterior wall of the left ventricle; subsequently, it followed the anterior interventricular sulcus along the usual course. The circumflex artery was very important, originating in the right coronary sinus in an independent ostium, following a retroaortic course, passing between the aorta and the left atrium and continuing through the left atrioventricular sulcus in the usual course. The right coronary artery was the dominant artery, reaching the posterior interventricular sulcus, where it gave rise to a posterior descending branch.

Preoperative coronary cineangiography was performed, which confirmed the tomography angiography findings, namely, proximal systolic narrowing of the anterior descending artery during its interarterial course; it did not reveal...
Figure 1 – Electrocardiographic tracing obtained during Myocardial Perfusion Scintigraphy. A 2mm downsloping ST segment depression is observed at the J point in lead CMS, along with ventricular repolarization changes in the inferior and anterolateral walls. Image provided by Eduardo Lins Paixão.

Figure 2 – Images from the stress and rest Myocardial Perfusion Scintigraphy performed using the 1-day protocol with Tc99m-MIBI. Normal tracer distribution is observed in the walls of the left ventricle. There are no signs of post-stress transient left ventricular dilation. Image provided by Eduardo Lins Paixão.
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Figure 3 – Image A: 3D reconstruction of a coronary angiography (AngioCT-3D), identifying the origin of the RCA, LAD, and LCx from the right coronary sinus; Image B: AngioCT-3D showing the interarterial course of the LAD; Image C: AngioCT-3D displaying the retroaortic course of the LCx; Image D: Interarterial course of the LAD; Image E: Retroaortic course of the LCx; Images F and G: Interarterial course of the LAD. RCA: Right Coronary Artery; LAD: Left Anterior Descending; LCx: Left Circumflex. Image provided by Eduardo Lins Paixão.

Obstructive arterial disease (Figure 4). The patient underwent surgical revascularization of the anterior descending artery through the anastomosis of the left internal mammary artery. The surgical procedure took place approximately 6 months after diagnosis, without complications. The patient resumed his physical activities and is asymptomatic.

Discussion

Anomalous origin of the coronary artery is a rare congenital heart disease. In cases of anomalous origin of the coronary artery, the coronary artery originates from the coronary sinus inappropriately, either through a separate or common ostium or through a sub-branch. There are several subtypes; in the interarterial subtype, the coronary artery follows a course between the aorta and the pulmonary artery, presenting a potential risk of sudden death. A comprehensive review published by Cheezum et al. suggests that the true prevalence of this abnormality is likely underestimated. In this series, 39% of the sample with some type of anomalous origin of the coronary artery had the interarterial subtype. There are few studies that have screened for anomalous origin of the coronary artery in the absence of clinical indications.

In current guidelines, coronary tomography angiography and cardiac magnetic resonance angiography are the only methods with a class I indication. In many centers, tomography angiography is the preferred method for this diagnosis. Invasive angiography allows for greater spatial and temporal resolution, and it has a class IIa indication for diagnosing anomalous coronary artery. Functional tests, such as exercise testing and myocardial perfusion scintigraphy, analyze the functional significance of the anomalous origin on coronary flow.

Grani et al. examined 46 adults (mean age of 56 years) diagnosed with anomalous origin of coronary arteries by tomography angiography, and they identified perfusion changes only in patients with concomitant coronary artery disease.

Physical stress is preferable to assess myocardial perfusion, since most cases of sudden death attributed to anomalous origin of the coronary arteries occur with strenuous exercise. Several publications have reported a large number of false positives and negatives with these methods. Among 27 young athletes with anomalous origin of the coronary arteries described by Basso et al., 6 patients had a normal exercise test before presenting sudden death. Therefore, the absence of ischemia during stress testing cannot be seen as sufficient.

This clinical case followed current recommendations. Even in the absence of documented ischemia, due to the interarterial course of the anterior descending coronary artery, revascularization was indicated.

The objective of this study is to report an unusual case of anomalous origin of the left coronary artery from the right coronary sinus with an interarterial course in an asymptomatic patient. It is necessary to reflect on this diagnosis in young patients with symptoms of angina and syncope on exertion.
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Figure 4 – Cinecoronariography: In image 1, the left anterior descending coronary artery originating from the right coronary sinus with an interarterial course is evident; highlighted (arrow) is the vessel diameter in diastole, within the interarterial segment (between the aorta and pulmonary arteries). In image 2, the vessel diameter in systole is observed, showing extrinsic compression of the vessel. Image 3 displays the normal right coronary artery, while image 4 illustrates the origin of the right coronary, circumflex, and left anterior descending arteries, all arising from the right coronary sinus. Imagens cedidas por Eduardo Lins Paixão.

Figure 5 – 3D reconstruction, in a transverse section with a craniocaudal view, showing the right coronary sinus, from which the right coronary artery originates and follows its normal course, through the right atrioventricular sulcus. Note also the circumflex artery, which originates from the right coronary sinus, in an independent ostium; follows a retroaortic course, passing between the aorta and left atrium; and subsequently follows the left atrioventricular sulcus in the usual course. CX: circumflex artery; RCA: right coronary artery.
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Figure 6 – Coronary tomography angiography: 2D reconstruction, in a multiplanar section, showing the pulmonary artery, the aorta, and, between them, the image of the anterior descending coronary artery. Posterior to the aorta, the circumflex artery can be seen. ADA: anterior descending artery; AO: aorta; CX: circumflex artery; PA: pulmonary artery.

Figure 7 – Coronary tomography angiography: 3D reconstruction, in a transverse section with a craniocaudal view, showing the pulmonary artery, the aorta, and the anterior descending artery, which has an anomalous origin in the right coronary sinus, following an interarterial course and subsequently following the usual course through the anterior interventricular sulcus. ADA: anterior descending artery; AO: aorta; PA: pulmonary artery.
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Figure 8 – Coronary tomography angiography: 3D reconstruction, showing the right coronary sinus and the common origin of the right coronary, circumflex, and anterior descending arteries in this sinus. ADA: anterior descending artery; CX: circumflex artery; RCA: right coronary artery.

Figure 9 – Coronary tomography angiography: 2D reconstruction, in a sagittal section, showing the pulmonary artery, the aorta, and, between them, the image of the anterior descending coronary artery. ADA: anterior descending artery; AO: aorta; PA: pulmonary artery.
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Figure 10 – Coronary tomography angiography: 2D reconstruction, in a sagittal section, showing the circumflex artery in its retroaortic course, subsequently following the usual course. AO: aorta; CX: circumflex artery.

Figure 11 – Coronary tomography angiography: 2D reconstruction, in a transverse section with a craniocaudal view, showing the pulmonary artery, the aorta, and the anterior descending artery, which has an anomalous origin in the right coronary sinus and follows an interarterial course. ADA: anterior descending artery; AO: aorta; PA: pulmonary artery; RCA: right coronary artery.
Nonetheless, the diagnostic challenge is enormous, considering that a large portion of these patients do not have symptoms before sudden death; this fact requires more research to find a cost-effective means of assessment, particularly in individuals who practice intense competitive sports.

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Author Contributions

Conception and design of the research and statistical analysis: Paixão EL; acquisition of data: Couto MC, Lages DB, Paixão EL; analysis and interpretation of the data: Couto MC, Pacífico FA, Lages DB, Santos LS, Farias MA, Paixão EL; obtaining financing: Couto MC, Lages DB, Santos LS, Farias MA, Paixão EL; writing of the manuscript: Couto MC, Lages DB, Santos LS, Farias MA, Paixão EL; critical revision of the manuscript for intellectual content: Pacífico FA, Paixão EL.

References