Case Report

Anomalous Origin of the Right Coronary from the Left Coronary Sinus: How to approach?

Origem Anômala de Coronária Direita a partir do Seio Coronariano Esquerdo: como Conduzir?

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Introduction

Coronary artery anomalies (CAAs) are characterized by a changed vessel origin, path, or structure.[1] It is a rare condition, present in approximately 1% of the general population.[2] These anomalies include the anomalous aortic origin of a coronary artery (AAOCA), which can travel retroaortic, subpulmonic, prepulmonic, and inter-arterial courses.[3]

In general, these abnormalities remain asymptomatic until adulthood, being incidental findings on complementary tests or necropsy. However, they can also cause angina, syncope, ischemia, arrhythmias, or sudden death.[1,2] Sudden death can be the first presentation of this pathology in young athletes, representing the second leading cause of death in this group.[4]

This case report describes the case of a patient diagnosed with anomalous origin of the right coronary artery (RCA) after an episode of moderate-risk unstable angina and presents important details of the clinical management of this condition.

Case report

A 56-year-old man was hypertensive, pre-diabetic, sedentary, and obese and presented with dyslipidemia and a positive family history of coronary disease. The patient was admitted to the hospital to evaluate two episodes of prolonged moderate chest pain and tightness in the previous 24 hours without irradiation or associated factors that had already resolved on admission. There were no increased cardiac enzymes. An electrocardiogram showed sinus rhythm and signs of left ventricular overload with slight ventricular repolarization changes. The patient was stratified as having moderate-risk unstable angina and received a Thrombolysis in Myocardial Infarction score of 3 for non-ST coronary syndrome. Despite his initial clinical stability, the attending physician opted for coronary angiography when the patient was still in the hospital due to the typical characteristics of his pain and the presence of risk factors for coronary artery disease.

Keywords

Arteries; Coronary vessel anomalies; Coronary vessels; Diagnostic imaging.

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Cine coronary angiography showed the origin of the RCA in the left coronary sinus with a slit ostium, possible inter-arterial course (between the aorta and pulmonary arteries), and moderate ostial stenosis due to probable extrinsic compression (Figure 1A). Left coronary artery without stenosis (Figure 1B). Transthoracic echocardiography showed mild left atrium dilation, eccentric hypertrophy, grade I diastolic dysfunction, and preserved global and segmental systolic function of the left ventricle.

Computed tomography angiography of the coronary arteries confirmed the improper RCA origin in the left Valsalva sinus with an inter-arterial course and adjacent to the origin of the left main coronary artery (Figures 2 and 3). There was angulation at the origin (~23°), moderate proximal luminal reduction (slit-like orifice), and presence of calcified atherosclerotic plaque in the middle third of the RCA without reducing the lumen. The Agatston calcium score was 13, and coronary calcification was associated with a plaque in the middle segment of the RCA.

The patient was referred for myocardial scintigraphy, which showed normal myocardial perfusion and no signs of ischemia.

Maintenance clinical drug treatment without surgical intervention was chosen. The patient uses olmesartan medoxomil associated with amlodipine besylate (40/5 mg daily); indapamide (1.5 mg daily); spironolactone (25 mg daily); nebivolol hydrochloride (5 mg daily); MR trimetazidine dihydrochloride (70 mg daily); rosuvastatin (20 mg daily); XR metformin hydrochloride (1 g daily), and allopurinol (300 mg daily), and he remains asymptomatic with no new angina attacks at 1 year after the initial diagnosis. He has controlled systemic blood pressure, good glycemic and cholesterol levels, and no activity limitations.

Discussion

Although most CAAs are benign, not progressing with hemodynamic implications or influencing the patient's prognosis, their discussion is important due to their association with sudden death,[5] mainly in young athletes and usually triggered by rigorous physical exercise.[6] AAOCA comprises a portion of CAA cases, being subdivided into left and right CAAs, the latter originating from the left sinus of Valsalva.[7]

The coronary vessel originating from the contralateral sinus can trace different courses to reach the territory it irrigates.[3] Most patients with right CAA present with an inter-arterial course (between the aorta and the pulmonary trunk).[3,5,8] These anomalies may involve a changed ostium and coronary artery

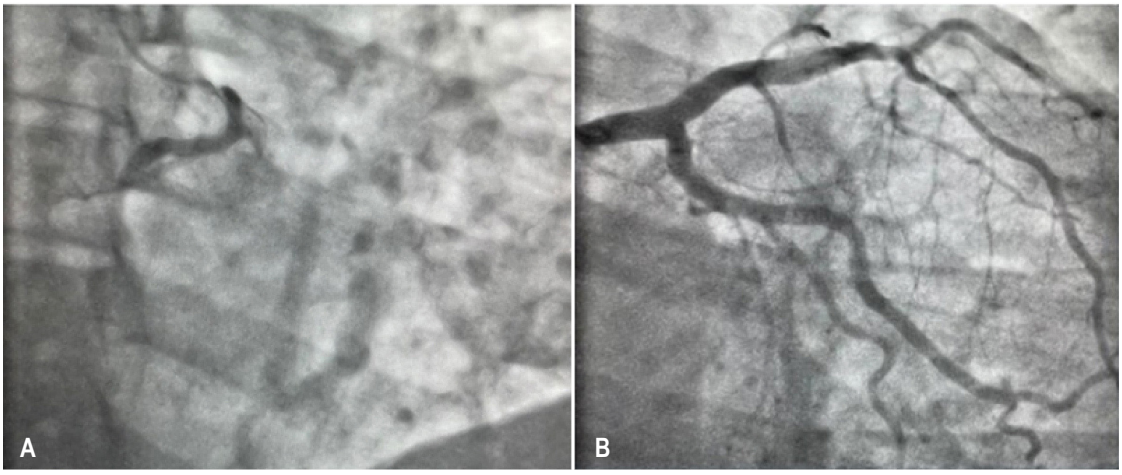

Figure 1 – *Cine coronary angiograpgy. A: Right, anomalus RCA and ostial/proximal stenosis (55%) B: Left coronary artery without stenosis.*

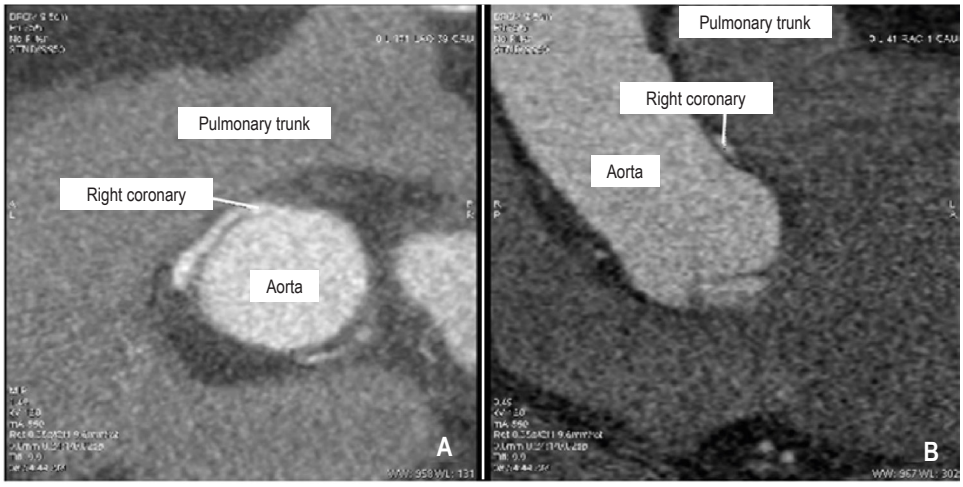

Figure 2 – *Computed tomograpgy angiography. A: Inter-arterial course between the aorta and pulmonar arteries. A e B: Improper right coronary artery origin.*

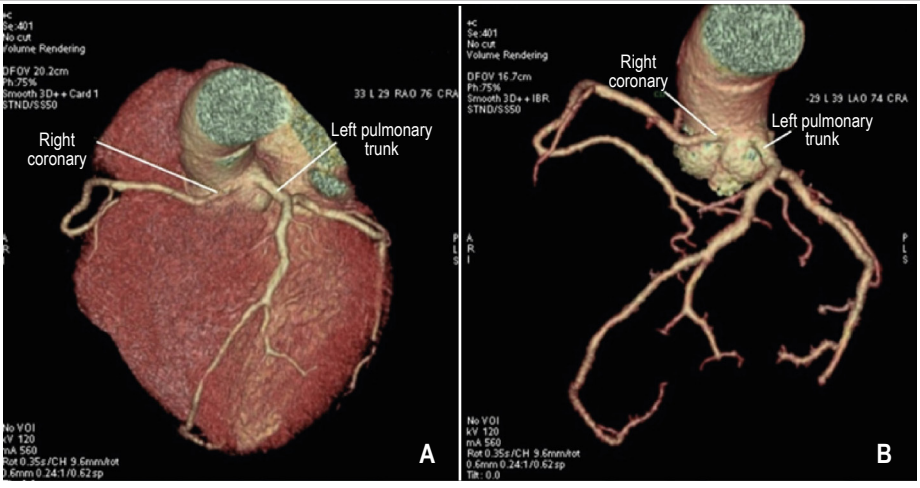

Figure 3 – *Computed tomograpgy angiography. A e B: Improper RCA origin in the left Valsalva sinus, adjacente to the origin of the left main coronary artery.*

angulation.[4,5] They result from an abnormal involution of the endothelial button-like junctions at the base of the common arterial trunk that join the coronary artery network to form the definitive arterial system or of the septation of the common arterial trunk.[5]

The inter-arterial course increases the degree of malignancy as well as the propensity for arrhythmias, myocardial infarction, and syncope.[3] The clinical presentation includes chest pain and dyspnea related to physical effort. However, there are asymptomatic cases in which sudden death is the first sign of the anomaly.[4]

The most accepted pathophysiological mechanism is that the proximal oblique course in the anomalous coronary artery shapes its ostium as a slit-like orifice instead of a circle, which can collapse with aortic expansion during systole, especially during exercise, decreasing blood flow supply to the myocardium.[7] During exercise, the systemic blood pressure and pulmonary territory increase, which can compress the inter-arterial coronary segment, causing dynamic obstructions and leading to ischemia and arrhythmias (including ventricular fibrillation). This can happen especially during more intense physical effort.[7] However, this explanation remains controversial.[3]

Abnormal changes can be classified into four classes by their functional repercussion and associated structural changes: I (benign and usually asymptomatic), II (relevant, associated with myocardial ischemia), III (severe ischemia, with potential risk of sudden death), and IV (severe, related to coronary atherosclerotic disease).[4]

There are no changes on physical examination, except in cases of structural cardiac injury. The diagnosis is usually made during an incidental finding during coronary angiography.[5]

Electrocardiography or stress test results may suggest ischemia, arrhythmia, or no evidence. Imaging tests allow better vessel visualization, anomaly classification, and risk stratification.[8]

Transthoracic echocardiography often identifies the origin and proximal segments of the coronary arteries. However, its sensitivity varies depending on the operator, patient age, echocardiographic window, and the analyzed anomaly.[9] Thus, coronary angiography is the best diagnostic method for identifying the anomaly. However, the definition of the coronary inter-arterial course is not accurate since it is a two-dimensional analysis method. Thus, the anatomical evaluation using three-dimensional methods has been increasingly used.[7] Computed tomography angiography, for example, more accurately defines the coronary location, shape, and angle of origin, as well as its course and relationship with the pulmonary artery and aorta.[5]

Surgical treatment is the standard of care in patients aged under 30 years with evidence of ischemia or ventricular arrhythmias and severe symptoms (such as previous infarction or sudden reversed death). However, the treatment of asymptomatic patients aged under 30 years and symptomatic patients without documented ischemia remains controversial, especially in older patients.[5]

The surgical risk outweighs the advantages of conservative treatment in many asymptomatic older patients.[5] Thus, the initial drug therapy (especially aiming at an effective beta-adrenergic block) can be implemented and periodically tested for its ability to lead to negative provocative ischemia tests.[3,5]

Percutaneous coronary intervention can be indicated, especially in patients at higher surgical risk, with good results in the short-term follow-up as previously described. In this case, a careful choice of therapeutic catheter and coronary guidewire is essential for greater support and an increased success rate. The use of drug-eluting stents should also be prioritized since the difficult selective catheterization of the anomalous coronary artery can hinder eventual reinterventions.[3]

To summarize, CAAs are rare and usually asymptomatic. However, the possibility of ischemia, myocardial arrhythmia, or even sudden death makes it mandatory for professionals to carefully screen patients for CAAs using more adequate imaging methods. An individual therapeutic plan should be implemented from the diagnosis depending on the patient's age and comorbidities, degree of habitual daily physical effort, and functional repercussion to reduce the morbidity and mortality related to CAAs, which can be treacherous, especially in young athletes.

In the case described here, the treatment of choice was the maintenance of clinical drug treatment since the patient showed normal myocardial perfusion without signs of ischemia. Although the inter-arterial course of this anomaly is associated with a higher occurrence of sudden death, especially after vigorous exercise, physical inactivity and, consequently, low daily physical effort made by the patient, associated with the absence of induced ischemia, reduce the risk of a fatal event and, thus, corroborate the therapeutic choice in this clinical case. Furthermore, the non-recurrence of anginal symptoms after drug treatment reinforces the initial noninterventional choice. The patient had a good clinical progression with the established therapy, remaining asymptomatic and without activity limitations in the subsequent evaluations.

Authors' contributions

Research concept and design: Almeida DC, Carrijo AMM, Souza MGS, O'Connell JL; data collection: Almeida DC, Carrijo AMM, Souza MG; data analysis and interpretation: Almeida DC, Carrijo MM, Souza MG, Martinelli FM, Fazzio FR, O'Connell JL; manuscript writing: Almeida DC, Carrijo AMM, Souza MG, Martinelli FM, Fazzio FR, O'Connell JL; critical review of the manuscript for important intellectual content: Martinelli FM, Fazzio FR, O'Connell JL.

Conflict of interest

The authors have declared that they have no conflict of interest.

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