

Diagnosis of Coronary Anomaly of the Circumflex Artery With Retroaortic Course by Transthoracic Echocardiography (Rac Sign) Confirmed by Computed Tomography Coronary Angiography: A Case Series

Diagnóstico de Anomalia Coronariana da Artéria Circunflexa com Trajeto Retroaórtico por meio do Ecocardiograma Transtorácico (RAC Sign) e Confirmação pela Angiotomografia de Artérias Coronárias: Série de Casos

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Introduction

Congenital coronary artery anomaly (CCAA) is a relatively rare condition with an estimated population prevalence of 0.21–5.79%.¹ CCAA presents a wide spectrum of variations and can be classified into origin and course, intrinsic, and termination anomalies.^{1,2} The CCAA with the highest prevalence is the anomalous origin of the circumflex artery (CxA) from the right coronary sinus (RCS) with an estimated prevalence of 0.3–0.8%.

CCAA usually travels along a retroaortic course and may present different originations, such as from a common ostium with the right coronary artery (RCA), from separate ostia in the RCS, or from the proximal segment of the RCA.^{2–4}

Although this anomaly is usually benign, i.e., with the characteristic of not causing symptoms, ischemia, or cardiovascular outcomes, numerous cases in the specialized literature associate this anomaly with episodes of angina, ischemia, and acute myocardial infarction. This infrequent progression is mainly associated with factors related to the coronary artery origin angulation to extrinsic compression caused by aortic root dilation and the presence of an intramural course in the aortic wall.^{5–10}

Transthoracic echocardiography (TTE) is a noninvasive and easily accessible tool that allows a limited assessment of some findings suggestive of CCAA.¹⁰ One of these findings is the retroaortic anomalous coronary sign (RAC sign), a signal proposed by Witt et al. in 2017.^{10–12}

The RAC sign is characterized by a hyperechoic tubular image located on the atrial face of the atrioventricular groove perpendicular to the aorta. This finding showed high sensitivity for the diagnosis; therefore, it has a high relationship with the presence of anomalous origin of the CxA from the RCS with a retroaortic course.^{10–12}

Keywords

Cardiovascular Abnormalities; Echocardiography; Computed Tomography Angiography.

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On the other hand, coronary computed tomography angiography (CCTA) is a robust diagnostic tool in the anatomical assessment of the coronary tree and, therefore, the diagnosis of CCAA. As it is a noninvasive method with high temporal and spatial resolution, CCTA is currently considered the first-choice method for suspected CCAA as described in the 2010 appropriate use criteria for CCTA of the American Heart Association (AHA).^{4,13}

Compared to invasive coronary angiography (ICA), CCTA is superior for CCAA assessments, especially in cases of course anomalies involving the ostium and proximal third of the coronary arteries, intrinsic anomalies (myocardial bridging), and termination anomalies (fistulas). CCTA also stands out compared to ICA due to the possibility of a three-dimensional evaluation that allows the analysis of the close relationship between the coronary and other cardiac and non-cardiac structures, mainly the aortic root and pulmonary artery, extremely important data in the prognostic assessment of CCAA.

This case series aimed to demonstrate the agreement between the echocardiographic finding (RAC sign) and the noninvasive anatomical assessment by CCTA, its high sensitivity, and the assessment of the presence or absence of myocardial ischemia in the absence of obstructive atherosclerotic lesions.

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Case 1

A 52-year-old man had a body mass index (BMI) of 27.3 kg/m², had dyslipidemia, was using a high-potency statin, and had a history of stable atypical angina and intermediate cardiovascular risk by the Diamond-Forrester classification. A stress test (ST) and three-dimensional transthoracic echocardiography (3D-TTE) were requested to complement the diagnosis.

The 3D-TTE showed preserved global and segmental systolic function (end-diastolic volume [EDV], 122 mL; end-systolic volume [ESV], 44 mL; left ventricular ejection fraction [LVEF], 64%) and global longitudinal strain (GLS; 18.6%) were within the normal range (Figure 1). The diastolic function parameters were preserved and the heart valves showed no significant anatomical or functional changes.



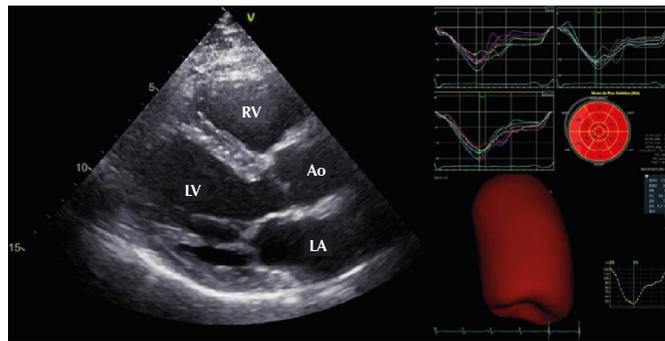
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However, a hyperechoic tubular image characteristic of the RAC sign was observed in the apical windows suggestive of a CxA anomalous retroaortic coronary course (Figure 2, Video 1).

The Bruce protocol was included in the ST and was effective and reached 99% of the age-predicted maximal heart rate. The patient presented no clinical symptoms on physical exertion and demonstrated excellent functional

capacity, adequate blood pressure behavior, and the absence of arrhythmias or electrocardiographic criteria for myocardial ischemia (Figure 3).

The CCTA findings complemented the diagnosis (Figures 4 and 5), corroborating the echocardiographic findings and showing an anomalous origin of the CxA from the RCS in a single ostium and at an acute angle (30°) traveling the



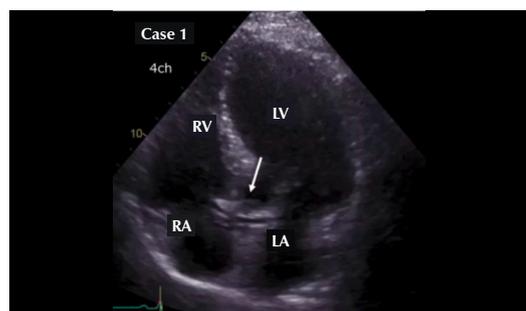
Ao, aorta; LA, left atrium; LV, left ventricle; RV, right ventricle. Source: Images obtained in the cardiovascular imaging sector of DASA S/A.

Figure 1 – Echocardiographic window in parasternal long-axis view showing the cardiac chambers with preserved dimensions (left). Graphs and bullseyes of preserved left ventricular global longitudinal strain (top right) and left ventricular systolic function by three-dimensional echocardiography showing a preserved LV ejection fraction (bottom right).



Ao, aorta; LA, left atrium; LV, left ventricle; RA, right atrium; RAC, retroaortic anomalous coronary; RV, right ventricle. Source: Images obtained in the cardiovascular imaging sector of DASA S/A.

Figure 2 – Echocardiographic four-, three-, and five-chamber sections showing the RAC sign (white arrow), which suggests an anomalous origin of the CxA with a retroaortic course.



LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle. Source: Images obtained in the cardiovascular imaging sector of DASA S/A.

Video 1 – Four-chamber apical echocardiogram showing the retroaortic anomalous coronary sign (white arrow).

retroaortic course without signs of intramural course or extrinsic compression by the aortic root (aortic root with a maximum diameter of 32 mm). As for atherosclerosis, CCTA showed a 29 Agatston calcium score with atherosclerotic plaques in two segments of the coronary tree and no significant luminal reduction.

Case 2

A 53-year-old man who was asymptomatic had a BMI 28 kg/m², had dyslipidemia, and was using a high-potency statin came to our service for routine tests.

He underwent successful myocardial perfusion scintigraphy (MPS) with ST on a treadmill (Ellestad protocol), reaching the age-predicted maximal heart rate (Figure 6). The

patient presented no clinical symptoms, arrhythmias, or electrocardiographic changes suggestive of ischemia during the ST. MPS showed a preserved LVEF at rest (58%) and under stress (65%) and no perfusion defects.

The 3D-TTE presented cardiac chambers with preserved dimensions and normal global and segmental ventricular systolic function (EDV, 123 mL; ESV, 46 mL; and LVEF, 63%). GLS demonstrated preserved values (20.9%), while the diastolic function analysis findings were normal. No anatomical or functional valve changes were identified (Figure 7). The apical section showed a hyperechoic tubular image compatible with the RAC sign (Figure 8).

CCTA performed as a diagnostic complement identified an anomalous CxA origin from the RCS with a common ostium

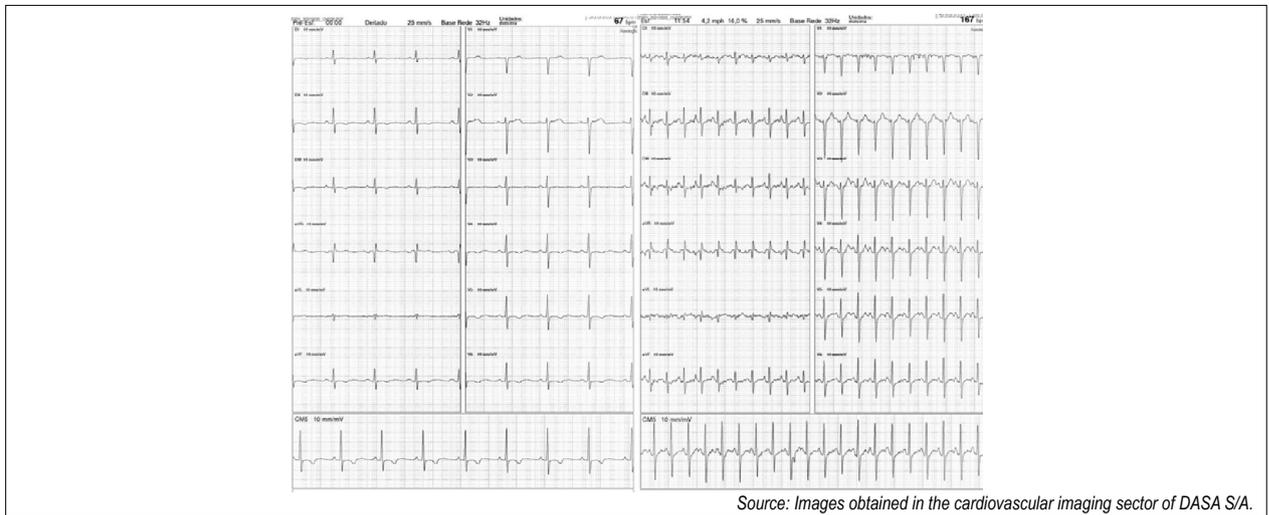


Figure 3 – Electrocardiographic tracing of the stress test (at rest on the left and peak exertion on the right) without electrocardiographic signs of myocardial ischemia on exertion.

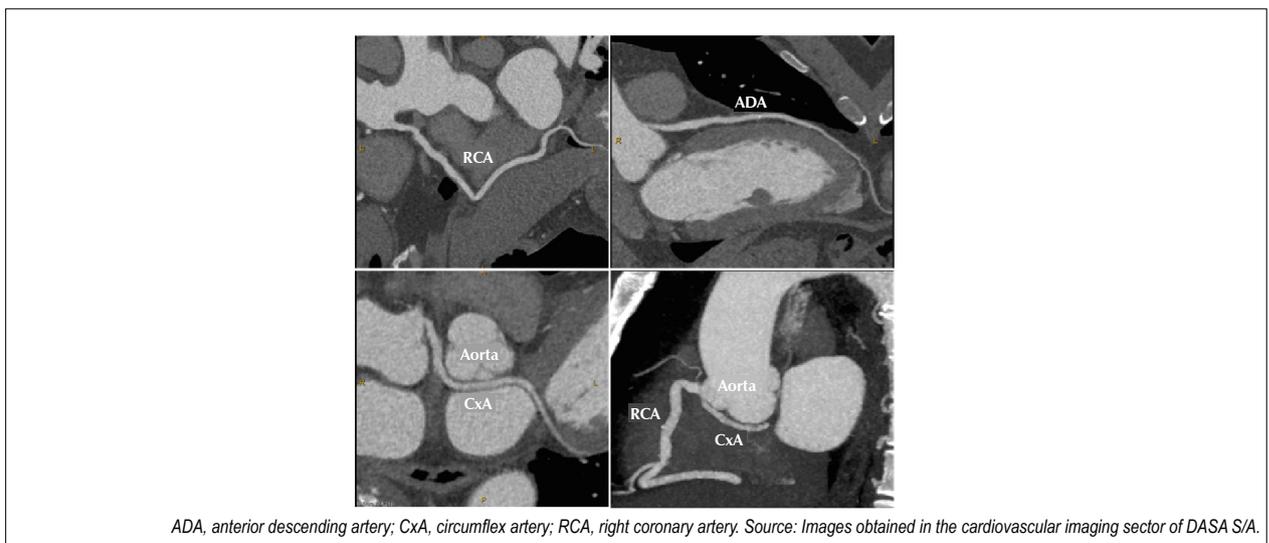
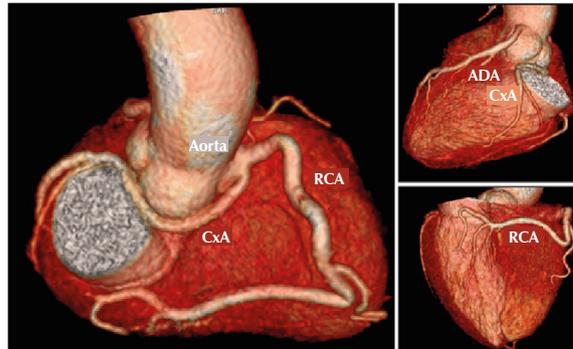


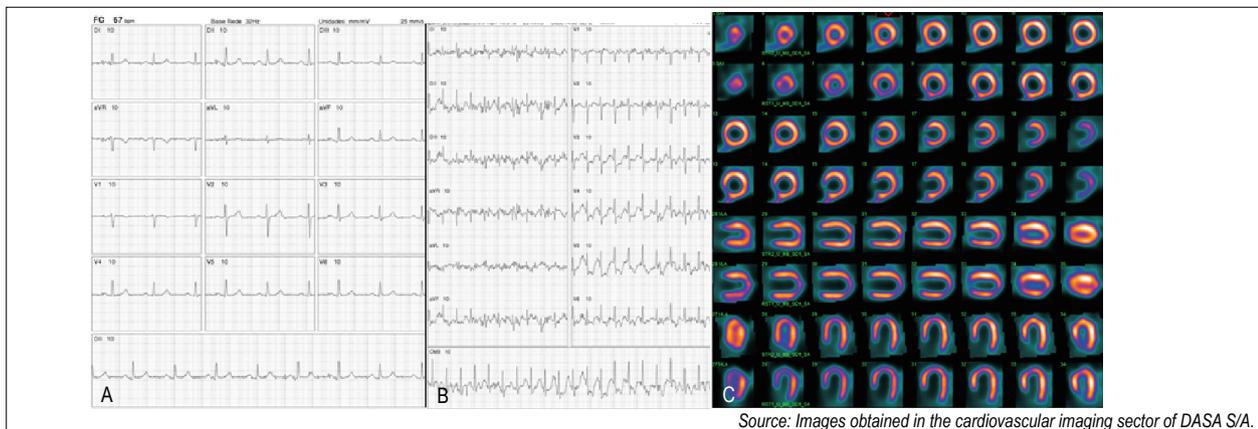
Figure 4 – Computed tomography coronary angiography showing the absence of significant luminal reduction and the presence of anomalous origin of the CxA from a single ostium and its retroaortic course.

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ADA, anterior descending artery; CxA, circumflex artery; RCA, right coronary artery. Source: Images obtained in the cardiovascular imaging sector of DASA S/A.

Figure 5 – 3D reconstruction of CT coronary angiography showing the coronary anatomy with an anomalous course and absence of atherosclerotic plaques.



Source: Images obtained in the cardiovascular imaging sector of DASA S/A.

Figure 6 – Electrocardiographic tracing (A, at rest; B, at peak exertion) without criteria for ischemia and myocardial perfusion scintigraphy with no perfusion changes (C).

with the RCA at an acute angle (35°) and with a retroaortic course. The course showed no luminal reduction and no signs of intramural course or extrinsic compression by the aortic root (maximum aortic root diameter, 33 mm) (Figures 9 and 10).

Case 3

A 31-year-old woman who had a BMI 20 kg/m², was asymptomatic, and had no known comorbidities was referred for routine TTE. The TTE showed preserved global and segmental biventricular systolic function. Her GLS (22.8%) and diastolic function were within the normal range. No significant anatomical and functional valve changes were identified. However, a hyperechoic tubular image was also visualized in apical sections perpendicular to the aorta, consistent with the RAC sign and suggestive of coronary anomaly (Figure 11, Video 2).

As a diagnostic complement, CCTA showed an anomalous origin of the CxA from the RCS in a separate ostium in an acute angle (26°), at a retroaortic course, and with no intramural course (Figures 12 and 13). No extrinsic compression by the aortic root was observed (largest aortic root diameter, 26 mm).

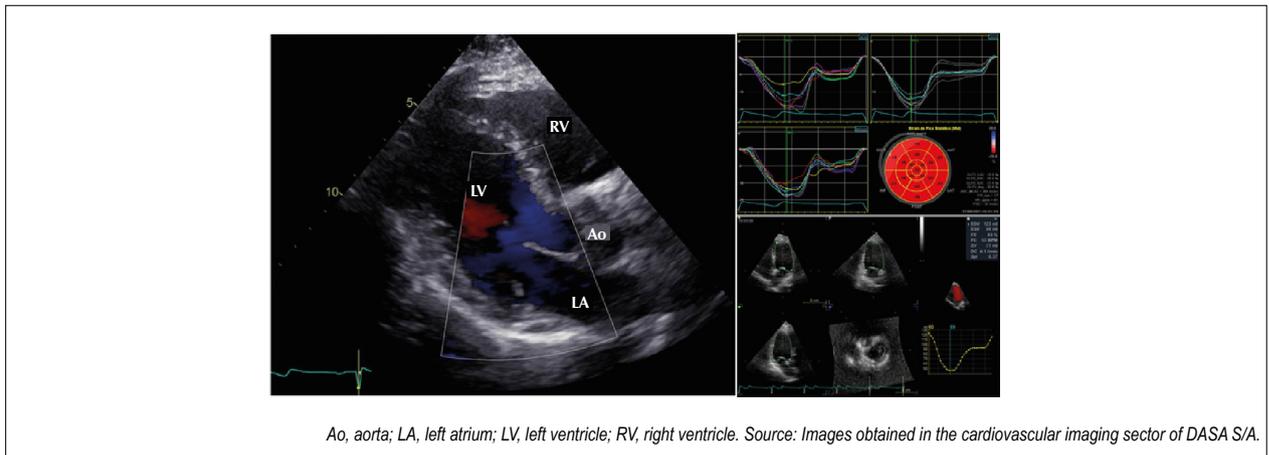
An atherosclerosis assessment presented a coronary calcium score of zero and absence of atherosclerotic plaques in the coronary segments.

In this case, no myocardial ischemia investigation was performed.

Discussion

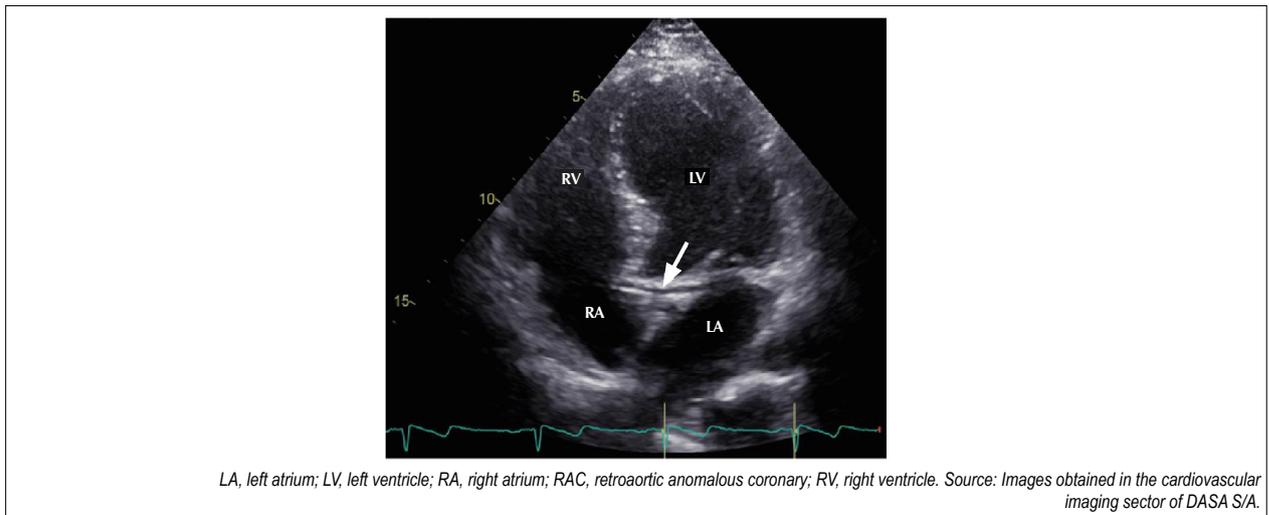
Here we reported a series of three cases of a suspected CxA anomalous retroaortic course due to a TTE characteristic finding known as the RAC sign. Corroborating the data in the literature, this finding presented high diagnostic sensitivity since the coronary anomaly was confirmed in all cases on a noninvasive anatomical CCTA evaluation.

We also highlighted that CCTA is the gold standard method for CCAA evaluations since it is an easy and noninvasive way to analyze the characteristics of the coronary anomaly and provides important complementary information that can suggest a worse prognosis, such as the coronary ostium outflow angulation, the presence of an intramural course, and the relationship between the coronary artery and adjacent cardiac or non-cardiac structures.



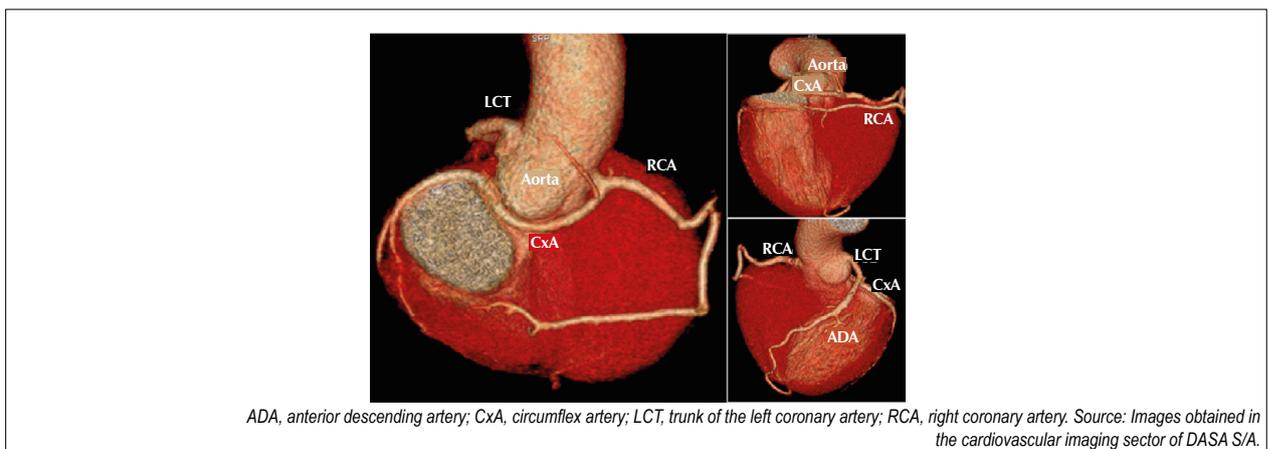
Ao, aorta; LA, left atrium; LV, left ventricle; RV, right ventricle. Source: Images obtained in the cardiovascular imaging sector of DASA S/A.

Figure 7 – Parasternal long-axis echocardiographic window (left), global longitudinal strain (top right), and preserved global systolic function by three-dimensional echocardiography (bottom right).



LA, left atrium; LV, left ventricle; RA, right atrium; RAC, retroaortic anomalous coronary; RV, right ventricle. Source: Images obtained in the cardiovascular imaging sector of DASA S/A.

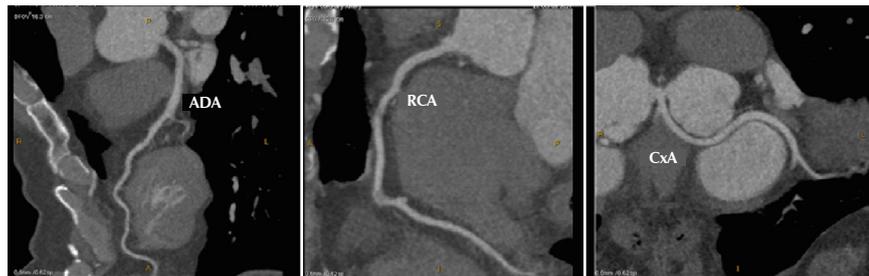
Figure 8 – Echocardiographic five-chamber section demonstrating the RAC sign (white arrow), characteristic of anomalous CxA with a retroaortic course.



ADA, anterior descending artery; CxA, circumflex artery; LCT, trunk of the left coronary artery; RCA, right coronary artery. Source: Images obtained in the cardiovascular imaging sector of DASA S/A.

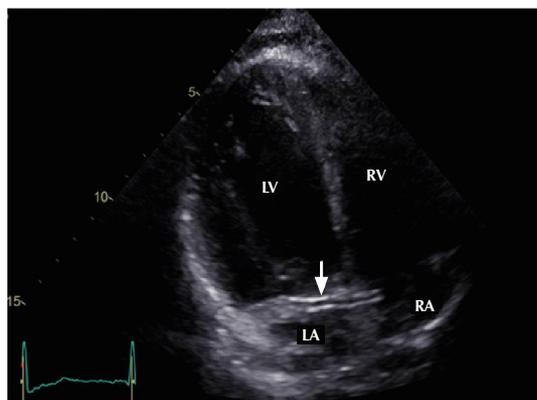
Figure 9 – Three-dimensional reconstruction of computed tomography coronary angiography showing the coronary anatomy with an anomalous course and absence of atherosclerotic plaques.

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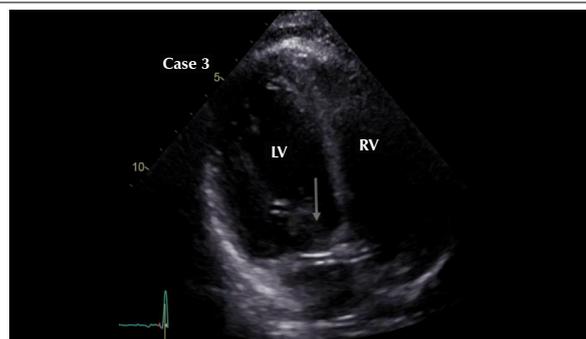
ADA, anterior descending artery; CxA, circumflex artery; RCA, right coronary artery. Source: Images obtained in the cardiovascular imaging sector of DASA S/A.

Figure 10 – Computed tomography coronary angiography showing the absence of significant luminal reduction and the presence of an anomalous CxA origin from a single ostium and retroaortic course.



VE: ventrículo esquerdo; VD: ventrículo direito; AD: átrio direito; AE: átrio esquerdo. Source: Images obtained in the cardiovascular imaging sector of DASA S/A.

Figure 11 – Apical four-chamber echocardiographic view showing tubular hyperechoic image – RAC sign (white arrow), suggestive of anomalous CxA origin with retroaortic course.



LV, left ventricle; RV, right ventricle. Source: Images obtained in the cardiovascular imaging sector of DASA S/A.

Video 2 – Four- and three-chamber apical view echocardiogram showing the retroaortic anomalous coronary sign (white arrow).

It is important to emphasize that an ST was performed in two of the cases described here to investigate myocardial ischemia. The results of both were negative, once again corroborating the findings in the literature, which mostly state that this anomalous course has a benign progression. However, as already mentioned, there are several reports of major and minor cardiovascular events related to this anomaly, a fact that supports the indication of ischemia research in cases of this diagnosed anomaly.

Conclusion

This case series highlights the fundamental role of TTE in the diagnostic confirmation of CCAA by the presence of the RAC sign, especially in cases with a retroaortic origin of the CxA. Despite the limitations inherent to the method, TTE has robust ability to identify this anomaly; thus, its use should always be considered so that a diagnostic opportunity is not missed.

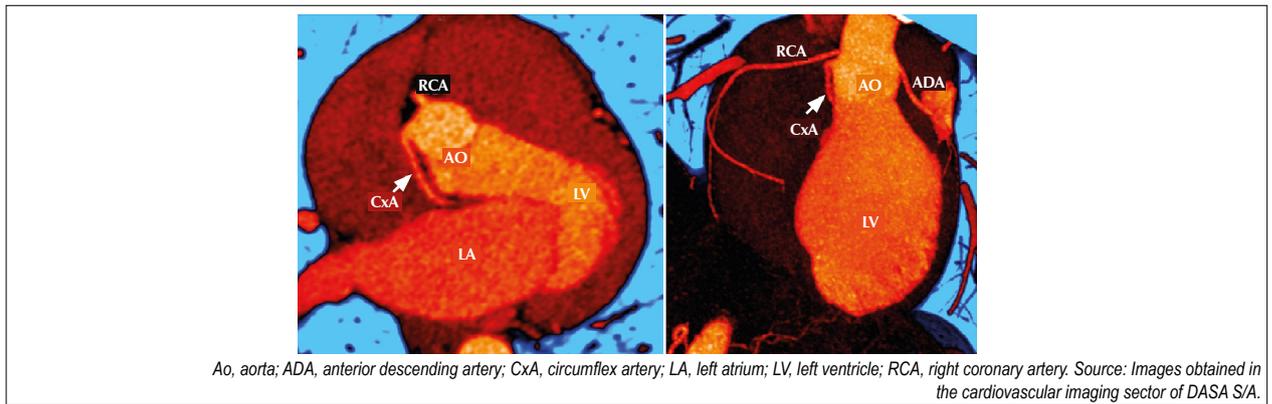


Figure 12 – Tomographic section reconstruction showing the anomalous origin of the CxA from the RCS in a separate ostium and with a retroaortic course (white arrow).

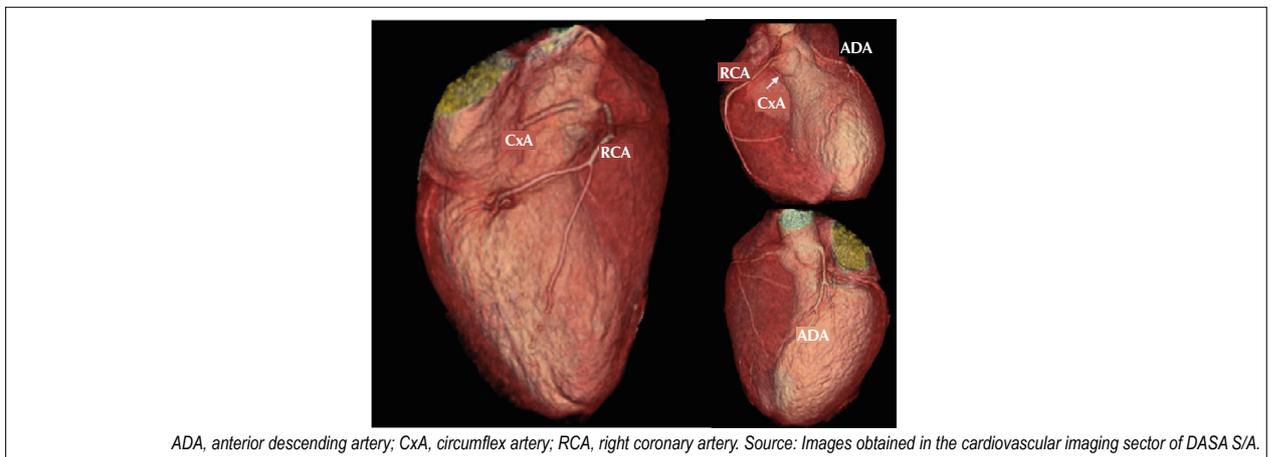


Figure 13 – Three-dimensional reconstruction of computed tomography angiography of the coronary arteries showing the coronary anatomy with an anomalous course and absence of atherosclerotic plaques.

Authors' contributions

Valério RS, Aguiar Filho LF: research conception and design; Silva CES, Aguiar Filho LF: data collection; Valério RS: manuscript writing; Uellendahl M, Rodrigues AAE, Silva CES, Aguiar Filho LF: critical review of the manuscript for important intellectual content.

Conflict of interest

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

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