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Case Report

Double coronary anomaly: A case report

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ABSTRACT

We describe an extremely rare case of a 37-year-old female patient who presented with exertional angina and was diagnosed with a unique coronary anomaly with an anomalous right coronary artery with origin in the left anterior descending artery and a fistula between this anomalous coronary artery and the pulmonary artery.

Learning objectives: Most patients with coronary anomalies are asymptomatic but some may have angina caused by a coronary steal phenomenon, myocardial infarction, or even sudden death depending on the circuit and characteristics of the anomaly.

The combination of multiple coronary anomalies is extremely rare.

Despite being a rare diagnosis, coronary anomalies should always be considered as a cause of myocardial ischemia, especially in young patients with low probability for coronary obstructive disease.

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Introduction

Coronary artery anomalies consist of a wide range of defects that alter coronary artery blood flow and are often congenital. Their prevalence ranges from 0.21% to 5.79% according to different sources and definitions [1]. Most patients are asymptomatic, but some may have angina, myocardial infarction, or even sudden death depending on the circuit and characteristics of the anomaly. The combination of multiple anomalies is extremely rare. Coronary to pulmonary fistulas are rare abnormal communications often found between the left coronary artery and the pulmonary artery (PA), origin in the right coronary artery (RCA) is less frequent. Anomalous origin of the right coronary artery (ARCA) as a branch of the left anterior descending artery (LAD) is another very rare anomaly [2].

Case report

A 37-year-old female was referred to a consultant of cardiology. She had complaints of exertional angina, localized in the precordium with irradiation to the left arm and relief at rest for one year and worsening in the past months. On examination, her heart rate and blood pressure were normal, and she had regular and symmetric pulses in the extremities. Heart sounds were normal, no murmur was audible on auscultation, and there were no signs of heart failure on examination.

As cardiovascular risk factors she had dyslipidemia and she was an active smoker. She had no other relevant past medical or family history.

Electrocardiogram (ECG) and thorax radiography were normal. No structural changes were found on the transthoracic echocardiogram. A cardiac stress test was performed and the maximal heart rate was achieved, there were no ECG changes, and the blood pressure and heart rate remained stable, but the patient complained of angina at peak exercise. A few days later a computed tomography angiography (CTA) was performed and a fistula to the PA with origin in the LAD and absence of the RCA were identified (Fig. 1).

The patient was hospitalized for further investigation and observation and two days later an invasive coronary angiography (ICA) confirmed the diagnosis of an ARCA with origin in the LAD and a fistula between this anomalous coronary artery and the PA (Videos 1–6). The case was discussed in a multidisciplinary team with the cardiac surgery center of referral and, considering the presence of a fistula from a high chamber pressure (coronary artery) to a low chamber pressure (PA) and the symptoms suggesting a coronary steal phenomenon, the decision to perform surgical correction was made and other ischemic examinations were dispensed with. A few weeks later, surgical correction was performed with success with ligation of the fistula near its connection to the PA, eradicating the anomalous coronary flow from the fistula

Abbreviations: ARCA, anomalous origin of the right coronary artery; CTA, computed tomography angiography; Cx, circumflex artery; ECG, electrocardiogram; ICA, invasive coronary angiography; LAD, left anterior descending coronary artery; LM, left main coronary artery; PA, pulmonary artery; PDA, posterior descending artery; RCA, right coronary artery; RI, ramus intermedius.

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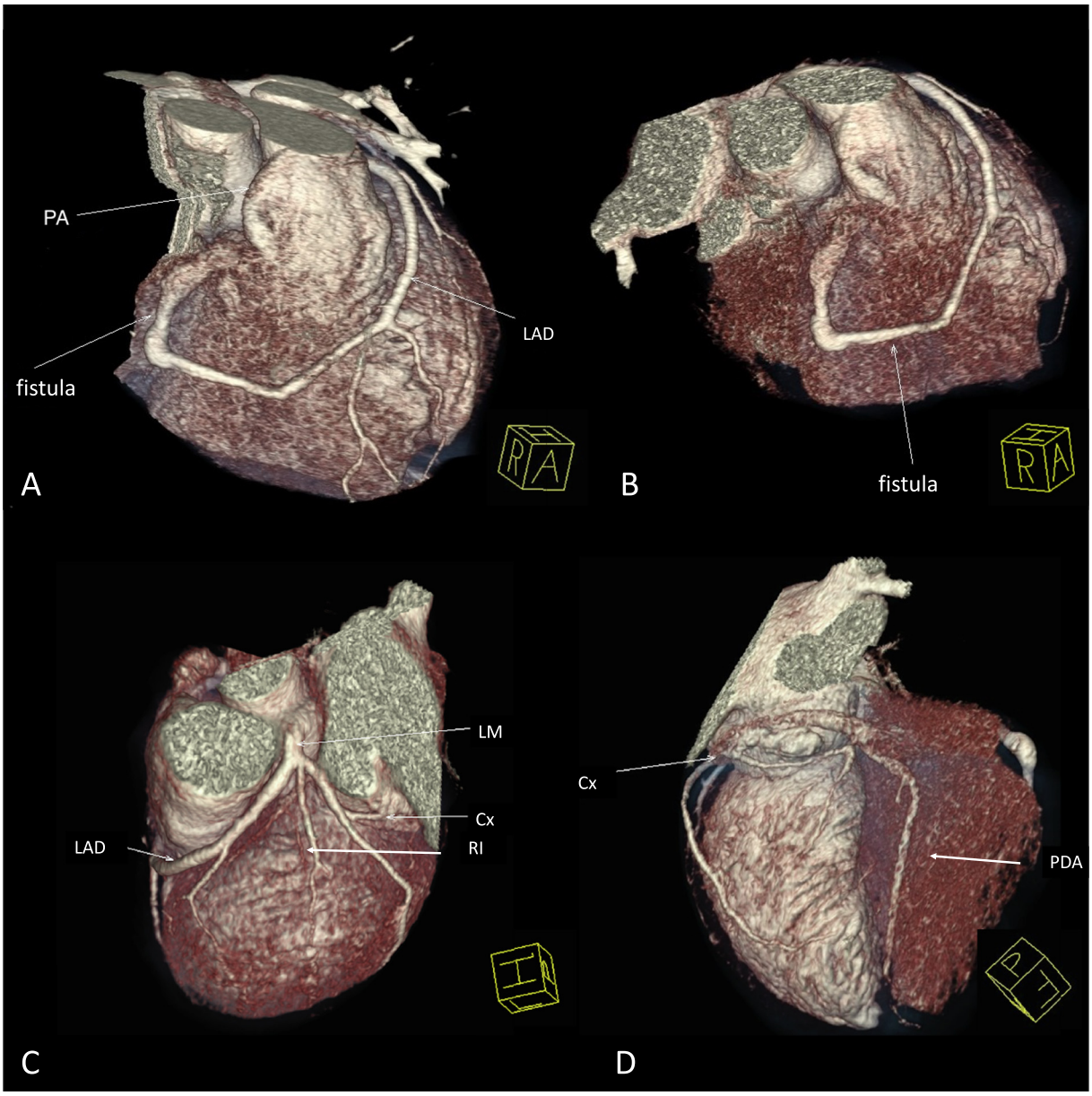

Fig. 1. (A–D). Computed tomography coronary angiography showing a fistula to the pulmonary artery with origin in the left descending artery.
Cx, circumflex artery; LAD, left anterior descending artery; LM, left main coronary artery; PA, pulmonary artery; PDA, posterior descending artery; RI, ramus intermedius.

to the PA (Video 7 and Fig. 2). The patient remained hemodynamically stable without recurrence of symptoms and was discharged a few days later. Since the procedure, the patient remains stable, without recurrence of symptoms or other complications and one year later after the surgical repair, a cardiac stress test confirmed no signs of ischemia or symptoms.

Discussion

We report an extremely rare case of a double coronary anomaly with an ARCA with origin in the LAD and a fistula between this anomalous coronary artery and the PA in a young female patient without other structural abnormalities.

Echocardiography is a non-invasive modality useful to detect and exclude other structural abnormalities and, although it was not focused in assessing the coronary arteries, it was performed earlier in the study of this case.

Considering this was a young patient with typical angina, the pre-test probability of having obstructive coronary artery disease was low [3] and so, the first diagnostic test recommended should be non-invasive. A cardiac stress test was chosen and was clinically positive, requiring a more accurate test. CTA is a less-invasive imaging modality that can confirm the diagnosis but ICA remains the gold standard for diagnosis.

Due to the low prevalence of this anomaly, the optimal treatment strategy remains unclear and depends on the size of the fistula, anatomy, presence of symptoms, age, and the experience of the center. Treatment options include conservative medical management, trans-catheter closure and surgical correction. Most patients with coronary anomalies are asymptomatic and do not require specific treatment. However, potential for life-threatening complications are relevant and may be present in up to 11–19% of young patients and athletes [4] requiring correction. This patient had myocardial ischemia caused by a coronary steal phenomenon

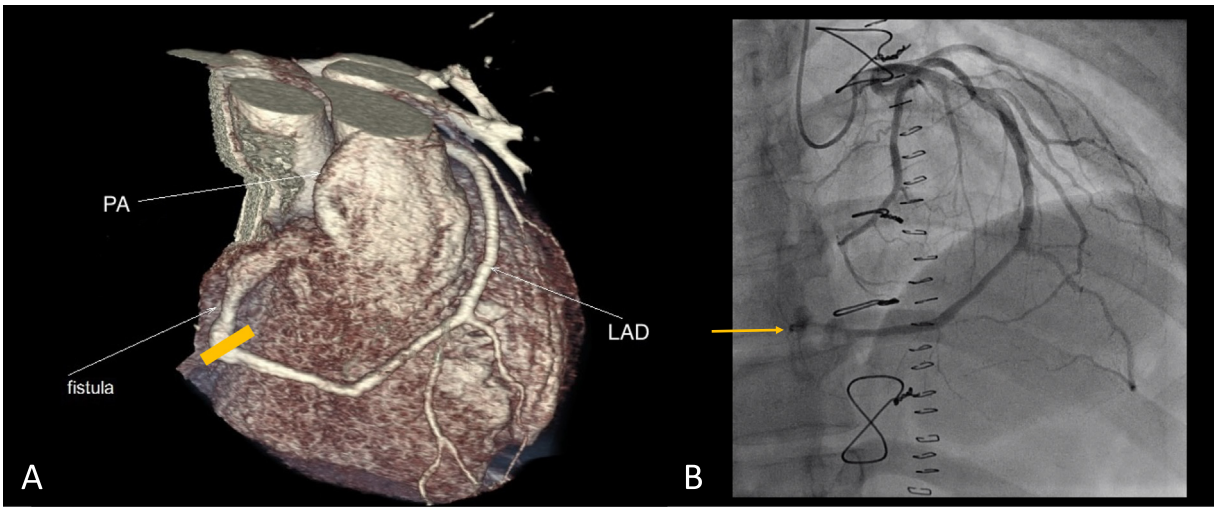

Fig. 2. (A) Computed tomography coronary angiography showing the surgical ligation site of the fistula. (B) Coronary angiography in the right anterior oblique caudal view after excision of the fistula. Yellow arrow and yellow box: incision site.
LAD, left anterior descending artery; PA, pulmonary artery.

secondary to left-to-right shunting from the anomalous coronary to the pulmonary artery and surgical correction was chosen and performed with success [5].

Conclusions

This report describes an extremely rare case of a double coronary anomaly with an ARCA with origin in the LAD and a fistula between this anomalous coronary artery and the PA causing a coronary steal phenomenon with symptoms of angina in a young female patient.

Despite being a rare diagnosis as cause of myocardial ischemia, coronary anomalies should always be considered as a cause of myocardial ischemia, especially in young patients with low probability for coronary obstructive disease.

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Declaration of competing interest

The authors declare that there is no conflict of interest.

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