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Variant Origin of the Right Coronary Artery from the Left Main Coronary Artery Together with Aortic Valve Stenosis: Report of a Rare Case

Origen Variante de la Arteria Coronaria Derecha a Partir de la Arteria Coronaria Izquierda Junto con Estenosis de la Valva Aórtica: Reporte de un Caso Raro

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SUMMARY: Variations in the origin of the right coronary artery have an incidence between 0.09 % and 0.92 %. Herein, we report a rare case of a coronary artery anomaly in which the right coronary artery originates from the left main coronary artery. This variant was found during routine coronarography, combined with an artificial aortic valve. Despite their rare occurrence, some variations in the origins of the coronary arteries can be life threatening and are associated with a higher risk of sudden cardiac death. They can also pose serious technical challenges and predispose to complications during coronary angiographic procedures. Thus, knowledge of such anomalies is paramount for managing the patients correctly.

KEY WORDS: Right coronary artery; Anatomical variation; Coronarography; Angiogram.

INTRODUCTION

The coronary arteries (CA) supply blood to the heart. Normally, there are two main coronary arteries, left and right. The left coronary artery (LCA) originates from the left aortic sinuses (sinus of Valsalva) and quickly bifurcates into the anterior interventricular artery (AIA) (left anterior descending artery) and the left circumflex artery (LCx). The AIA mainly supplies the anterior portion of the left ventricle, whereas the LCx supplies the lateral portion of the left ventricle and the superior region of the inferior part of the interventricular septum. The right coronary artery (RCA) usually originates from the right aortic sinuses (sinus of Valsalva) and has a descending course on the right margin of the heart. It gives branches supplying blood to the right ventricle, right atrium and the sinoatrial node (Standring *et al*., 2008).

The frequency of coronary artery anomalies (CAAs) is reportedly between 0.3 % (Alexander & Griffith, 1956) and 3.06 % (Sidhu *et al*., 2019). Variations in the origin of the RCA are rare and range between 0.09 % (Ayalp *et al*., 2002) and 0.92 % (Angelini *et al*., 1999). CAAs can be divided into two broad categories, benign and malignant, depending on their point of origin and course. Approximately 20 % of all CAAs are associated with increased

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cardiovascular risk and sudden cardiac death (SCD) (Taylor & Virmani, 2001; Datta *et al*., 2005).

Despite their rarity, detailed knowledge of possible CAAs is essential for invasive cardiology and cardiac surgeons to diagnose and treat such cases correctly. The aim of this article is to report a rare anatomical variation in the origin and course of the RCA and to highlight the importance of distinguishing such a variation for correct diagnosis and subsequent treatment of the patient.

An RCA arising from the left main coronary artery (LMCA) is an extremely rare anomaly described in a few case reports (Singam *et al*., 2019). The combination of this anomaly with aortic valve stenosis is even rarer. According to Vallabhajosyula *et al*. (2021) the frequency of an anomalous coronary origin from the opposite aortic sinuses (sinus of Valsalva) is higher in patients with a left atrioventricular valve (bicuspid aortic valve) than a right atrioventricular valve (tricuspid aortic valve).

CASE REPORT

We report an extremely rare variation in the origin of the RCA from the LMCA combined with a severe aortic stenosis, previously corrected surgically by implanting a bioprosthesis.

A 55-year-old female was admitted to our department with complaints of exertional angina and shortness of breath for two weeks. Her cardiovascular risk factors were arterial hypertension, dyslipidemia and smoking. The ECG revealed sinus rhythm, criteria for left ventricular hypertrophy and new dynamic ST depressions in anterior leads. The laboratory tests were normal.

Medical history: In 2017, she was admitted to our clinic for heart failure symptoms and 2diagnosed with severe aortic stenosis according to transthoracic echocardiography data: fused aortic valve leaflet with severe calcification, mean gradient of 50 mmHg and aortic valve area of 0.6 cm^2. However, there was no medical documentation of the treatment. Because of the patient's young age, we could only suppose there was a left atrioventricular valve (bicuspid aortic valve) anatomy. She was advised to proceed to surgical treatment, which she chose to undergo abroad.

At the current hospital admission she gave information about the surgical replacement of the aortic valve five years earlier, but no medical documents were available. Transthoracic echocardiography revealed a preserved ejection fraction of 70 %, symmetric left ventricular hypertrophy, no cardiac wall motion abnormalities, and an aortic bioprosthesis with preserved function.

Despite optimal medical therapy, the patient had recurrent chest pain, so a coronary angiography was performed. The LCA was successfully catheterized with a 5F JL 3.5 catheter, which revealed an unexpected finding: an RCA originating entirely from the LMCA (Fig. 1a). Furthermore, coronary angiography revealed non-obstructive atherosclerostic coronary artery disease in the ostio-proximal part of the anomalous RCA. Aortography showed an aortic bioprosthesis with normal function, lack of insufficiency, and no visible coronary artery arising from the right aortic sinuses (sinus of Valsalva) (Fig. 1b).

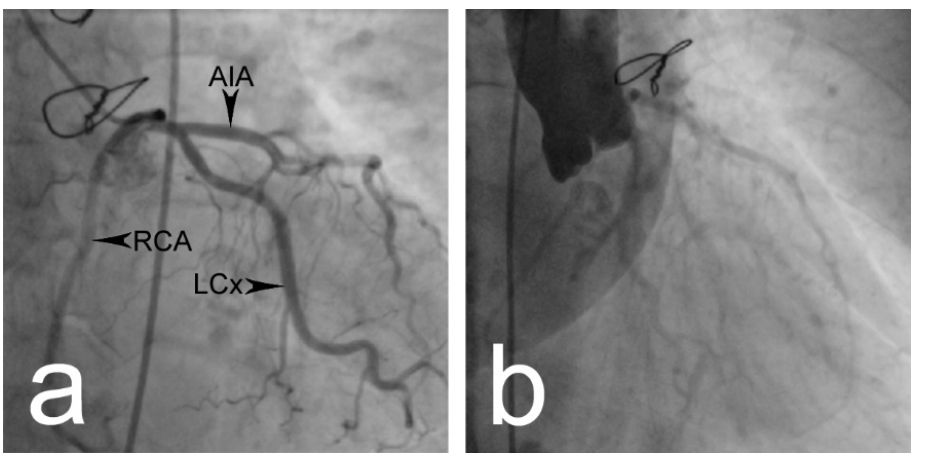

Fig. 1. A) Coronary angiographic views showing variant origin of the right coronary artery (RCA) from the left main coronary (LMCA); anterior interventricular artery (AIA) and left circumflex artery (LCx) at their usual location; B) aortography, showing no coronary arteries arising from the right aortic sinuses (sinus of Valsalva) with a competent aortic bioprothesis.

DISCUSSION

Variations in the origins of CAs are seldom encountered, but the literature provides several studies of CAAs. Alexander & Griffith (1956) reported a 0.3 % incidence rate of CAAs during an autopsy study. According to Yamanaka & Hobbs (1990), the mean incidence rate of CAAs among the 126,595 patients included in their angiographic study was 1.3 %. Sidhu *et al*. (2019) recorded a 3.06 % incidence rate of CAAs during their angiographic study of 3,233 patients. In those studies, the most commonly encountered variations were in the origins of the LCx and AIA (Alexander & Griffith, 1956; Yamanaka & Hobbs, 1990; Sidhu *et al*., 2019). Variations in the RCA are less common. According to the angiographic study by Ayalp *et al*. (2002) the mean incidence of anomalous origin of the RCA was 0.09 % of 5253 patients. These authors reported a 0.03 % incidence of an RCA originating from the left aortic sinuses (left sinus of Valsalva) and 0.05 % from an ectopic ostium above the left aortic sinuses (left sinus of Valsalva) (Ayalp *et al*., 2002). The incidence of RCAs originating from the left aortic sinuses (left sinus of Valsalva) has been reported as 0.107 % (Yamanaka & Hobbs, 1990), 0.018 % (Yuksel *et al*., 2013), and 0.37 % (Sidhu *et al*., 2019). However, in our case, there was a single coronary ostium and the RCA originated from the main part of the LCA. The frequency of a single coronary ostium is reported to be 0.024 % (Lipton *et al*., 1979). It has previously been reported by Arteaga *et al*. (2006), Swaminath *et al*. (2013), Singam *et al*. (2019) and similar to our case. This variation can be suspected when no right coronary ostium has been localized in the RSV after multiple attempts and no collateral is visible (Yamanaka & Hobbs, 1990). A CT scan is advised to visualize the course of the aberrant RCA (Singam *et al*., 2019).

Several classifications have been proposed for appropriate systematization and categorization of CAAs. Angelini *et al*. (2002) provided the most complete and complex one, considering the origin, course, anastomoses, intrinsic anatomy and termination of CAAs. According to this classification, our case would be identified as A4a4. The classification by Dollar & Roberts (1989) is more straightforward and only considers the number of coronary ostia. According to this classification, our case is Type I.

In most cases, CAAs are discovered accidentally or during autopsy (Alexander & Griffith, 1956, de Oliveira *et al*., 2012). Their origin and course indicate that they can be divided into benign and potentially serious (Yamanaka & Hobbs, 1990). An RCA originating from the left aortic sinuses (left sinus of Valsalva) or the LCA has three different courses: anterior to the aorta, between the aorta and pulmonary trunk (intraarterial) and posterior to the pulmonary trunk (Yamanaka & Hobbs, 1990). Only the intraarterial course is deemed potentially serious, being associated with angina, arrhythmias, myocardial ischemia and increased risk of SCD (Yamanaka & Hobbs, 1990). In our case, the fact that cardiac surgeons avoided reimplantating the anomalous RCA led us to suppose that a benign course of this artery had been found intraoperatively. According to D'Ascenzi *et al*. (2022) 7.2 % of SCDs among athletes and 1.9 % among nonathletes are caused by CAAs. Moreover, CAAs can lead to serious diagnostic and therapeutic difficulties, especially for less experienced interventional cardiologists, owing to the complexity of the procedure. There is no agreed protocol for managing such patients since there are no guidelines on the matter because the cases are rare.

CONCLUSION

Encountering CAAs during routine coronary angiography is a rare yet plausible scenario. Thus, knowledge of such variations is paramount for identifying and managing such patients correctly. Moreover, it is crucial to know whether the anomaly is combined with aortic valve stenosis.

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RESUMEN: Las variaciones en el origen de la arteria coronaria derecha tienen una incidencia entre el 0,09 % y el 0,92 %. En este documento, informamos un caso raro de una anomalía de la arteria coronaria en la que la arteria coronaria derecha se originaba en la arteria coronaria izquierda. Esta variante se encontró durante una coronariografía de rutina, combinada con una válvula aórtica artificial. A pesar de su rara aparición, algunas variaciones en los orígenes de las arterias coronarias pueden poner en peligro la vida y se asocian con un mayor riesgo de muerte súbita cardíaca. También pueden plantear serios desafíos técnicos y predisponer a complicaciones durante los procedimientos angiográficos coronarios. Por tanto, el conocimiento de dichas anomalías es fundamental para el manejo correcto de los pacientes.

PALABRAS CLAVE: Arteria coronaria derecha; Variación anatómica; Coronarografía; Angiograma.

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