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Case Report

Rare Congenital Anomalous Origin of the Coronary Arteries: A Series of Three Cases

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Abstract

Introduction: Congenital coronary anomalies are found in 0.3% to 5.6% of the population and can be classified according to alterations in origin, distribution or course. Although benign in most cases, they can be associated with sudden death in young adults, especially after strenuous physical exercise. We report three rare cases of anomalous coronary arteries found incidentally by coronary angiography.

Case Report: The first case refers to a 79-year-old man who underwent cardiac catheterization for etiological investigation of heart failure, showing a non-obstructive interruption of the left anterior descending artery (LAD) after the origin of the first diagonal branch, with the continuation of its middle and distal segments from the right coronary artery (RCA), which crosses to the left side at the atrial level and reaches the apex cordis. The second case is a 56-year-old woman with stable angina and a treadmill exercise test suggestive of myocardial ischemia. Coronary angiography revealed an anomalous origin of the RCA from the distal segment of the left circumflex artery (LCxA). The third and last case is a 64-year-old woman in whom coronary angiography revealed an anomalous origin of the LCxA from the proximal third of the RCA.

Discussion: Coronary artery anomalies are a heterogeneous group whose diagnosis mostly occurs incidentally. They differ from anatomical variations in their rarity and greater association with adverse events, such as myocardial ischemia and sudden death. The cases presented address different anomalous origins: a LAD with duplicated origin, a single coronary artery with the emergence of the RCA in the distal third of the LCxA, and the anomalous origin of the LCxA from the RCA, one of the most common coronary anomalies. The three cases reported were patients over 50 years old, which shows the need to individualize cases and risks as regarding to anomalous origins of the coronary arteries.

Keywords

Coronary Artery, Coronary Artery Disease, Sudden Cardiac Death, Vascular Anomaly.

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Introduction

Coronary anomalies are congenital malformations of the origin, course or termination of the coronary arteries, found in 0.2 to 5.6% of the population [1,2]. The study of these anomalies is especially important when observed in the context of sports and athletics, since they represent the second leading cause of sudden death among athletes, accounting for 12.2% to 17.7% of deaths [3]. However, there is a lack of multicenter studies, in addition to great difficulty in estimating the real prevalence of the anomalies, which are overestimated when investigated in the occurrence of cardiac events [1,2]. This poses a major obstacle to their functional and prognostic definition. The most documented prevalences, however, are the left circumflex coronary artery (LCxA) originating from the right coronary sinus, the right coronary artery (RCA) originating from the left coronary sinus, and the left main coronary artery (LMCA) emerging from the right coronary sinus [2-4]. This study reports three rare cases of congenital coronary anomalies as an incidental finding seen on coronary angiography.

Case Report

Case 1

Fifty-year-old female outpatient with no previous comorbidities, presenting symptomatic PVCs. The resting electrocardiogram (ECG) showed frequent PVCs probably originated in the right ventricle outflow tract (Figure 1). She reported frequent palpitations compromising quality of life. The 24-hour Holter monitorization showed 13,008 PVCs in 24 hours (14.1% of all heartbeats). During the investigation, a coronary angiography was performed, which did not show obstructive lesions. Magnetic resonance imaging of the heart showed late epicardial enhancement of non-ischemic etiology in the anterolateral (basal) and inferolateral (basal and middle) segments of the left ventricle, with normal cavity diameters and without ventricular dysfunction.

Case 2

A 56-year-old woman presented with stable angina and ischemic findings on electrocardiogram during a treadmill exercise test. Coronary angiography via the right radial artery revealed mild stenosis in the LAD and anomalous origin of the RCA from the distal segment of the LCxA, making it a single coronary artery (Figure 1C).

Case 3

A 64-year-old woman with type 2 diabetes, hypertension and active smoking, presented an inversion of the T wave in the entire anterior wall on the electrocardiogram during the preoperative period of orthopedic surgery. She was referred for coronary angiography, which revealed coronary arteries without stenosis and anomalous

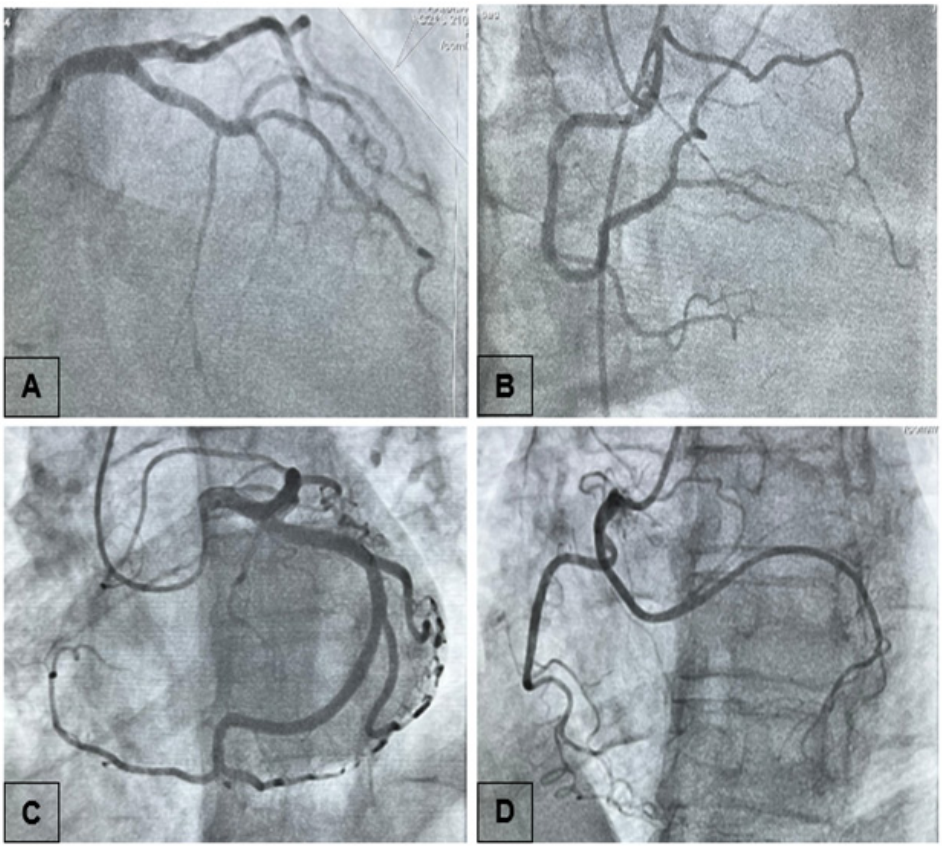

Figure 1: Coronary angiography images. **A.** Nonobstructive interruption of the left anterior descending (LAD) artery after originating the first diagonal branch. **B.** Right coronary artery (RCA) originating the middle and distal segments of the LAD, which crosses to the left side at the atrial level and reaches the apex cordis. **C.** Anomalous origin of the RCA from the distal segment of the left circumflex artery (LCxA). **D.** Anomalous origin of the LCxA from the proximal segment of the RCA.

origin of the LCxA from the proximal segment of the RCA (Figure 1D).

Discussion

Coronary artery anomalies are a group of congenital conditions characterized by the abnormal origin or course of any of the three coronary epicardial arteries: LAD, LCxA, and RCA [1]. This is a heterogeneous group, and its incidence varies greatly in literature, since most are diagnosed incidentally, through routine coronary angiography or autopsy. In addition, they are frequently confused with anatomical variabilities, being differentiated by their rarity (< 1% among unselected individuals) and greater association with adverse events and unfavorable outcomes [1,5]. Regarding clinical presentation, most cases are asymptomatic; however, they can be the cause of myocardial ischemia, manifesting as angina, arrhythmias, acute myocardial infarction and sudden death, the latter being related to vigorous physical exercise [6]. Despite their rarity in the population, Coronary anomalies are the second leading cause of sudden death in young athletes, with the anomalous origin of the RCA in the left coronary sinus with an interarterial course (between the aorta and pulmonary artery) being the most associated with this outcome [5,6].

The cases presented here address the incidental finding of three distinct anomalous origins. The first reports a LAD with duplicated origin, in which the emergence of a short LAD from the LMCA can be observed, and a terminal segment is originated from the RCA. This is a benign and rare abnormality, with a prevalence of less than 1%, with no association with hemodynamic repercussions, despite being a complicating factor in surgical approaches [7,8].

The second case concerns the presence of a single anomalous coronary artery, which may originate from either the right or left coronary sinus [7]. In this case, it arises on the left, following the normal course of the LMCA bifurcation in LAD and LCxA, but with the emergence of the RCA from the distal segment of the LCxA. Once again, this is an extremely rare occurrence, with a prevalence of 0.024%-0.066% in the general population and with few reports in the literature [7]. There is an association with myocardial ischemia in approximately 15% of patients with this anomaly, regardless of the presence of coronary atherosclerotic disease [9]. Similar cases have been reported, with the presence of a single coronary artery arising from the left trunk of Valsalva, with dominance of the LCxA and an aberrant branch arising from its distal portion, crossing the crux and continuing to the right atrioventricular groove [10], exactly as seen in our case report.

The third case refers to the anomalous origin of the LCxA from the proximal segment of the RCA, one of the most prevalent coronary anomalies, occurring in approximately 0.32% to 0.67% of the population [11]. This anomaly may present several patterns of origin, such as originating in a common or separate ostium of the RCA or originating in its proximal segment, as in the case presented. In most of these cases, the LCxA will have a retroaortic course, between the posterior region of the aorta and the interatrial septum, a location where coronary vessels are not commonly found and, when present, are generally associated with anomalies of the LMCA or the LCxA [12]. Transthoracic echocardiography can display a sign that is suggestive, but not diagnostic of this anomaly. The so-called retroaortic anomalous coronary sign (RAC sign), is defined by a hyperechoic, tubular image located on the atrial face of the atrioventricular groove and perpendicular to the aorta [13]. Coronary computed tomography angiography (CCTA) can be used as a confirmatory method for this coronary anomaly. A series of three cases demonstrated the agreement between the RAC sign and the anomalous LCxA from the right coronary sinus with a retroaortic course on CCTA [14].

Imaging methods play a central role in the diagnosis of coronary anomalies, since, in most cases, they are incidental findings during the investigation of other diseases. Our three cases were identified by coronary angiography, a highly accurate method that is considered the gold standard for detecting coronary stenosis. Since this is an invasive examination that uses nephrotoxic contrast and ionizing radiation, CCTA and magnetic resonance imaging have often been preferred over coronary angiography [3,6]. The American Heart Association strengthens the recommendation of CCTA for the diagnosis of coronary artery anomalies, with the disadvantage of using ionizing radiation compared to magnetic resonance imaging. Defining the course of an anomalous coronary artery is more appropriate with the use of CCTA, whilst it may be difficult with coronary angiography alone. Hence, novel techniques have emerged that use lower doses of radiation while maintaining a high standard of resolution [15,16].

Congenital coronary anomalies constitute a broad spectrum of rare congenital abnormalities with diverse clinical manifestations, ranging from asymptomatic patients to the occurrence of sudden death in young athletes. These anomalies are still poorly investigated and, consequently, diagnosed, hence the importance of advancing imaging methods as a diagnostic tool and the need for further studies on this subject. We reported three cases involving anomalous origins of the LAD, the RCA and the LCxA. The age over 50 years in all reported patients adds a challenge to clinical decision making.

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