JACC: CASE REPORTS VOL. 29, 2024


VASCULAR DISEASE

CASE REPORT: CLINICAL CASE

# Left Anterior Descending Coronary Artery Mimic

## A Case of Coronary Artery to Pulmonary Artery Fistula

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**ABSTRACT**

Coronary artery fistulas (CAFs) are rare coronary anomalies involving the communication of an epicardial coronary artery and another cardiovascular structure. CAFs are usually easily distinguished from nearby coronary arteries. Here, we report a unique case of CAF that mimics the size, branching pattern, and appearance of a native epicardial left anterior descending artery. (J Am Coll Cardiol Case Rep 2024;29:102187) © 2024 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 49-year-old man with a history of obesity, diabetes mellites, hypertension, and dyslipidemia presented to the hospital with acute chest discomfort. He denied shortness of breath, nausea, vomiting, or diaphoresis. Vital signs demonstrated a temperature of 98 °F, blood pressure of 147/91 mm Hg, pulse of 68 beats/min, respiratory rate of 20 breaths/min, and oxygen saturation of 98% on ambient air. Cardiac examination revealed precordial chest wall tenderness; a regular rate; and rhythm without murmur, rub, or gallop.

**LEARNING OBJECTIVES**

- To recognize CAFs as a rare contributing factor to premature MI.
- To recognize the rarity of this clinical scenario as a unique case of CAF mimicking the appearance of the LAD.

### PAST MEDICAL HISTORY

The patient's cardiovascular history was notable for years of atypical chest pain. Prior echocardiograms and myocardial perfusion imaging studies were unremarkable. Coronary angiography had not been performed at that time.

### DIFFERENTIAL DIAGNOSIS

The patient's risk factors and clinical presentation are most concerning for acute coronary syndrome. Other etiologies include pericarditis/myopericarditis; hypertensive crisis; valvular disease; vasospastic angina; spontaneous coronary artery dissection; pulmonary embolism; and congenital coronary anomalies, including anomalous origin of a coronary artery or coronary artery fistula (CAF).

### INVESTIGATIONS

An electrocardiogram showed normal sinus rhythm with 1-mm ST-segment horizontal depressions in

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

Manuscript received August 27, 2023; revised manuscript received December 4, 2023, accepted December 6, 2023.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2023.102187

**ABBREVIATIONS AND ACRONYMS**

**CAF** = coronary artery fistula

**CTA** = computed tomography angiography

**LAD** = anterior descending artery

**MI** = myocardial infarction

**PA** = pulmonary artery

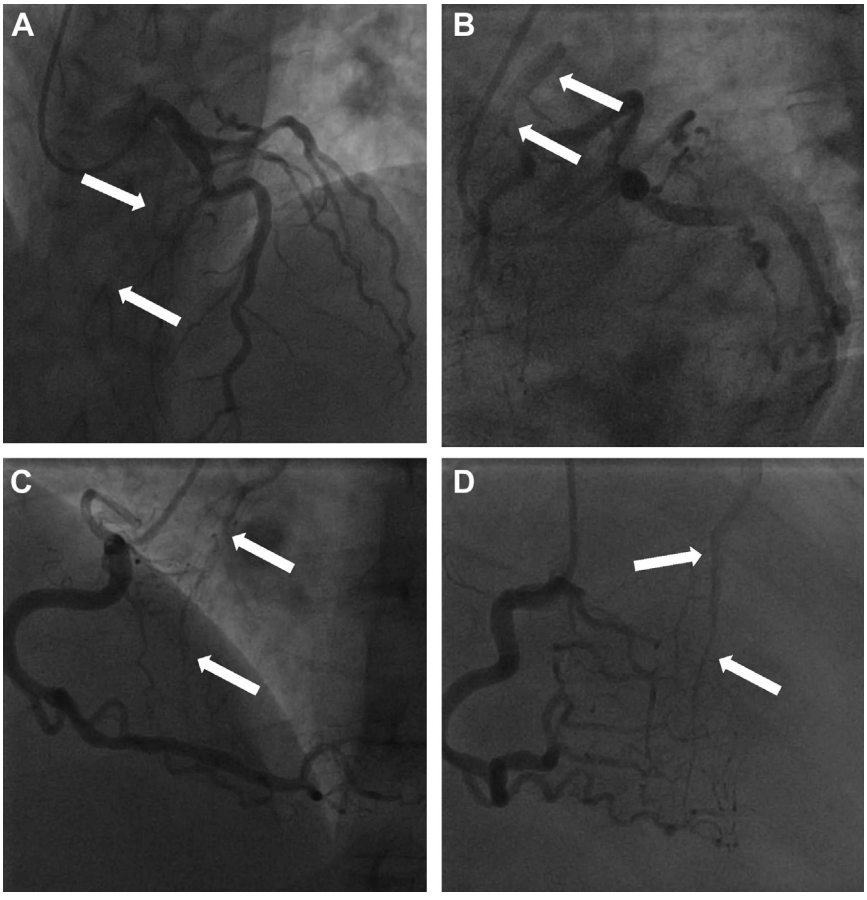
**VSD** = ventricular septal defect

leads $V_4$ to $V_6$. High-sensitivity troponin I was 6,702 ng/L on presentation and peaked at 16,295 ng/L (reference range: 3-57 ng/L). Basic metabolic panel, coagulation study, hepatic function panel, and complete blood count findings were unremarkable. Echocardiogram revealed normal biventricular size and systolic function with subtle hypokinesis of the basal and midanterolateral walls. Left ventricular ejection fraction was 55%. No significant stenosis or regurgitation was seen with any of the cardiac valves. Valve structures and Doppler flows were normal.

The patient was taken for urgent right and left heart catheterization with coronary angiography. Right heart catheterization revealed normal filling pressures with normal cardiac output. Coronary angiography was notable for diffuse moderate epicardial coronary atherosclerosis with high-grade small vessel and branch vessel disease; occlusion of the distal left anterior descending artery (LAD); and a coronary-to-pulmonary fistula, appearing to originate at the level of the mid-LAD (**Figures 1A and 1B**, Video 1) with additional distal connections to a large right ventricular marginal branch (**Figures 1C and 1D**, Video 2).

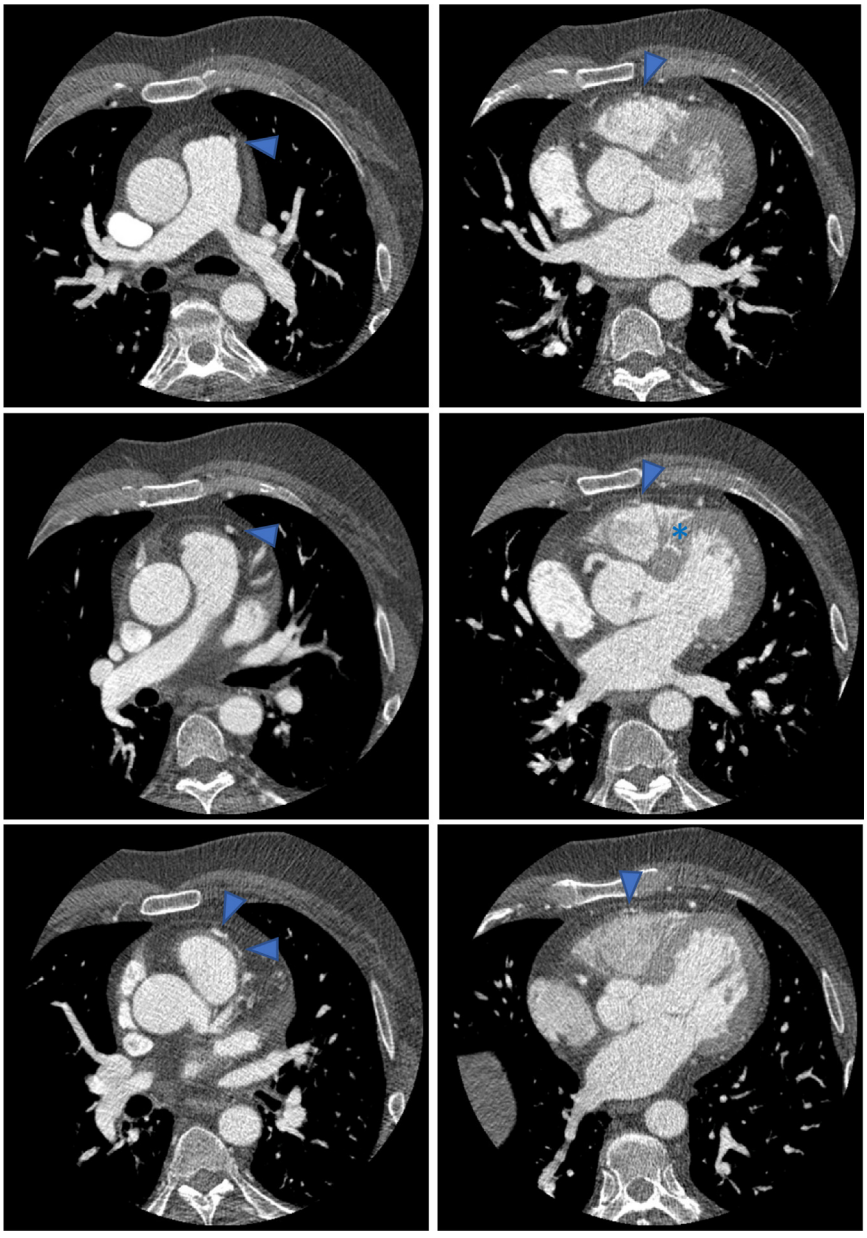
Subsequent computed tomography angiography (CTA) (**Figure 2**) was confirmatory, revealing a network of small superficial vessels along the anterior right ventricle that coalesced into a single large vessel resembling the LAD draining into the main pulmonary artery (PA). A 3-dimensional volume rendering of the computed tomography images was performed (**Figure 3**). A small muscular

**FIGURE 1 Coronary Angiography**




(A) LAO cranial projection showing faint visualization of fistula overlapping with a heavily diseased LAD (arrows). (B) LAO caudal projection showing the coronary artery fistula near the interventricular groove (arrows). (C) LAO and (D) RAO projections of the right coronary artery showing moderate to severe diffuse posterolateral branch disease and coronary artery–to-pulmonary artery fistula (arrows) involving distal RV marginal branches. LAD = left anterior descending artery; LAO = left anterior oblique; RAO = right anterior oblique; RV = right ventricular.

**FIGURE 2** **Representative Transaxial CTA Images Demonstrating Coronary Artery-to-Pulmonary Artery Fistula**



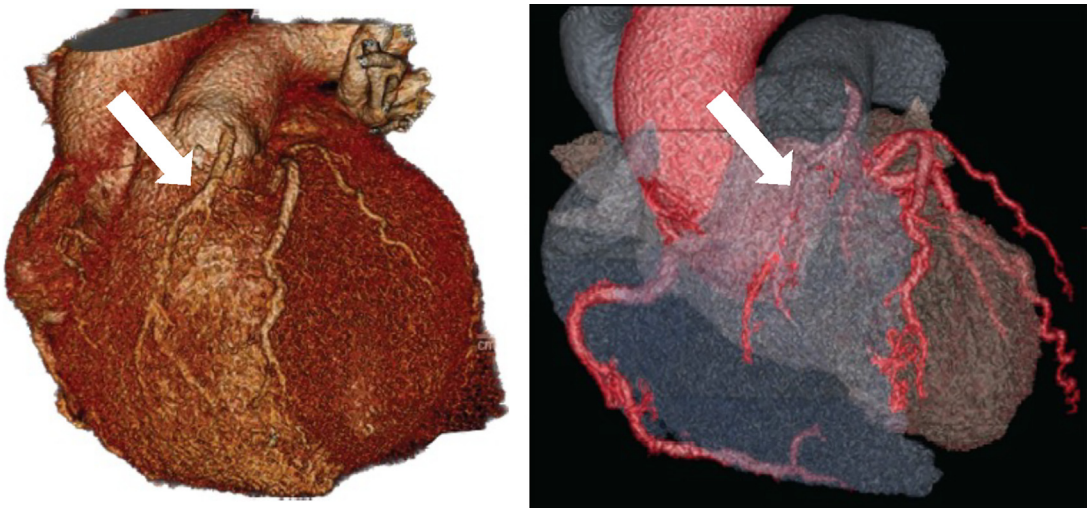
The communication (arrowheads) arises between the distal LAD and a network of small superficial anterior RV branches that coalesce to form a large vessel similar in caliber to the proximal LAD that drains into the supravalvular main pulmonary artery. The patient's VSD is also visible (star). CTA = computed tomography angiography; VSD = ventricular septal defect; other abbreviations as in Figure 1.

ventricular septal defect was incidentally noted (Figure 4).

## MANAGEMENT

The patient was discharged on optimal medical therapy, including dual-antiplatelet therapy and a high-intensity statin. The patient did not meet criteria for transcatheter or surgical intervention of his fistula or ventricular septal defect (VSD), because the fistula and VSD were not initially thought to be the primary etiologies of the patient's ischemic symptoms and myocardial infarction (MI), and preliminary management focused on patient stabilization and reperfusion. The VSD was not believed to be hemodynamically significant because of its small size.

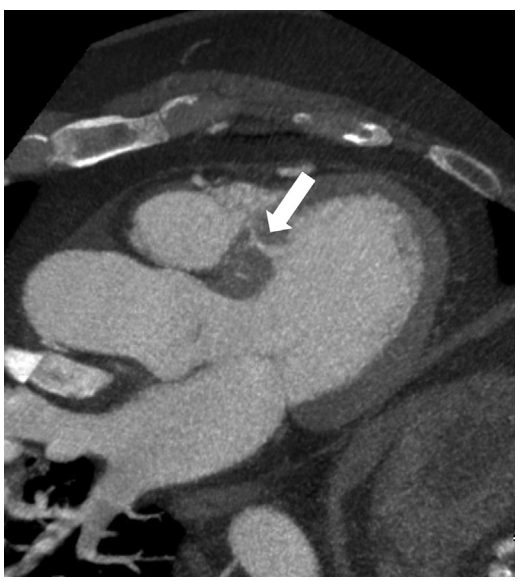
**FIGURE 3 Reconstructed Images of the Heart and Coronary Vessels**



A 3D volume-rendered CT (left) and 3D volume-rendered CT with transparent right ventricle (right) demonstrating CAF (arrows) mimicking the LAD size, shape, and branching pattern. CAF = coronary artery fistula; CT = computed tomography; LAD = left anterior descending artery.

Suspicion of the patient's fistula contributing as a secondary risk factor to the ischemic event remained, and so outpatient cardiac magnetic resonance imaging was ordered to evaluate his VSD hemodynamics and fistula location but has not yet been obtained.

**FIGURE 4 CTA Showing a Small Muscular Ventricular Septal Defect Located in the Proximal Anteroseptal Segment**



Defect in muscular ventricular septum allowing blood flow between right and left ventricles (arrow). CTA = computed tomography angiography.

## DISCUSSION

Coronary anomalies are a heterogenous group of disorders with variable clinical relevance. CAFs are either congenital or acquired and are rare, representing only 0.13% of all congenital heart defects.[1] Although these lesions are typically benign and incidentally discovered during coronary angiography or CTA, they can present with various clinical syndromes, including myocardial ischemia and congestive heart failure, secondary valve disease, and endocarditis.[2-4] CAFs can arise from either the right coronary artery, left coronary artery, or conus branch (infundibular artery) and communicate with either a cardiac chamber ("cameral fistula") or a pulmonary artery, pulmonary vein, or superior vena cava ("coronary arteriovenous fistula").[2] The Sakakibara CAF classification system categorizes fistulas as arising from the proximal or distal coronary arteries.[5] When the fistula originates within the proximal one-third of the coronary artery, the proximal feeding arteries may dilate while the diameters of the coronary arteries distal to the CAF remain normal. In contrast, the entire vessel is often dilated when the feeding arteries originate distally. In the present case, the fistulous tract arises from the distal LAD, and the entire vessel is diffusely dilated. A curious feature is the single large draining vessel terminating at the main pulmonary artery, itself comparable in size and branching morphology to the dilated LAD. Although there are existing cases of medium and large LAD

fistulas in the literature,[6] to our knowledge, ours is the first to report a fistula that closely mimics the LAD. Given its resemblance to the native LAD, it is important to distinguish this case as a fistulous tract rather than another coronary artery anomaly, such as a dual LAD or anomalous left coronary artery from the pulmonary artery.

Although the initial etiology for this patient's MI was thought to be a type I MI, his coronary angiogram revealing chronic epicardial coronary artery disease without a clear culprit makes his LAD-PA fistula a possible source. Multimodality imaging is of paramount importance to assess the anatomic and hemodynamic implications of congenital coronary anomalies. Historically, conventional angiography with right heart catheterization was the standard for evaluation and treatment of CAFs because it yields anatomic and hemodynamic information and facilitates the diagnosis and therapeutic embolization of CAFs. However, the dynamic 2-dimensional projection images can hinder delineation of complex abnormal fistulous tracts.[1] Electrocardiogram-gated CTA is now regarded as the gold standard imaging tool for known or suspected congenital or developmental coronary artery abnormalities because of its 3-dimensional volumetric display; its high temporal and spatial resolution; and its rapid, noninvasive acquisition.[1] Both the limitations in coronary angiography and benefits of advanced imaging with computed tomography are highlighted in our case.

Management of CAFs is dependent on patient symptoms and anatomic considerations. Small CAFs are typically benign, will not cause symptoms, and can be monitored with imaging every 3 to 5 years.[3] Conversely, intervention may be considered in cases of symptomatic medium-sized (1-2× the vessel diameter) or large-sized (>2× the vessel diameter) fistulas.[3] The technical aspects of transcatheter closure depend on whether the CAF is simple or complex in anatomy and whether or not the origin is proximal or distal. Current American College of Cardiology/American Heart Association guidelines provide a Class 1 recommendation for treatment of large CAFs regardless of symptoms or in cases of small- to moderate-sized fistulas with symptoms including myocardial ischemia, endarteritis, arrhythmia, and ventricular dysfunction.[7]

## FOLLOW-UP

The patient was asymptomatic on outpatient follow-up 1 month later. Optimal medical therapy including dual-antiplatelet therapy and statin therapy was continued. Exercise stress testing was inconclusive because of submaximal heart rate. The patient was referred for cardiac rehabilitation and reported improved stamina and endurance upon completion. He was scheduled for cardiac magnetic resonance with flow mapping for monitoring, assessment of shunt severity, and potential intervention but was lost to follow-up.

## CONCLUSIONS

CAFs are rare coronary anomalies that can result in myocardial ischemia and congestive heart failure. Transcatheter intervention is indicated in patients with symptomatic moderate or large CAFs.

## FUNDING SUPPORT AND AUTHOR DISCLOSURES

The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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**KEY WORDS** computed tomography, congenital heart defect, coronary angiography, coronary vessel anomaly, imaging, ventricular septal defect

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