Rare Presentation of an Anatomical Congenital Anomaly of the Coronary Arteries on Computed Tomography Angiography

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Introduction

Anomalous origin of the coronary arteries represents a heterogeneous group of alterations in the origin and course of these vessels, ranging from the most common, asymptomatic and low risk, to the rarest and highest risk, which may eventually be symptomatic or have sudden death as their first manifestation.[1, 2] Their prevalence corresponds to 1.3% of patients undergoing coronary angiography, but this number may be higher depending on the diagnostic method and the population selected, representing the second leading cause of sudden death in young athletes, behind only hypertrophic cardiomyopathy.[3, 4]

Within this group, coronary anomalies with a single ostium are present in a smaller proportion of the population, approximately 0.004% to 0.098%. They are most often asymptomatic, but they may manifest with chest pain, dyspnea, palpitation, myocardial infarction, and sudden death.[5, 6] The presentation of this anomaly has worse prognosis in the presence of an interarterial vessel course (between the aorta and pulmonary trunk), an acute angle of origin, and atherosclerotic plaque; the latter change has a higher incidence in anomalous vessels on account of the greater stress on the arterial wall.[7,8] In addition to presenting in isolation, they may also be associated with other alterations such as cardiac fistulas[8] or a bicuspid aortic valve.[9]

There is still no consensus regarding prognosis, especially the risk of sudden death in rarer anomalies or their combinations. Accordingly, the decision to indicate an eventual surgical correction is challenging and should be individualized.[10]

We report an extremely rare case of a combination of coronary anomalies, where the anterior descending artery originated from the right coronary artery and followed an interarterial course at a low subpulmonary (intramural) level, while in the presence of a circumflex artery also originating from the right coronary artery and following a retroaortic course. This combination of two anomalies, both classified as low risk, constitutes an unusual anatomical variation.

Keywords

Coronary Vessel Anomalies; Coronary Vessels; Anatomic Variation; Computed Tomography Angiography; Chest Pain

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Clinical Presentation

A 45-year-old female patient came to the clinic seeking a third opinion.

A few months prior, following an episode of atypical chest pain, she had undergone several functional and anatomical cardiology exams. The patient brought with her a resting electrocardiogram, 24-hour Holter monitoring, transthoracic echocardiogram, myocardial perfusion scintigraphy with physical stress, and cardiac magnetic resonance imaging with pharmacological stress. None showed pathological changes or findings compatible with ischemia.

She had also undergone coronary computed tomography angiography in the investigation carried out during consultations with two other cardiologists. This exam revealed an anterior descending artery originating from the right coronary artery, with an interarterial course at the intramural subpulmonary level, as well as a circumflex artery originating from the right coronary artery with a retroaortic course (Figures 1, 2, and 3).

Since the diagnosis 3 months prior, she had been presenting occasional symptoms of palpitations and nonspecific malaise without an apparent relation to physical exertion. She did not report any new episodes of the chest pain that had led to the investigation.

The patient had no previous diseases and was not taking any medications continuously. She denied tobacco and alcohol use, and she performed aerobic physical activity weekly, in addition to consuming a balanced diet. Upon physical examination, no alterations were identified during cardiovascular assessment.

Discussion

This case report presented here is extremely rare. It consists of the combination of two anatomical variations that are considered low risk. There is no consensus on the treatment of a single coronary ostium, without signs of

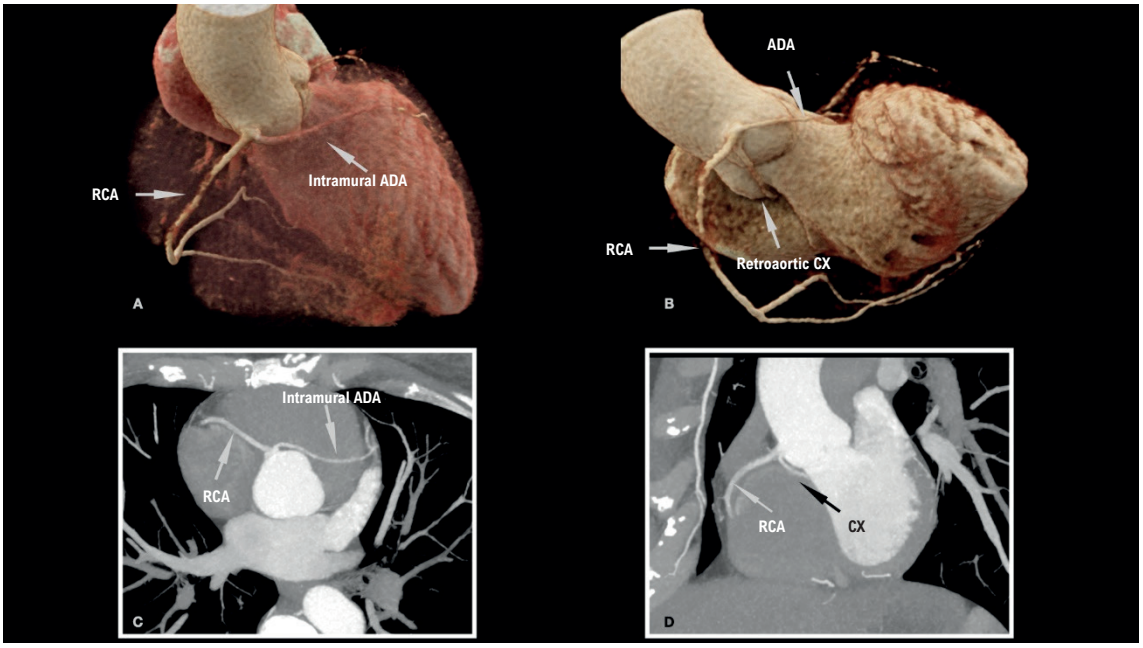

Figure 1 – *A) Three-dimensional reconstruction of the heart in anterior view, demonstrating the origin of the RCA and the origin of the ADA with a course anterior to the aorta. B) Three-dimensional reconstruction of the heart in right lateral view, demonstrating the origin of the RCA, the origin of the CX with a retroaortic course, and the origin of the ADA with a course anterior to the aorta. C) Axial reconstruction demonstrating the origin of the RCA and the origin of the ADA, highlighting the course anterior to the aorta and posterior to the intramural pulmonary trunk. D) Sagittal reconstruction demonstrating the origin of the RCA and the origin of the CX with a retroaortic course. ADA: anterior descending artery; CX: circumflex artery; RCA: right coronary artery.*

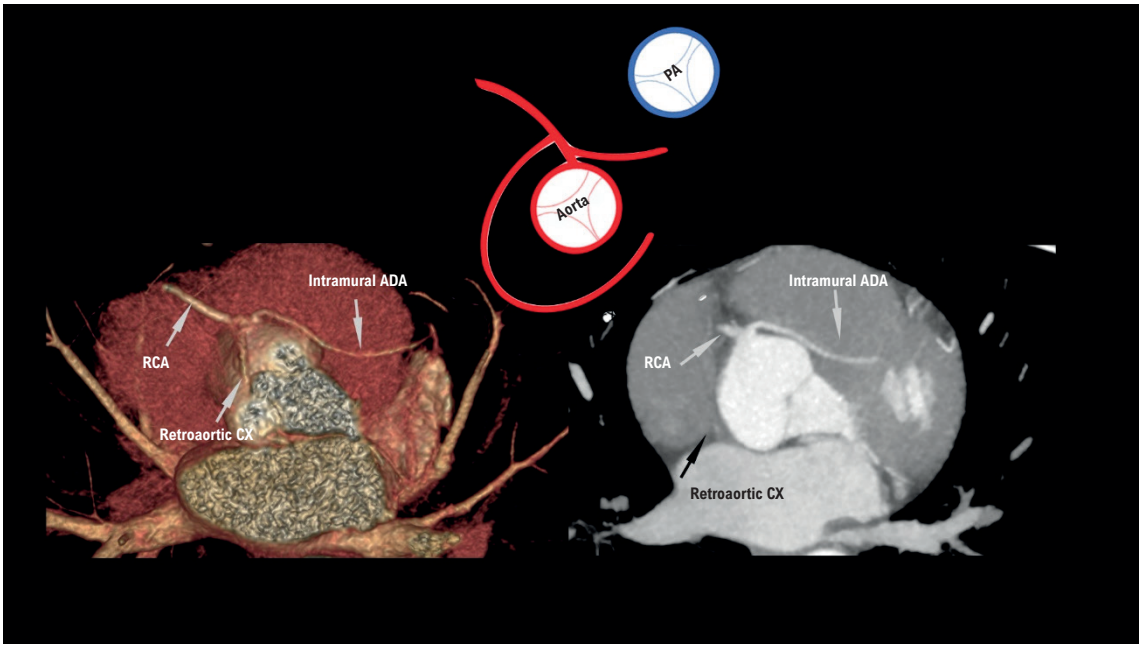

Figure 2 – *Comparison of the images in 3-dimensional and axial reconstruction and anatomical correlation with the graphical representation. ADA: anterior descending artery; CX: circumflex artery; PA: pulmonary artery; RCA: right coronary artery.*

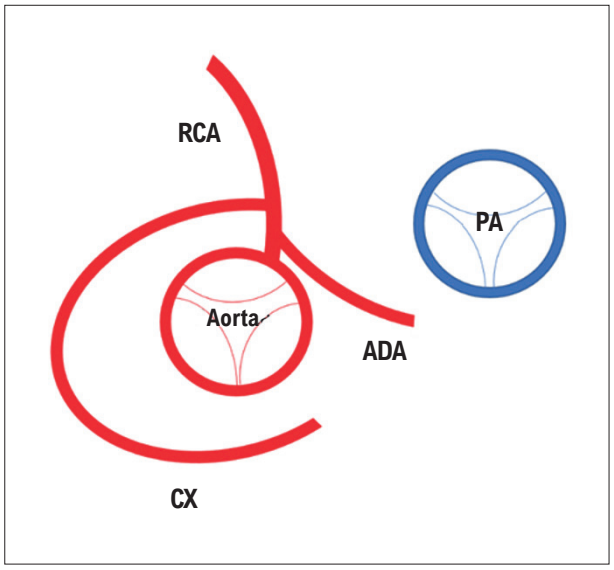

Figure 3 – *Graphical representation of the combination of anomalous origin of the LAD and CXA and their correlations with the aorta and pulmonary artery. ADA: anterior descending artery; CX: circumflex artery; PA: pulmonary artery; RCA: right coronary artery.*

ischemia, or of this variation associated with interarterial course. All provocative tests for ischemia performed did not show any changes, which leads to the presumption that the symptoms of palpitations, anxiety, and malaise were not related to this coronary anomaly.

The majority of anatomical variations of the coronary arteries are not symptomatic and do not require any type of treatment, be it conservative or surgical. Nonetheless, a small portion are associated with symptoms and a higher risk of sudden death, which are defined by cases of interarterial course, ostial stenosis, anomalous origin from the pulmonary trunk, and acute takeoff angle.[10] In these specific cases, interventions and corrections end up being necessary to reduce the risk.[11]

From an anatomical point of view, the main diagnostic methods are coronary computed tomography angiography and invasive coronary cineangiography; in selected cases and in specialized centers, coronary artery magnetic resonance imaging may also be used. For functional assessment, the main methods available are exercise testing, myocardial scintigraphy, stress echocardiography, and stress magnetic resonance imaging. Each procedure has specific characteristics, and it is the physician's responsibility to choose which form of investigation to use (Table 1).[12-14] It is important to underscore that invasive angiography has the greatest diagnostic accuracy[15] for evaluating coronary obstruction and was considered the gold standard for diagnosis for a long time. However, coronary computed tomography angiography has been rapidly evolving; it is capable of 3-dimensional evaluation of the vessel course and comparison with the underlying structures, and many consider it to be the new gold standard for diagnosis, in addition to being a non-invasive method.[6, 16] Coronary artery magnetic resonance imaging also has the advantage of demonstrating the anatomical relationship between cardiac structures and the anomalous vessel, in addition to using less contrast, but it has lower temporal and spatial resolution when compared to tomography angiography. Furthermore, it is less available, and its use is restricted in patients with pacemakers.[10]

There is currently a great deal of debate and uncertainty regarding the indication for surgical correction. The main algorithms involve the presence of symptoms or manifestations of ischemia and interarterial course, especially of the left coronary artery trunk.[17,18] Interarterial course can also be classified as suprapulmonary when the vessel is above the level of the pulmonary valve and infrapulmonary when it is below, the latter having the lowest risk.

Conclusion

This case highlights the complexity and rarity of congenital coronary artery anomalies, emphasizing the importance of detailed diagnostic assessment. The observed combination of the anterior descending artery originating from the right coronary artery with an intramural subpulmonary course, as well as the circumflex artery originating from the right coronary artery with a retroaortic course is rare, and the risk is uncertain according to the available literature. The management of these cases should be individualized, considering the absence of significant symptoms and a likely low risk associated with the specific anomaly. The decision for more invasive interventions should be cautious and based on rigorous criteria, for instance, the presence of clear symptoms of angina or documented ischemia. This case reinforces the need to individualize patient care and perform multidisciplinary assessment in order to determine the best therapeutic approach.

Author Contributions

Conception and design of the research, acquisition of data, writing of the manuscript and critical revision of the manuscript for intellectual content: Zapparoli M, Alessi A, Zapparoli FC, Alessi MR.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

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Ethics Approval and Consent to Participate

This article does not contain any studies with human participants or animals performed by any of the authors.

Table 1 – Diagnostic methods for investigating coronary artery disease

Diagnostic method	Advantages	Disadvantages
Anatomical imaging		
Coronary computed tomography angiography	Non-invasive technique Detects obstructive and non-obstructive coronary disease Direct visualization of coronary anomalies	Ionizing radiation Use of iodinated contrast Does not provide functional data Limited in non-ideal conditions (arrhythmia, obesity, excess coronary calcification)
Coronary cineangiography	Detects obstructive and non-obstructive coronary disease Direct visualization of coronary anomalies Permits physiological tests such as FFR and intravascular imaging such as IVUS/OCT Allows treatment of coronary lesions High accuracy	Ionizing radiation Use of iodinated contrast Invasive method
Functional imaging		
Exercise test	Non-invasive Low cost Physiological data (aerobic capacity, blood pressure behavior, and heart rate) High availability	Limited performance for detecting coronary disease in populations with low and intermediate risks Limited in patients with motor limitations
SPECT	Non-invasive technique Can be performed with pharmacological or physical stress Detection of ischemia and viability	Ionizing radiation Attenuation artifacts Contraindication to the stressor agent
Echocardiography (resting)	Non-invasive technique Can assess valvular disease, diastolic function parameters, pulmonary hypertension, and myocardial disease Can visualize CX anomaly (RAC sign)	Inappropriate acoustic window
Echocardiography (stress)	Non-invasive technique Can be performed with pharmacological or physical stress	Inappropriate acoustic window Contraindication to the stressor agent
Cardiac magnetic resonance imaging	Non-invasive technique Assessment of anatomy, function, myocardial perfusion, and myocardial viability	Claustrophobia Contraindication to the stressor agent Long acquisition time Accessibility Metallic implants or implanted devices

CX: circumflex artery; FFR: fractional flow reserve; IVUS: intravascular ultrasound; OCT: optical coherence tomography; RAC: retroaortic anomalous coronary artery; SPECT: single photon emission computed tomography.

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