CASE REPORT

Unroofing as treatment for anomalous aortic origin of the left coronary artery. A case report

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Anomalous aortic origin of the left coronary artery from the right Sinus of Valsalva is extremely rare and is a cause of sudden cardiac death of non-atherosclerotic ischemic etiology in patients younger than 35 years of age. We present a case of a patient with an anomalous aortic origin of the left coronary artery with an intramural trajectory treated with the unroofing technique.

Key words: Coronary arteries, anomalies; Sudden cardiac death; Unroofing.

El origen aórtico anómalo de la arteria coronaria izquierda del Seno de Valsalva derecho es en extremo raro y es una causa de muerte súbita cardiaca de etiología isquémica no ateroesclerótica en pacientes menores a 35 años. Reportamos un caso de una paciente con origen aórtico anómalo de la arteria coronaria izquierda con trayecto intramural tratada por medio de la técnica unroofing.

Palabras clave: Arterias coronarias, anomalías; Muerte súbita cardiaca; Unroofing, .

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Coronary artery anomalies (CAA) are a diverse group of congenital disorders whose manifestations and pathophysiological mechanisms are highly variable [1]. According to the literature, coronary anomalies affect 1% of the general population; this percentage is derived from cineangiograms performed for suspected obstructive disease [2]. Many of the CAA have no clinical repercussion, without needing any intervention. However, anomalous aortic origin of a coronary artery (AAOCA) deserves special attention, since it can impede myocardial perfusion intermittently or chronically, leading to clinical manifestations such as angina, myocardial infarction, congestive heart failure, ventricular aneurysms and sudden cardiac death [3].

The most recent guidelines of the European Society of Cardiology for the treatment of congenital heart disease in adults (2020) recommend surgical treatment in patients with AAOCA in the following circumstances: asymptomatic patients with evidence of ischemia or high-risk anatomy (IIa C); symptomatic patients even without evidence of ischemia or high-risk anatomy (IIb C), and for asymptomatic patients younger than 35 years with AAOCA of the left coronary artery even without evidence of ischemia or high-risk anatomy (IIb C) [4]. Surgical strategies for the treatment of AAOCA basically consist of the following procedures: aortocoronary bypass; ostium reimplantation; unroofing and modified unroofing [5]. We report the case of a patient with anomalous aortic origin of the left coronary artery from the right sinus of Valsalva with an intramural route.

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CLINICAL CASE

A 39-year-old female patient, with no significant chronic degenerative or surgical history began one month prior to her admission with typical angina and posterior syncope. She was taken to the emergency room where the diagnosis of non-ST elevation myocardial infarction was made. Diagnostic coronary angiography was carried out, observing an anomalous origin of the left coronary artery, which originates from the right Sinus of Valsalva, the rest of the coronary arteries did not present lesions or abnormalities. The diagnostic approach is complemented with coronary CT angiography, corroborating the origin of the left coronary artery from the right sinus of Valsalva with an interarterial path (**Fig. 1**) between the pulmonary artery and the aorta. A transthoracic echocardiogram was also performed, reporting an ejection fraction of the left ventricle of 54% without valve disease or other significant findings. In this context, the cardiology team decided to take the patient for surgical correction of the congenital coronary anomaly.

Surgical procedure

The cardiac approach was performed through a median sternotomy, corroborating the anomalous origin of the left

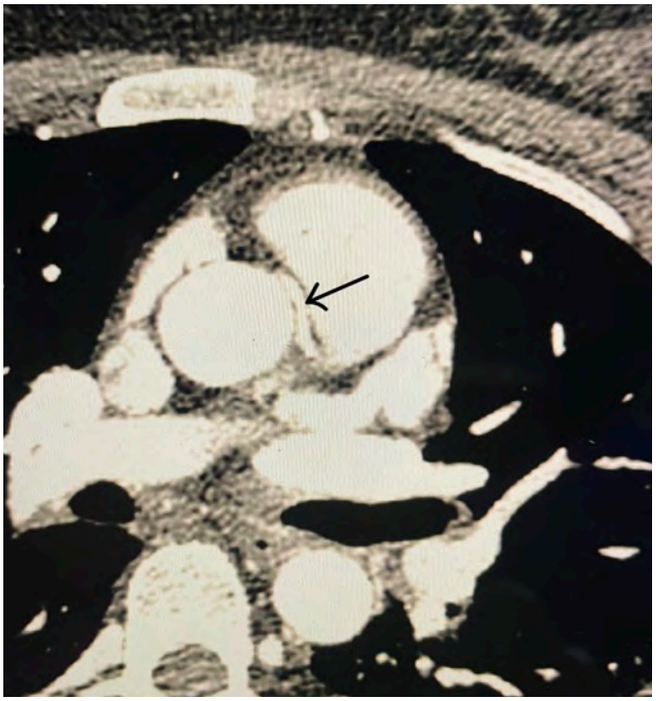

Figure 1. Figure 1. Axial Computed Tomography (CT) scan section showing the origin of the left coronary artery from the right Sinus of Valsalva and its interarterial course (arrow).

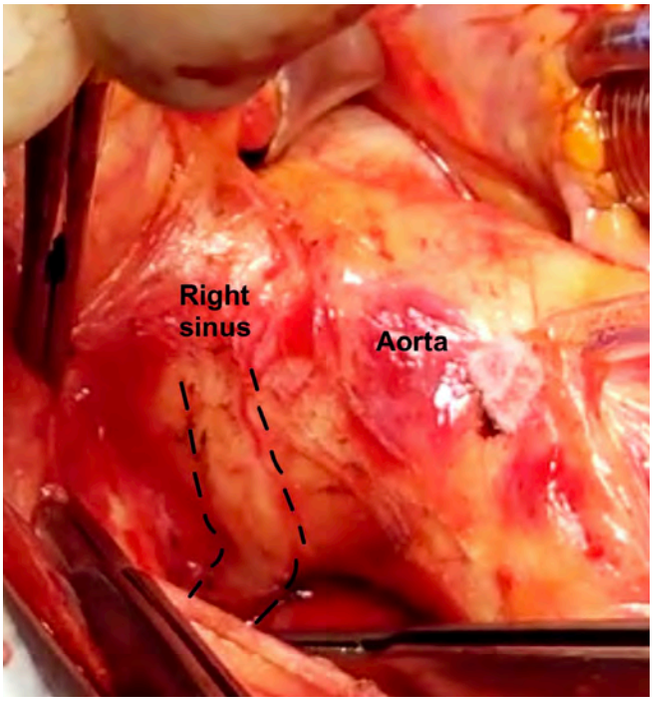

Figure 2. Surgical image showing the origin of the left coronary artery in the right Sinus of Valsalva and its intramural segment (dotted line).

main coronary artery arising from the right sinus of Valsalva with an intramural trajectory (**Fig. 2**). It was decided to use cardiopulmonary bypass with central cannulation in the ascending aorta and right atrium, aortic clamping, and cardiac arrest with anterograde cardioplegia. Transverse aortotomy was performed, the left coronary ostium in the right sinus of Valsalva is identified, and a coronary probe is inserted to identify the intramural trajectory. The coronary "unroofing" technique is performed to create a wide neo-ostium up to the coronary emergency site in its "correct" sinus. Fixation sutures with 7-0 polypropylene were placed between the neo-ostium and the aorta to prevent subsequent intimal tearing. A coronary probe was inserted into the neo-ostium through the emergence of the left coronary artery on the aorta, observing a new course without angulation and without extrinsic compression (**Fig. 3**). The aortotomy was closed. Aortic unclamping and cardiopulmonary bypass weaning as usual, with a total cardiopulmonary bypass time of 91 minutes and aortic cross-clamping time of 64 minutes. She had a satisfactory postoperative evolution, being discharged from hospital on the fifth postoperative day. The control echocardiogram did not show aortic insufficiency. At 3-month follow-up, the patient had no other cardiovascular event, such as syncope, angina, dyspnea, remaining in functional class I (NYHA).

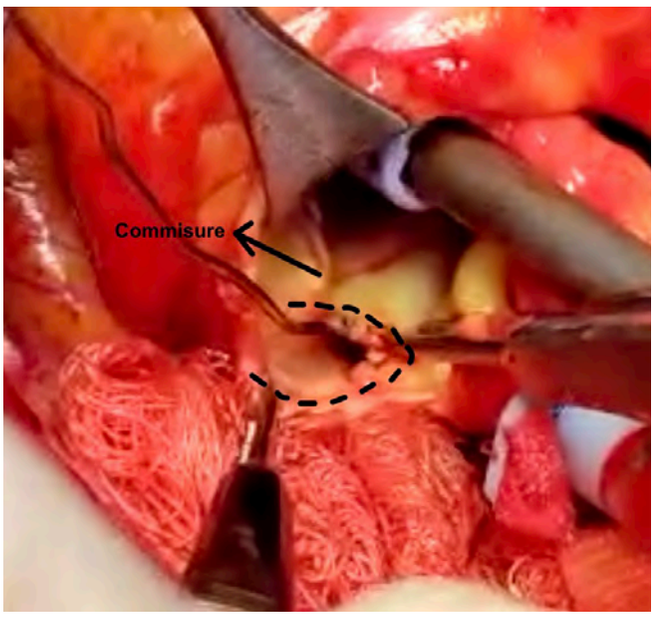

Figure 3. Surgical image showing the neo-ostium after the unroofing and its distance from the commissure.

COMMENT

CAA affect about 1% of the general population; this percentage is derived from cineangiograms performed for suspected obstructive coronary disease [2]. Autopsies show an even lower incidence: in 18,950 autopsies, Alexander and Griffith observed only 54 coronary anomalies (0.3%) [6]. Anomalous aortic origin of the left coronary artery arising from the right sinus of Valsalva is extremely rare: in 1950 angiograms performed by Angelini, this variant was only found in 3 patients (0.15%) [1]. AAOCA has been identified as a cause of sudden death of non-atherosclerotic ischemic etiology in patients younger than 35 years of age, especially in athletes [7]. Therefore, the most recent guidelines of the European Society of Cardiology

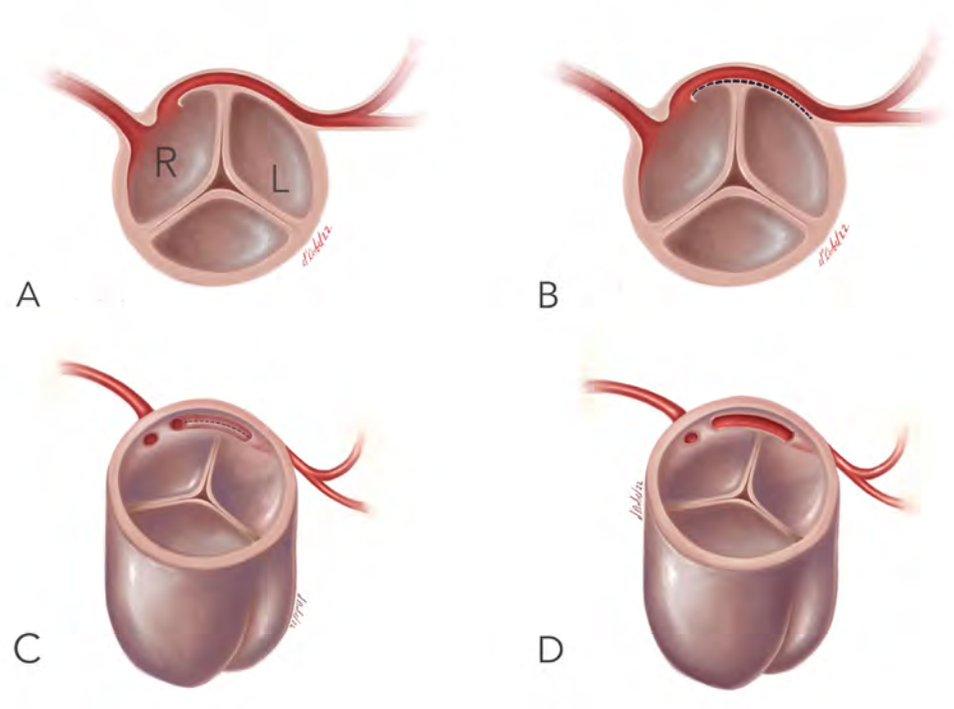

Figure 4. Diagram that exemplifies the realization of the unroofing technique.

for the treatment of congenital heart disease in adults (2020) recommend surgical treatment in patients with AAOCA in the following circumstances: asymptomatic patients with evidence of ischemia or high-risk anatomy (IIa C); symptomatic patients even without evidence of ischemia or high-risk anatomy (IIb C) and asymptomatic patients <35 years with anomalous aortic origin of the left coronary artery even without evidence of ischemia or high-risk anatomy (IIb C) [4].

There are 4 surgical techniques for the treatment of AAOCA, each with its advantages and limitations: 1) Aortocoronary bypass surgery, 2) Reimplantation of the artery in the appropriate coronary sinus, 3) Unroofing and 4) Modified Unroofing [5]. Jegatheeswaran et al. reviewed the surgical results of 395 surgical patients in a cohort of 682 patients from 45 North American centers, finding that the most widely used repair technique was unroofing (87%) [8]. This technique should be applied only in patients with a long intramural course to the appropriate cusp and without contact with the commissure to avoid the postoperative risk of aortic regurgitation. In patients with a short intramural course, unroofing may not address extrinsic compression of the coronary artery between the aorta and pulmonary artery [9]. In our case, the patient had a long intramural path (approximately 2.5 cm) to the "correct" sinus and an upper commissure separation between the right and left coronary leaflets of approximately 0.7 cm, which allowed coronary unroofing to be performed without damaging the aortic valve, avoiding any potential residual compression of the left coronary artery between the aorta and the pulmonary artery.

We can conclude by saying that anomalous origin of the left coronary artery arising from the right Sinus of Valsalva is extremely rare and is a cause of non-atherosclerotic ischemic sudden cardiac death in patients younger than 35 years of age, especially athletes. Unroofing consists of exploration and confirmation of the intramural course with a coronary probe. The common wall between the aorta and the coronary artery is precisely excised or an incision is made over the probe throughout its entire intramural course proximal to the site where it emerges from the aortic wall. Sutures are placed to ensure continuity of the intima and prevent future dissection. A wide neo-ostium is formed in the "correct" Sinus of Valsalva, therefore the unroofing procedure includes a coronary osteoplasty component (**Fig. 4**).

Unroofing is the preferred technique by surgeons to treat the AAOCA. To guarantee the success of this technique, it is essential that the intramural trajectory be long enough to avoid residual compression of the coronary between the aorta and the pulmonary artery, as well as its superior separation from the aortic valve commissure to avoid postoperative aortic regurgitation.

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