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INTERMEDIATE

CASE REPORT: CLINICAL CASE

Circumflex Artery Arising From the Pulmonary Artery

Always a Malignant Coronary Anomaly?

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ABSTRACT

Some coronary artery anomalies are associated with increased risk of sudden cardiac death and myocardial infarction in young patients. There are few data on the clinical and prognostic relevance of isolated origin of the left circumflex artery from the pulmonary artery, an extraordinarily rare variant of anomalous left coronary artery from the pulmonary artery. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:1702–7) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A 39-year-old man was admitted to the emergency department with long-lasting palpitations preceded by abdominal pain, vomiting, and diarrhea. At presentation, the patient was afebrile and hemodynamically stable, with an unremarkable physical examination. The electrocardiogram (ECG) showed sinus rhythm without ST-segment deviation. There was no alteration in blood test results despite a slightly elevated C-reactive protein. In serial determination of markers of myocardial damage, there was an elevation of high-sensitivity troponin I (5.28 ng/ml).

LEARNING OBJECTIVES

- To make a differential diagnosis of myocardial damage in young adults with multimodality imaging.
- To relate coronary anomalies with other CHD.
- To understand the importance of combining clinical presentation, past medical history, and clinical impact of the findings to select the best approach when high-quality published reports are lacking.

PAST MEDICAL HISTORY

The patient had aortic coarctation partially corrected during childhood with a bypass between the left subclavian artery and the descending thoracic aorta. He underwent reoperation with a Hemashield (Maquet, Rastatt, Germany) aortic graft implantation at 24 years of age. Follow-up cardiac magnetic resonance confirmed a good result of the aortic graft but significant stenoses at the proximal and distal

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anastomosis of the subclavian-to-aortic bypass (**Figures 1A and 1B**). He was previously diagnosed with type 1 bicuspid aortic valve with normal function (Video 1). He had no known cardiovascular risk factors.

DIFFERENTIAL DIAGNOSIS

In this scenario, different entities must be considered in the presence of elevated troponin, such as myocarditis or acute myocardial infarction type 1 or type 2.

INVESTIGATIONS

First, a transthoracic echocardiogram demonstrated normal left ventricular ejection fraction with no wall motion abnormalities (Videos 2, 3, and 4), bicuspid valve normal function (Video 5), and absence of recoarctation (Video 6, **Figure 2**). Because of his previous symptoms of viral infection, cardiac magnetic resonance was requested to rule out myocarditis, and the imaging showed not only normal left ventricular systolic function but also no edema (Video 7) or fibrosis (Video 8). To complete the differential diagnosis, cardiac computed tomography angiography was performed. This technique demonstrated the presence of left circumflex artery origin from the pulmonary artery (LCxPA) (Video 9, **Figures 3A and 3B**) without significant stenosis (**Figure 3C**). Small aortobronchial fistulous communications (**Figure 3D**) were noted as remnants of aortic coarctation. Finally, invasive cardiac catheterization confirmed the diagnosis by showing that the left circumflex artery had good retrograde filling through collateral vessels from the left anterior descending and right coronary arteries (Videos 10 and 11).

MANAGEMENT

During hospitalization, no arrhythmias were detected in continuous ECG monitoring. In the absence of chest pain or new clinical events, the patient was considered at low risk for sudden cardiac death (SCD). He was conservatively managed and discharged to be closely followed.

FOLLOW-UP

The patient has been followed up for 1 year. A recent treadmill test with single-photon emission computed tomography with sestamibi was informed as clinical and ECG negative but showed mild basal inferolateral ischemia on perfusion imaging (**Figures 4A and 4B**). Given the absence of symptoms and the limited perfusion defect, expectant management was maintained.

DISCUSSION

The clinical implications of LCxPA are not well established. Garcia et al. (1) first described it in adults in 1992, in a patient with no other congenital heart disease (CHD) who reported dyspnea and chest pain at rest. Recently, few additional cases have been described in adults: 2 presented as SCD, 1 with double-outlet right ventricle (2), and 1 without CHD (3), and another patient presented with mild exertional chest pain, wall motion abnormalities noted on echocardiography, and aortic coarctation (4). In contrast, in the larger experience with LCxPA among pediatric patients (5–7), the presentation was mostly exertional chest pain and dyspnea, and the association with other CHDs was higher. Two main questions arise from the scarce data described in published reports. First, given the higher life expectancy of patients with CHD nowadays, coronary anomalies, and this anomaly in particular, should be considered in the differential diagnosis of exertional symptoms or SCD. In this regard, noninvasive imaging techniques comprise the first diagnostic approach, requiring also invasive angiography in some cases, which enables physicians to: 1) assess adequate retrograde flow from collateral vessels; and 2) have a basal angiogram for comparison in the middle to long term in case the patient presents with extensive myocardial ischemia. In contrast, there are no consensus and guidelines defining the management of LCxPA. Although the latest guidelines (8) recommend surgery for asymptomatic anomalous left coronary artery from the pulmonary artery, they do not specify guidance on variants such as LCxPA; therefore, the only evidence is based on case reports. In these cases, adult patients presented with resting angina, dyspnea, New York Heart Association functional class III or IV, or aborted cardiac death in shockable rhythm were surgically treated. Only 1 reported patient (9) had angina and no pathological findings on ECG or echocardiography but was surgically treated. However, most of the adults described in published reports have had no previous heart operations (3,5–7,9,10). Besides, survival depends on collateral development (5,9),

ABBREVIATIONS AND ACRONYMS

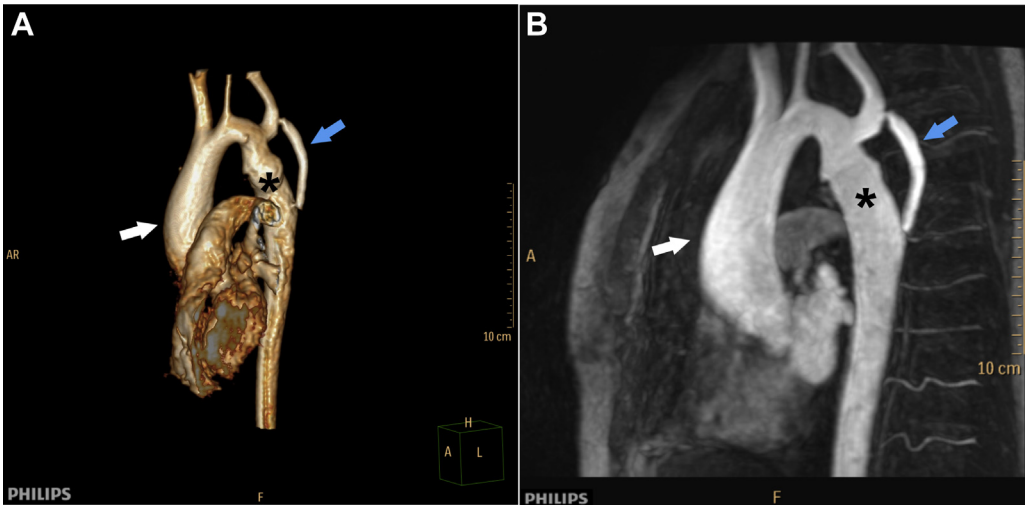
CHD = congenital heart disease

ECG = electrocardiogram

LCxPA = left circumflex from the pulmonary artery

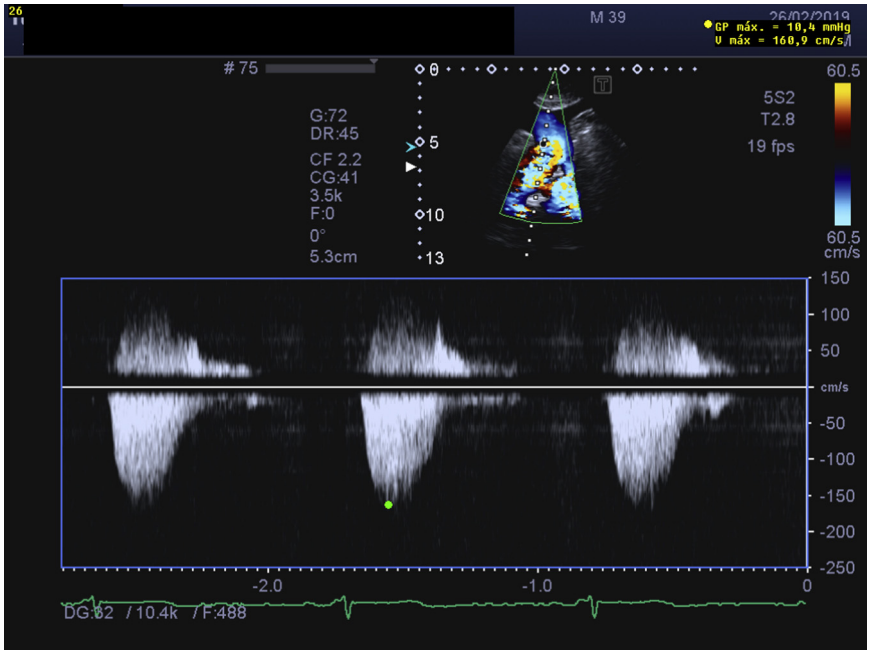
SCD = sudden cardiac death

FIGURE 1 Magnetic Resonance Angiography



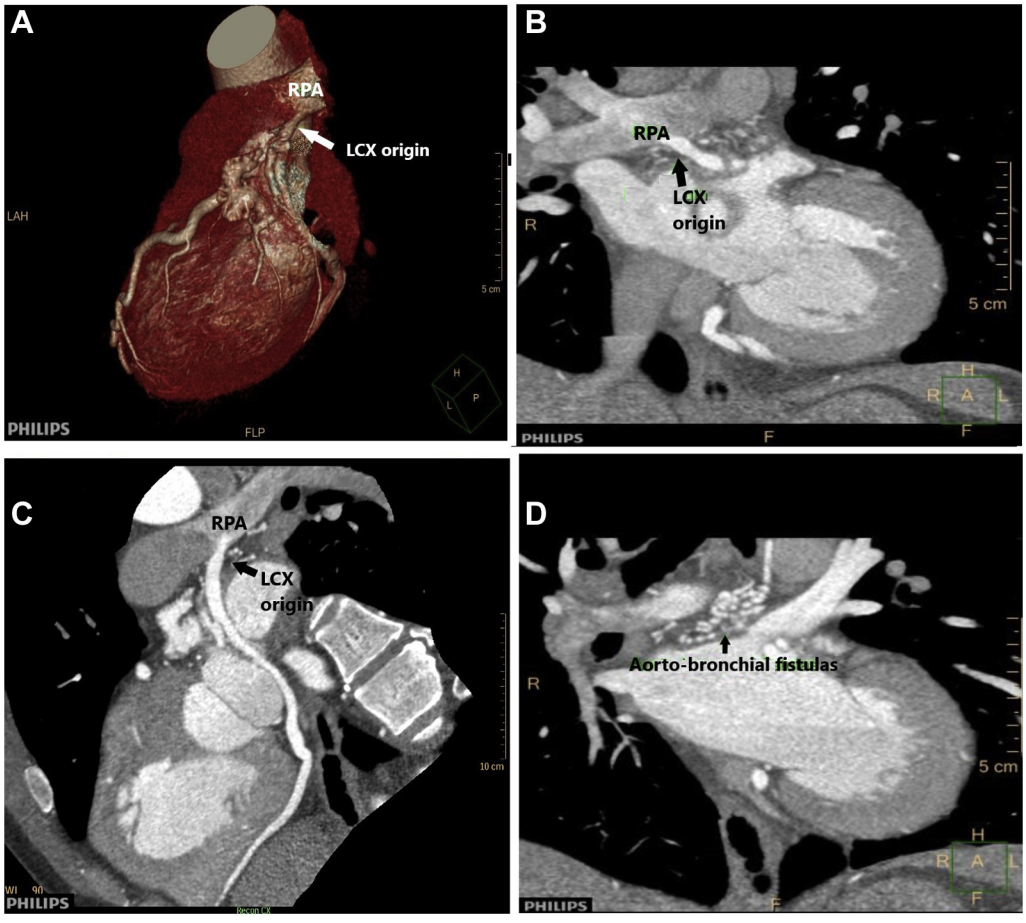
(A) Volume-rendering technique. **(B)** 2-dimensional magnetic resonance angiography. Mild dilatation of ascending aorta **(white arrows),** subclavian-descending aorta bypass (**blue arrows)** with stenosis at the proximal and distal anastomoses. Permeable Hemashield (Maquet, Rastatt, Germany) graft **(asterisks)**.

FIGURE 2 Doppler Echocardiography of the Descending Aorta



Peak systolic velocity is 161 cm/s, and peak systolic gradient is 10 mm Hg.

FIGURE 3 Coronary Computed Tomography Angiography Reconstruction Images



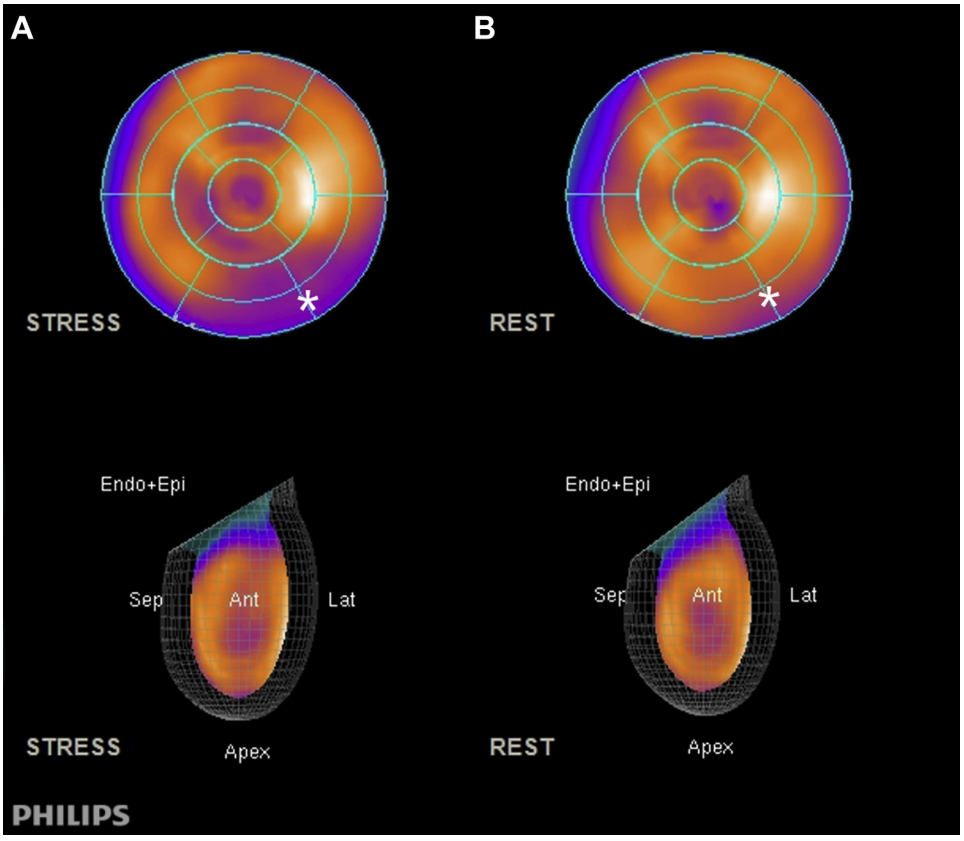
(A and B) Left circumflex (LCX) artery origin from the right pulmonary artery (RPA). **(C)** Curved multiplanar image of the left circumflex artery. **(D)** Coronal plane of aortobronchial fistulas.

and surgery is recommended when symptoms are attributed to ischemia (10). Conversely, our patient reported palpitations with no chest pain on admission, and although findings of single-photon emission computed tomography with sestamibi were slightly positive for ischemia, the treadmill test result was clinically negative at high load, and he remained free from angina during follow-up. This patient had considerable collateralization, 2 previous thoracic surgical procedures, and symptoms that could be attributed to supraventricular tachycardia triggered by relative hypovolemia secondary to gastroenteritis. After careful consideration, we chose conservative management and close follow-up. Certainly, we cannot ensure completely the appropriateness of our management. Nonetheless, we present the case of a patient who, more than a year after the diagnosis, has no symptoms or cardiovascular rehospitalizations. This outcome supports medical treatment as an option in selected scenarios.

CONCLUSIONS

LCxPA is a rare coronary anomaly, but it has been described in association with CHD, especially aortic coarctation. Awareness of this entity will enable early

FIGURE 4 SPECT-MIBI Stress-Testing Bullseye



(A) Peak of stress. Tracer hypocaptation in basal-inferolateral wall resulting from exercise-induced ischemia **(asterisk). (B)** At rest, perfusion tracer uptake increases **(asterisk).** Ant = anterior; Endo = endocardium; Epi = epicardium; Lat = lateral; Sep = septal; SPECT-MIBI = single-photon emission computed tomography sestamibi.

diagnosis and provide potential prognostic benefit. Furthermore, there is a gap of evidence regarding the best treatment of this anomaly. Although cardiac surgery has been extensively used, our case shows that conservative management may be an option in some circumstances.

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REFERENCES

1. Garcia CM, Chandler J, Russel R. Anomalous left circumflex coronary artery from the right pulmonary artery: first adult case report. Am Heart J 1992;123:526–8.

2. Harky A, Bashir M, Garner M, Hsia T. Anomalous origin of the circumflex coronary artery presenting with ventricular fibrillation cardiac arrest. BMJ Case Rep 2017;2017:bcr2016219184.

3. Liu B, Fursevich D, O'Dell MC, Flores M, Feranec N. Anomalous left circumflex coronary artery arising from the right pulmonary artery: a rare cause of aborted sudden cardiac death case presentation. Cureus 2016;8:e499.

4. Bolognesi R, Alfieri O, Tsialtas D, Manca C. Surgical treatment of the left circumflex coronary artery from the pulmonary artery in an adult patient. Ann Thorac Surg 2003;75:1642–3.

5. Sarioglu T, Kinoglu B, Saltik L, Eroglu A. Anomalous origin of circumflex coronary artery from the right pulmonary artery associated with subaortic stenosis and coarctation of the aorta. Eur J Cardiothorac Surg 1997;12:663–5.

6. Sekelyk R, Mykychak Y, Fedevych O, Yemets I. Anomalous origin of circumflex coronary artery from right pulmonary artery associated with coarctation of the aorta: a case report of surgical treatment. World J Pediatr Congenit Heart Surg 2014;5:97–9.

7. Al-muhaya MA, Syed A, Najjar AHA, Mofeed M, Al-mutairi M. Anomalous origin of circumflex coronary artery from right pulmonary artery associated with atrial septal defect. J Saudi Heart Assoc 2017;29:219–22.

8. Stout KK, Daniels CJ, Aboulhosn JA, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. J Am Coll Cardiol 2019;73:e81–192.

9. Danov V, Knornovski V, Hazarbasanov D, Panayotov P. anomalous origin of left circumflex coronary artery from the right pulmonary artery in adult. Thorac Cardiovasc Surg 2009;57:110–8.

10. Levin RL, Degrange MA, Salvagio F, Blanco N, Botbol A, Porcile R. Origen pulmonar anómalo de la arteria circunfleja en un paciente adulto. Rev Argent Cardiol 2009;77:524–6.

KEY WORDS ALCAPA, anomalous circumflex artery, coronary anomaly sudden cardiac death

APPENDIX For supplemental videos, please see the online version of this paper.