

## Atypical origin of the left coronary artery originating from the right coronary sinus with interarterial course: A case report

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### ABSTRACT

**Background:** A left coronary artery with atypical origin originating from the right coronary sinus with interarterial course was found in a 68-year-old man, who had been referred to the hemodynamics unit for ventricular extrasystoles, observed on ECG, which raised suspicions of suspect coronary heart disease.

**Methods:** Atypical origin was observed on a coronary angiography, and interarterial course was confirmed with a coronary CT.

**Results:** The left coronary artery is observed arising from the right aortic sinus, in an ostium separated by 5 mm to the left of the right ostium. The left coronary trunk was 28 mm long, with eccentric calcium plaque in the distal third of the coronary trunk, and 50% stenosis. The right coronary artery was of normal caliber.

**Conclusions:** Atypical origin of the left coronary artery is a rare condition, normally associated with sudden death.

### 1. Introduction

Atypical origin of the coronary arteries is a rare condition, with a reported incidence of 1.3%. The atypical origin of the left coronary artery from the right sinus with interarterial course has a prevalence of 0.17%, and a risk of death of 0.24/100,000 per person-year [1]. However, abnormal coronary arteries are the second leading cause of sudden cardiac death (SCD) in Europe and North America, with an incidence ranging from 12.2% to 17.2% [2]. Left coronary artery anomaly (LCAA) is most commonly associated with young patients during or after strenuous exercise, but elderly patients are usually asymptomatic. We may need to use Left coronary artery anomaly (LCAA) detection during or after strenuous exercise abnormality symptoms. Both the symptomatology and the SCD in these patients are often the result of a reduced diastolic coronary flow, which can be explained by several mechanisms: Interarterial compression during exertion, narrowing of the ostium, intramural course stenosis, exit at an acute angle with possible torsion, and spasm or twisting of the abnormal coronary artery [3,4]. Anatomical variations have a wide range of morphological diversity, from the most typical pattern to infrequent variations [5]. Five anatomical

subtypes are reported, which are classified according to the relationship of the abnormal coronary artery with the aorta and the pulmonary artery. These are “anterior”, “inter”, “septal”, “posterior” and “combined”. The “septal”; subtype is the most common, while the “inter” type present in this case is rare [3].

Given the high risk of SCD, patients with an anomalous left coronary artery should undergo surgical correction. There are several possible surgical techniques to address these underlying pathophysiological mechanisms, such as fenestration or unroofing of the intramural component, reimplantation of the anomaly into the correct sinus, coronary artery bypass grafting, and pulmonary artery translocation to avoid interarterial compression [6,7].

This work is important for daily clinical practice because it presents coexisting clinical situations that are often seen in very different contexts. On one hand, coronary anomalies have a much greater impact on people under 35 years of age. On the other hand, atherosclerosis is a more prevalent phenomenon in older people like our patient. The presence of obstructive coronary disease also impacted the surgical treatment plan. In addition, many asymptomatic patients undergo CT angiography, so we must be prepared to manage patients with coronary

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anomalies such as the one described in this work.

## 2. Case presentation

A 68-year-old Chilean man presented with a history of hypertension, carbohydrate intolerance, and occasional smoking. His brother had died suddenly of an acute myocardial infarction. During preventive control, bradycardia was screened. Frequent ventricular extrasystoles were observed in a 12-lead electrocardiogram (ECG). The initial study was complemented with a two-dimensional echocardiogram which demonstrated mild left ventricular systolic dysfunction, and 24-h ECG monitoring that showed very frequent ventricular extrasystoles (Fig. 1). In the 24-h electrocardiogram, ventricular extrasystoles of two different morphologies were observed, mostly isolated and also bigeminate and trigeminate. As a whole, ventricular extrasystoles represented 24% of the total recorded beats. Most of the ventricular extrasystoles had a left bundle branch block morphology, suggesting a right ventricular origin.

We thus decided to perform a coronary angiography, showing a left coronary artery originating in the right coronary sinus (Fig. 2A) with possible interarterial course. An anterior descending artery of discrete caliber was observed as well, presenting diffuse atheromatosis with a mild proximal lesion and fine distal bed (Fig. 2B). A circumflex artery of medium caliber with good development originated two branches of good caliber, with a mild obstructive distal lesion. The right coronary artery is of large caliber and development, without significant obstructive lesions. Ventriculography showed a left ventricle of normal size and left ventricular ejection fraction, although the study was limited by frequent ventricular extrasystoles (Fig. 3).

In order to confirm the interarterial route of the left coronary artery, a coronary angio-CT was performed, which showed a heart of normal general size, without any intracavitary filling defects suggestive of thrombi. The right coronary artery shows habitual origin in the right aortic sinus. An atypical origin of the left coronary artery was observed (Fig. 3A), which also originates from the right aortic sinus, in an ostium

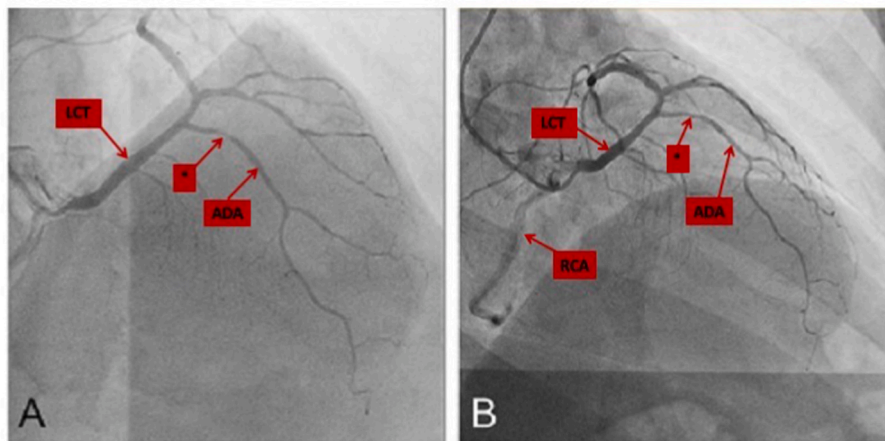
separated approximately 5 mm to the left of the origin of the right coronary artery. Under these conditions, we observed the left coronary trunk of interarterial trajectory measuring approximately 28 mm in length, demonstrating normal caliber. There is eccentric calcium plaque in the distal third of the coronary trunk, indicating stenosis of approximately 50% (Fig. 3B). The circumflex artery is permeable, with eccentric calcium plaques in its proximal and middle third, which do not determine significant stenosis. The anterior descending artery presents calcium plaque in its origin, which gives an indication of stenosis greater than 50%. The right coronary artery (dominant vessel) is of normal caliber and permeable throughout its extension, with multiple foci of calcium atheromatosis in its origin, middle third and distal third, without luminal stenosis foci.

## 3. Discussion

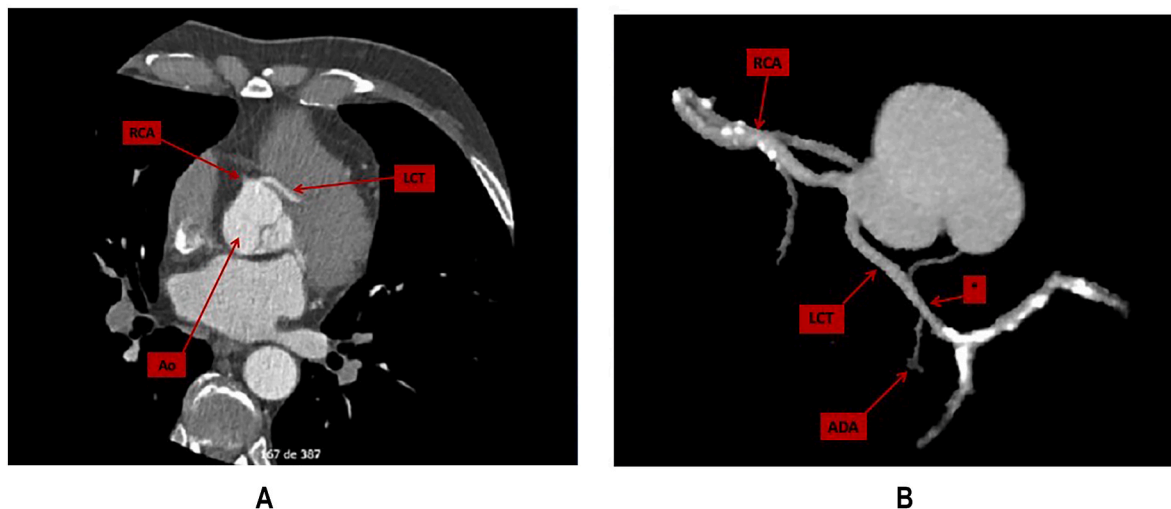
Coronary arteries with abnormal origin are a rare pathology. The anomaly of the left coronary artery arising from the right aortic sinus is even more rare [3]. Subtypes have been described according to the course of the anomalous vessel, in reference to the great vessels. The interarterial course or malignant course [8] is especially severe, since it is directly related to an increased risk of sudden cardiac death. The literature contains various cases associated with major cardiovascular events. Different theories have been postulated as a cause of sudden death in these patients. The most accepted theory is the higher incidence of ostium occlusion, secondary to a more slit-like orifice and occlusion during physical activity, due to compression between the major arteries [4]. This makes it all the more notable that our patient had no history of classic cardiac symptoms (angina, syncope, dyspnea, and palpitations), and also had no history of previous cardiovascular events. It is worth mentioning that cases of elderly people with asymptomatic coronary artery anomalies have already been described, especially the difference in sudden death risk compared to young patients, where it has been proposed that arterial wall thickenings could serve as protection against



**Fig. 1.** Results of the rhythm holter performed with Mortara equipment. Total record of 22 hrs. with 92271 total beats, showing 22876 ventricular extrasystoles: A. Trace of the aVL derivative, containing 22452 monomorphic isolated ventricular beats lasting 7.5 seconds (arrows) B. Trace of the aVL derivative, with ventricular bigeminism (\*).



**Fig. 2.** Coronary angiography performed with a Siemens allura F20 A angiograph. Right cranial projection 10/40° showing a long left coronary trunk (LCT) with a thin anterior descending artery (ADA), with tortuosities and proximal stenosis of 40% (\*). B. Anterior oblique right projection (30°) showing a long LCT with a thin ADA, with tortuosities and proximal stenosis of 50% (\*). While partially opacifying the right coronary artery (RCA).



**Fig. 3.** Angio-TC A. Cut at the level of origin of the LCT from the right aortic sinus and path between aorta artery (Ao). B. Processed image demonstrating severe calcification of distal LCT, with 50% stenosis (\*) (arrows).

interarterial segment compression, thus avoiding ischemia by occlusion [3,9–11].

Defining the course followed by an artery in a two-dimensional projection such as coronary angiography is not easy, even in expert hands. Some methods can help for this purpose [12,13], but they are not 100% accurate. Determining abnormal arteries' courses is important in these patients, as it determines the severity of the alteration. There are useful alternative tests for this, such as conventional or transesophageal echocardiography, and other more recent ones such as coronary angio-CT or cardiac magnetic resonance, which let us precisely define the origin and course of each coronary artery [14].

Angio-CT has been a great contribution to characterizing the anomalous origin of coronary arteries, since it provides essential information unavailable from invasive angiography. In this particular case, it was necessary to determine the course of the coronary left artery after its beginning in the right coronary sinus, since we know that the “inter-arterial” course is the one associated with an increased risk of sudden cardiac death. The latter options have the advantage of obtaining three-dimensional images, but they are expensive, are not always available, and also have some risk due to the use of radiation and contrast medium. They are thus not indicated for every case.

Our patient underwent a coronary angiography, and although it managed to determine the type of anomaly regarding the origin of the left coronary artery, its performance was insufficient to identify the course. This made it necessary to expand the study with a coronary angio-CT that finally revealed the interarterial course. It is highly important to do so with these patients, because it provides vital information and allows for definitive treatments [11,15–19], such as coronary angioplasty [20] or bypass surgery [21].

Our patient underwent an invasive coronary study due to frequent ventricular arrhythmia detected on a 24-h ECG. After starting treatment with carvedilol 6.25 mg every 12 hours, the patient had his ventricular extrasystoles drop, and he has remained asymptomatic. His case was discussed by the Heart Team, and he was offered coronary revascularization surgery, which the patient ultimately refused. As of today, he remains asymptomatic and is receiving medical treatment that includes Enalapril (5 mg every 12 hours), Atorvastatin (20 mg daily), carvedilol (6.25 mg every 12 hours), and acetylsalicylic acid (100 mg daily). What we were looking for was atherosclerotic coronary disease. An angio-CT was performed because invasive coronary angiography is inefficient for interarterial course determination. Angio-CT guarantees the inter-arterial course, and also increases the presence of calcified plaque with



50% stenosis in the distal left main coronary artery. The AHA guidelines on congenital heart disease in adults [22] recommend surgery for asymptomatic patients with this pathology with a class II A rating. However, during a meeting of the local Heart Team, it was decided that surgery should be performed, due to the presence of significant coronary disease of the left main coronary artery and frequent ventricular arrhythmia. We opted not to perform additional ischemia testing because previous research has shown that the absence of ischemia does not eliminate the risk of sudden death in these patients [23].

Studies indicate that coronary artery embryology depends on vasculogenesis, and may have genetic determinants [24]. The literature presents a few cases of coronary artery anomalies of similar characteristics within the same family. These relationship findings [25], although sporadic, would point to a potential genetic substrate linked to coronary artery anomaly development. More scientific evidence is still required for confirmation. Similarly, our patient has a history of a first-degree relative who died as a result of sudden cardiac death. This information could indicate a similar phenomenon, linked to as yet unproven genetic factors.

We believe that this work is important for daily clinical practice, because it shows the great contribution of Angio-CT for characterizing various anatomical subtypes of coronary anomalies. This case also presents coexisting clinical situations that are usually seen in very different contexts. On the one hand, coronary anomalies have a much greater impact on people under 35 years of age. However, in older people like our patient, atherosclerosis is a more prevalent phenomenon. The presence of obstructive coronary disease even impacted the surgical treatment plan, opting for a traditional coronary revascularization strategy over a coronary artery reimplantation technique. Finally, with current developments in preventive medicine, many asymptomatic patients are submitted to Angio-CT. We must therefore be prepared to manage patients with coronary anomalies such as the one described herein, among asymptomatic patients.

#### 4. Limitations and conclusions

- A viability ischemia study (myocardial scintigram or magnetic resonance imaging) was not performed to investigate ischemia, so we cannot determine whether the trunk lesion or coronary anomaly in the patient is causing the ischemia. This is due to the scarcity of public health resources in developing countries.
- The patient refusing to undergo surgery creates a limitation, so we cannot confirm the evolution of the case.
- Atypical origin of the left coronary artery is a rare condition that is usually associated with sudden death.
- Subtypes have been described according to the course of the anomalous vessel, in reference to the great vessels. The interarterial course or malignant course is especially severe.
- Conventional, transesophageal echocardiography, coronary CT angiography, or cardiac magnetic resonance are necessary tests to accurately define the origin and course of each coronary artery.
- Coronary angiography is insufficient to determine the type of anomaly regarding the origin of the coronary artery, as well as to identify the course.

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#### Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence

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