

CASE REPORT

INTERMEDIATE

CLINICAL CASE

Coronary Artery and Pulmonary Artery Fistula

Rare Congenital Coronary Artery Fistula



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ABSTRACT

Coronary artery fistula is a rare condition that can be congenital or acquired. Most patients with these anomalies remain asymptomatic, but symptoms and complications may develop with increasing age. We discuss the clinical presentation, diagnostic work-up, and management options for this condition. **(Level of Difficulty: Intermediate.)** (J Am Coll Cardiol Case Rep 2020;2:286-8) © 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 57-year-old female patient presented to the emergency department with left-sided chest pain, which was described as intermittent pressure-like pain, associated with numbness of the left arm. The patient did not describe any aggravating or relieving factors. She denied shortness of breath or palpitations. She also denied having fever, chills, dyspnea, or recent illness. On admission, her blood pressure was 124/62 mm Hg, her heart rate was 68 beats/min, her respiratory rate was 16 breaths/min, and her

temperature was 98.2°F. Her chest auscultation findings and heart sounds were normal, with no murmurs audible on auscultation. Jugular venous pressure was not raised, and there were no peripheral signs of heart failure.

From her past medical history, she was known to have systemic lupus erythematosus treated with long-term steroid therapy. She also had hypertension and gastroesophageal reflux disease, and she was a current smoker. The patient was admitted for further work-up.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis included cardiac chest pain that could represent acute coronary syndrome or, less likely, myocarditis and pericarditis. The differential diagnosis of her chest pain also included noncardiac chest pain, such as musculoskeletal chest pain or esophageal pain.

INVESTIGATIONS

Blood test results on admission are presented in [Table 1](#).

LEARNING OBJECTIVES

- The clinician will be able to understand different presentations of coronary artery fistula.
- The importance of a conservative approach in the management of coronary artery fistula in light of the 2018 American Heart Association and American College of Cardiology guidelines for the management of adults with congenital heart disease will be appreciated.

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Informed consent was obtained for this case.

Manuscript received April 8, 2019; revised manuscript received October 7, 2019, accepted November 4, 2019.

TABLE 1 Blood Test Results

Blood Tests	Result	Normal Values
White blood cells	5,500/ μ l	3,600-10,800/ μ l
Hemoglobin	11.2 g/dl	12-16 g/dl
Platelets	377,000/ μ l	150,000-400,000/ μ l
Creatinine kinase	333 U/l	21-215 U/l
Creatinine kinase-MB	3.3 ng/ml	05-3.6 ng/ml
Troponin I	<0.015 ng/ml	<0.054 ng/ml
Troponin I after 6 h	<0.015 ng/ml	<0.045 ng/ml
B-type natriuretic peptide	32 pg/ml	<100 pg/ml

MANAGEMENT

The patient continued to have chest pain despite normal troponin levels. Echocardiography showed normal findings, including contractility and chamber size (Table 2). Taking into consideration her cardiac risk factors, we decided to proceed to a nuclear stress test to exclude ischemic heart disease. The test revealed an inferior wall defect, along with normal left systolic function. The next step was to perform a computed tomography (CT) coronary angiogram to exclude significant coronary artery stenosis. CT angiography revealed a possible fistula from the left anterior descending coronary artery to the pulmonary trunk (Figure 1). Chest pain then improved significantly. The patient was discussed in a multidisciplinary meeting, and it was decided to proceed to a coronary angiogram to confirm this congenital abnormality. Coronary angiography demonstrated normal coronary arteries as well as a coronary artery fistula measuring 2 to 3 mm in diameter from the midportion of the left anterior descending artery, immediately after the first diagonal branch, to the main pulmonary artery above the pulmonic valve (Figure 2, Videos 1 and 2). The fistula was thought to be too small to explain any symptoms, and there was no evidence of left-to-right shunt. Therefore, no intervention was deemed necessary. The pain was attributed to gastroesophageal reflux disease, and she was advised to follow up with the gastroenterology clinic.

TABLE 2 Echocardiographic Measurements

Features	Measurements	Normal Value
LV diastolic diameter	35.7 mm	30.9-50.3 mm
LV systolic diameter	23.1 mm	20.1-40 mm
IVS diastolic thickness	10.7 mm	6-12 mm
RV diastolic diameter	29 mm	10.9-30.8 mm
Ejection fraction	65%	—
Aortic root diameter	30 mm	—
LA volume index	13.2 ml/m ²	—

IVS = interventricular septum; LA = left atrial; LV = left ventricular; RV = right ventricular.

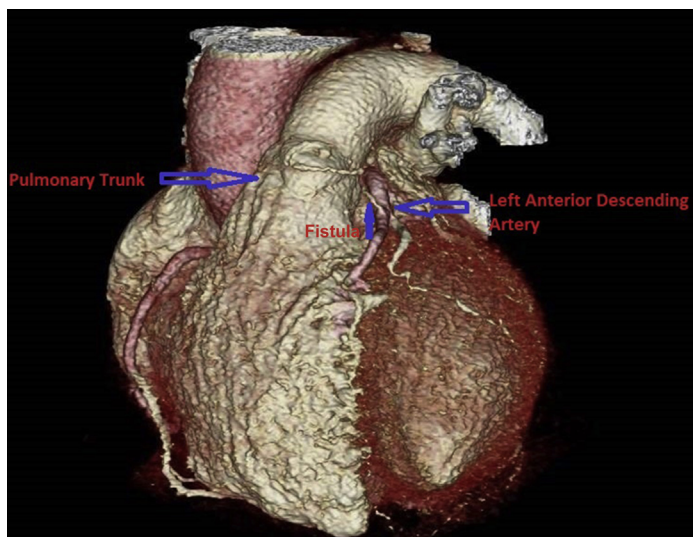
DISCUSSION

Coronary artery fistulae are congenital or acquired connections between the coronary vessels and the cardiac chambers or other vascular structures, such as the vena cava, the pulmonary artery, or pulmonary veins. The incidence of coronary artery fistula is 0.1% to 0.2% in all patients undergoing coronary angiography (1). These fistulae may arise from any of the major 3 coronary arteries; however, in most cases coronary artery fistula arises from the right coronary artery (2). In our patient, the fistula connected the left descending coronary artery with the pulmonary trunk. Although patients with coronary artery fistulae remain asymptomatic most of the time, the physiological derangement depends principally on the resistance of the fistulous connection and on the site where the fistula terminates. The resistance is determined by the size, tortuosity, and length of the pathway. These fistulae tend to enlarge with age. In addition, blood follows the lower-resistance pathway through the fistula, rather than traversing the smaller arterioles and capillaries of the myocardium. With larger fistulae, diastolic runoff may occur, drawing blood away from the normal coronary pathway with widened pulse pressure and coronary steal. All of this could lead to symptoms and complications, including myocardial ischemia, myocardial infarction, congestive heart failure, or sudden death (3). In general, the

ABBREVIATIONS AND ACRONYMS

- CT = computed tomography
- IVS = interventricular septum
- LA = left atrial
- LV = left ventricular
- RV = right ventricular

FIGURE 1 Coronary Artery Fistula



Computed tomography coronary angiogram shows a fistula connection between the main pulmonary trunk and the left anterior descending coronary artery.

FIGURE 2 Coronary Artery Fistula



Coronary angiography reveals the fistula between the midportion of the left anterior descending coronary artery and the main pulmonary trunk with normal coronary arteries. See Videos 1 and 2.

symptoms depend on the size of the fistula. They also depend on where the coronary artery fistulae drain. If they drain into the right-sided chambers or systemic veins, the presentation is with a left-to-right shunt, whereas when the fistula drains into the left-sided chambers, the presentation is with a left-sided volume overload. Although the nuclear stress test in our patient showed a defect in the inferior wall, the catheter angiographic study revealed normal coronary arteries. Rubini et al. (4) reported a case with a reversible myocardial perfusion defect, with nuclear

perfusion imaging associated with a coronary artery-to-pulmonary artery fistula in the anterior wall and apex. However, we do not believe that was the case in our patient because the nuclear study showed a fixed inferior wall defect that did not reflect the anatomic location of the fistula.

In the 2018 American Heart Association and American College of Cardiology guidelines for the management of adults with congenital heart disease (5), there was no consensus on management strategies for coronary artery fistulae. Balanescu et al. (6), in their review of published reports, proposed indications for closure of fistulae that included symptoms of heart failure and myocardial ischemia, and in asymptomatic patients with high-flow shunting, to prevent occurrence of symptoms or complications, especially in pediatric patients. Surgery and catheter techniques can be used to close the fistulous connection. In patients treated with surgical fistula ligation, post-operative myocardial infarction occurred in 11% because of low flow in the dilated coronary artery, proximal to the fistula closure. Late survival was also significantly reduced compared with an age-matched group (1).

CONCLUSIONS

Percutaneous closure is associated with lower procedural risk and has therefore become the preferred method of treatment. In our patient, after discussing the risk versus benefit of each intervention with the patient, the team decided to continue with the conservative approach. Patients treated conservatively should be followed up closely for the appearance of symptoms.

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KEY WORDS chest pain, coronary angiography, coronary vessel anomaly

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