

CASE REPORT

Right Coronary Artery Agnesia Diagnosed by Coronary CT Angiography: A Case Report

Agnesia De Arteria Coronaria Derecha Diagnosticada Por Angiotomografía Coronaria: Reporte De Caso

Agnesia da artéria coronária direita diagnosticada por angiotomografia coronária: relato de caso

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ABSTRACT

Agnesia of the coronary arteries is a rare anomaly, with an incidence ranging from 0.024% to 0.066% in the population. We present the case of a 49-year-old female patient with typical chest pain on moderate exertion for one year and moderate functional limitation (NYHA III), with an abnormal exercise stress test and subsequent referral for coronary computed tomography angiography. This revealed absence of the right coronary artery. The left coronary artery and its two branches showed a normal course and left dominance, predominantly through the circumflex artery, with increased collateral circulation to the right ventricle. The left coronary artery showed no significant lesions; therefore, symptomatic medical treatment was initiated. The patient showed clinical improvement after medical treatment and cardiovascular rehabilitation

Keywords: Right coronary artery agnesia, Collateral circulation, Computed tomography angiography.

RESUMEN

La agnesia de arterias coronarias es una anomalía poco frecuente con una incidencia entre el 0,024% al 0,066% de la población. Se presenta el caso de una paciente femenina de 49 años, con dolor torácico típico a los medianos esfuerzos de un año de evolución y limitación funcional moderada (NYHA III), con prueba de esfuerzo alterada, y posterior derivación a una angiotomografía coronaria, la misma que mostró ausencia de arteria coronaria derecha, la arteria coronaria izquierda con sus dos ramas presentó un trayecto normal y dominancia izquierda a predominio de la arteria circunfleja, con aumento de la circulación colateral para ventrículo derecho. La arteria coronaria izquierda no presentó lesiones significativas por lo que se administró tratamiento médico sintomático. Paciente con mejoría clínica posterior a tratamiento médico y rehabilitación cardiovascular

Palabras Clave: Agnesia de la arteria coronaria derecha, Circulación colateral, Angiotomografía computarizada.

RESUMO

A agenesia das artérias coronárias é uma anomalia rara, com incidência entre 0,024% e 0,066% da população. Apresenta-se o caso de uma paciente do sexo feminino, de 49 anos, com dor torácica típica aos médios esforços, com um ano de evolução, e limitação funcional moderada (NYHA III), com teste ergométrico alterado e posterior encaminhamento para angiotomografia coronariana, a qual evidenciou ausência da artéria coronária direita. A artéria coronária esquerda, com seus dois ramos, apresentou trajeto normal e dominância esquerda com predomínio da artéria circunflexa, além de aumento da circulação colateral para o ventrículo direito. A artéria coronária esquerda não apresentou lesões significativas, motivo pelo qual foi instituído tratamento médico sintomático. A paciente apresentou melhora clínica após o tratamento médico e a reabilitação cardiovascular.

Palavras-Chave: Agenesia da artéria coronária direita, Circulação colateral, Angiotomografia computadorizada.

INTRODUCTION

During the sixth and seventh weeks of embryonic development, the coronary arteries are formed through an angioblastic bud that extends over the epicardial layer of the heart and originates from the sinus of Valsalva of the aorta.⁽¹⁾ The right coronary artery courses along the right atrioventricular groove, supplying the sinoatrial node in 55% to 65% of patients, giving rise to the marginal branch, and ending in the interventricular and posterolateral branches. In contrast, the left coronary artery divides into the left anterior descending artery, which gives off septal and diagonal branches, and the circumflex artery with its marginal branches, which supply the anterior and lateral portions of the left ventricle. In 70% to 89% of cases, coronary dominance is right-sided, whereas in 7% to 13% it is left-sided or codominant.^(2,3)

Coronary anomalies occur in 1.7% of the population and account for 33% of sudden deaths worldwide.⁽²⁾ Congenital absence of the right coronary artery has a very low incidence, ranging from 0.024% to 0.066%, and arises from abnormalities during the embryonic period.⁽⁴⁾ These anomalies may be classified from an anatomical perspective, considering the affected segment, its origin, and termination, and from a functional perspective, considering whether or not they are hemodynamically significant ⁽⁵⁾, such as in the presence of arrhythmias or sudden death.^(6,4,7) Therefore, these patients should be evaluated and treated in specialized centers in order to reduce cardiovascular risk.

CASE PRESENTATION

A 49-year-old female patient with a past medical history of gastritis and untreated arterial hypertension presented with moderate-intensity oppressive precordial pain, triggered by moderate exertion and predominantly occurring during nighttime hours, with radiation to the head and left upper limb. The symptoms had been present for one year and had worsened over the last six months, accompanied by constant holocranial headache.

On physical examination, her blood pressure was 160/95 mmHg, heart rate was 89 bpm, respiratory rate was 18 breaths/min, and temperature was 36.8°C.

Laboratory tests showed hematocrit (Hct) of 37.7%, hemoglobin (Hb) of 13.4 g/dL, glucose of 98 mg/dL, urea of 21.45 mg/dL, creatinine of 0.73 mg/dL, total cholesterol of 300 mg/dL, triglycerides of 350 mg/dL, HDL cholesterol of 20 mg/dL, and LDL cholesterol of 130 mg/dL. Troponin and CPK levels were normal, as were the remaining paraclinical tests.

A resting electrocardiogram (Figure 1), transthoracic echocardiogram (Table 1), exercise stress test (Table 2), post-exercise electrocardiogram (Figure 2), and coronary CT angiography (Figures 3, 4, and 5) were performed and are presented below.

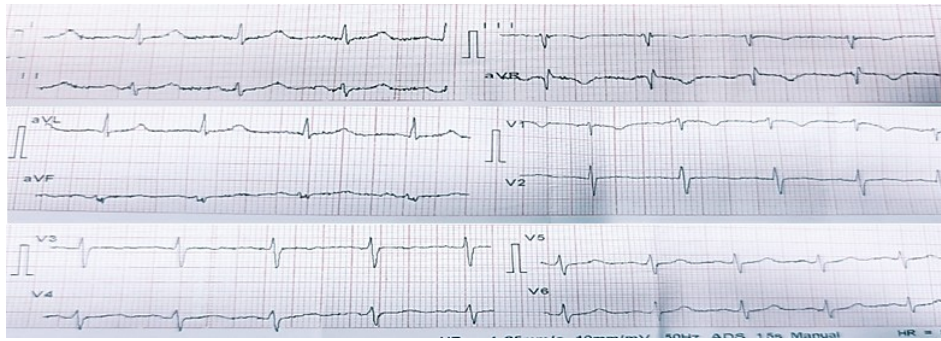


Figure 1. Electrocardiogram performed at the initial consultation, showing an incomplete right bundle branch block.

Table 1. Transthoracic Echocardiogram

Transthoracic Echocardiogram	Transthoracic Echocardiogram
LVEF	75%
LV diastolic dysfunction	Absent
Wall motion abnormality	Absent
Mass	Concentric LV hypertrophy, grade I hypertensive heart disease
Valves	Competent
RV dysfunction	Absent

Table 2. Exercise Stress Test

Blood pressure response	Mildly elevated systolic-diastolic response to exercise
Arrhythmias	Frequent PVCs during exercise and at rest
T waves	T-wave inversion from V1 to V6 during exercise, with slow recovery
Functional class	NYHA III

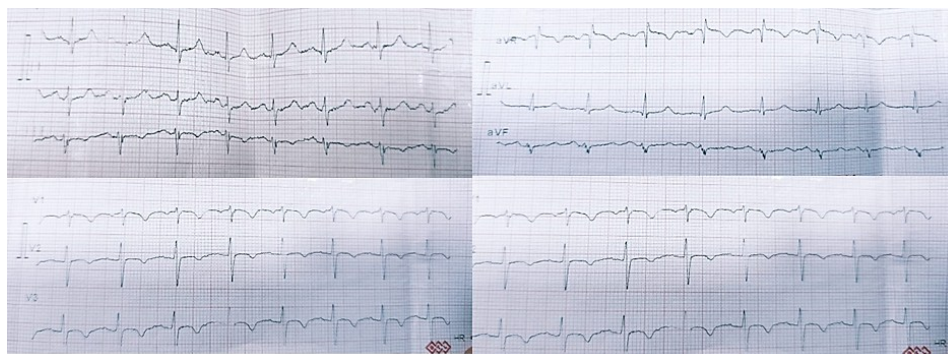


Figure 2. Electrocardiogram performed after exercise, showing an incomplete right bundle branch block and negative T waves from V1 to V6.

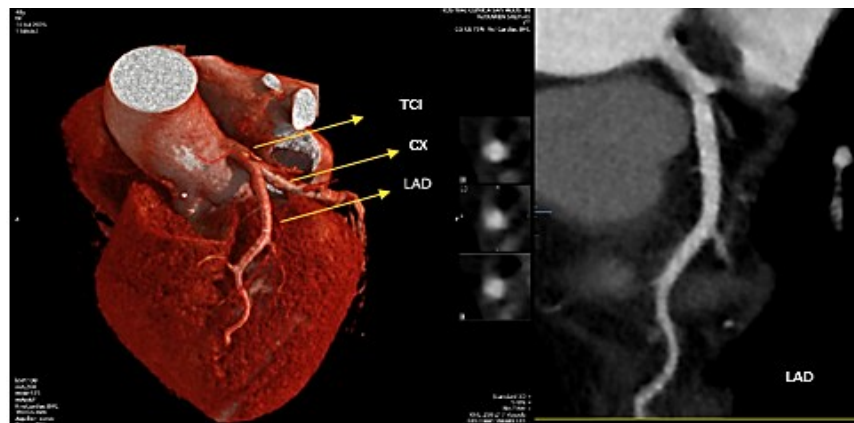


Figure 3. Coronary CT angiography: Left: Absence of the right coronary artery and origin of the left coronary artery are demonstrated. CX: Circumflex artery; LAD: Left anterior descending artery; LCA: Left coronary artery. Right: Absence of stenosis in the LAD.

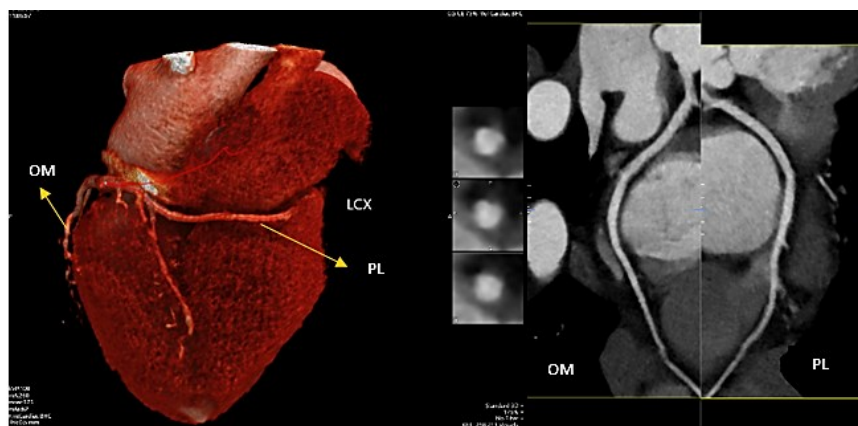


Figure 4. Coronary CT angiography: Left: The circumflex artery is visualized, with the presence of the obtuse marginal (OM) artery and the posterolateral (PL) artery. Right: Absence of stenosis in the OM and anterior PL branches. LCA: Left coronary artery. Right: Absence of stenosis in the LAD.

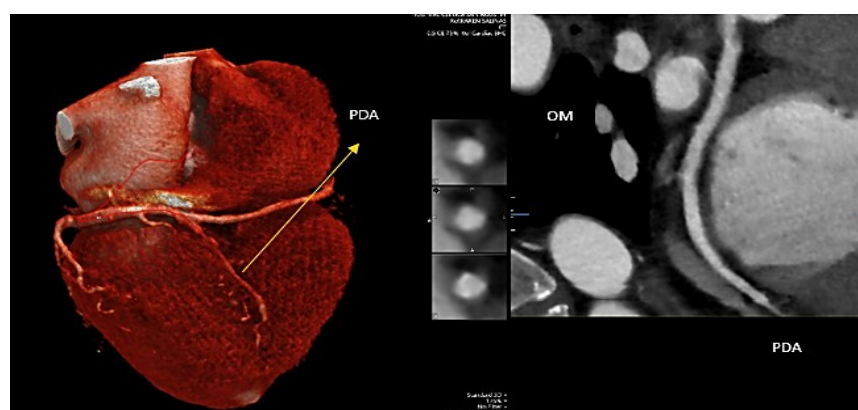


Figure 5. Coronary CT angiography: Left: The circumflex artery is visualized, with the presence of the posterior descending artery (PDA). Right: Absence of stenosis in the PDA.

The patient was treated with olmesartan 20 mg once daily. Due to intolerance to verapamil, bisoprolol 2.5 mg once daily, rosuvastatin 20 mg daily, and cardiac rehabilitation were initiated, resulting in clinical improvement, with a reduction in precordial pain and extrasystoles, improvement to NYHA functional class II, and adequate blood pressure control according to 24-hour ambulatory blood pressure monitoring (ABPM): 128/78 mmHg (reference value: 130/80 mmHg).

DISCUSSION

The absence of the right coronary artery is a condition with low incidence and prevalence, mainly caused by alterations in the rotation and migration of the sixth aortic arch during embryogenesis.⁽⁸⁾ Inactivation of the signaling pathways involved in normal coronary development, such as Tbx5, Tbx18, and Vegf-A/Vegfr2, may lead to relevant coronary artery anomalies, including hypoplasia or congenital absence.⁽⁹⁾

There are several classifications for coronary artery anomalies, among them the Greenberg and Angelini classification, which categorizes them according to their origin and course, intrinsic anomalies, and termination anomalies. A single coronary artery is characterized by a single ostium and may follow the course of the left anterior descending artery or the right coronary artery.⁽²⁾

In general, arterial agenesis is asymptomatic; however, when it coexists with cardiovascular risk factors such as hypertension, diabetes, dyslipidemia, or ischemic heart disease, the single coronary artery may be unable to supply blood both to its own territory and to that of the missing artery. This, together with abnormal vascular or microvascular damage and the long course traveled by the anomalous coronary artery, results in hypoxemia.^(9,10) This may lead to acute coronary syndrome, syncope, ventricular fibrillation, bundle branch blocks, bradycardia, or sudden death.⁽¹¹⁾ This may or may not produce electrocardiographic abnormalities. In the case of our patient, risk factors such as hypertension and dyslipidemia were present, which led to microvascular damage; when myocardial demand increased during physical activity, she developed anginal symptoms. The electrocardiogram showed T-wave inversion in the anterolateral leads after exercise and the presence of frequent extrasystoles.

Coronary CT angiography is the gold standard for patients with typical and atypical anginal symptoms, as it provides excellent visualization of anomalies, extracardiac blood vessels, and coronary arteries.^(4,10) This noninvasive study has a high negative predictive value of 90%–97% and also allows evaluation of other cardiac structures such as the left ventricle, right ventricle, and atria.^(12,4) In the present case, right coronary agenesis was identified through this method, with increased collateral circulation; since there was no significant stenosis in the existing coronary artery, no invasive treatment was performed.

At present, there is no specific treatment for coronary agenesis; however, symptomatic treatment with antiplatelet agents, lipid-lowering drugs, antihypertensive agents, and control of cardiovascular risk factors has been shown to be beneficial.^(9,11,12) In this case, adequate blood pressure control was achieved; the beta-blocker reduced both the arrhythmic and anginal burden, while the lipid-lowering therapy and cardiac rehabilitation improved the patient's functional class and quality of life.

CONCLUSION

Right coronary artery agenesis has a low incidence and prevalence; therefore, coronary CT angiography is the diagnostic technique of choice.

This condition requires multidisciplinary symptomatic management with an individualized follow-up plan, based on each patient and their comorbidities.

The cornerstone of treatment is the preservation of the remaining coronary arteries, cardiac rehabilitation, specialist follow-up, and adequate adherence to treatment.

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CONSENT

Informed consent was obtained from the patient for the preparation of this study.

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CONFLICT OF INTEREST

The author declares that there is no conflict of interest.

AUTHOR CONTRIBUTIONS

Conceptualization: Karen Salinas

Investigation: Karen Salinas

Methodology: Karen Salinas

Project administration: Karen Salinas

Writing – original draft: Karen Salinas

Writing – review & editing: Karen Salinas