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MINI-FOCUS ISSUE: CHEST WOUNDS

INTERMEDIATE

CASE REPORT: CLINICAL CASE

Multiple Coronary Fistulas After Several Penetrating Chest Wounds

A Rare Case of Refractory Heart Failure

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ABSTRACT

Coronary artery fistulas are rare coronary abnormalities. Most of these fistulas have a congenital origin, and only a few are acquired. We report the case of a patient with late-acquired multiple coronary fistulas secondary to a stab wound, diagnosed in the setting of ischemic heart failure secondary to coronary steal syndrome. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:34–8) © 2021 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/).

A 68-year-old man presented with a history of worsening shortness of breath, exercise intolerance, and new onset angina. Physical examination showed bilateral inspiratory crackles on lung auscultation, lower pitting edema, and jugular venous distention. Heart sounds were arrhythmic with no murmurs or gallops. Multiple stab wound scars were noted in the thorax and upper abdomen, with a 5-cm scar in the epigastrium. Blood pressure was 100/75 mm Hg, pulse rate was 75 beats/min, respiratory rate was 14 breaths/min, and oxygen saturation was 95% at room air.

LEARNING OBJECTIVES

- To understand the importance of a systematic evaluation of each patient's clinical history and work-up to establish an accurate diagnosis of refractory heart failure and to provide appropriate treatment.
- To recognize the possible clinical presentations and manifestations of acquired coronary fistulas.

PAST MEDICAL HISTORY

One year before, in the outpatient clinic of another city, he received a diagnosis of heart failure (HF) with preserved ejection fraction (HFpEF) in the context of what was thought to be permanent atrial fibrillation (AF). Clinical records from previous hospitalizations were not available. The patient relayed that he was initially prescribed warfarin, metoprolol-succinate, and nonpharmacological interventions. However, after multiple hospitalizations for HF decompensation, he was administered digoxin and spironolactone and was referred to our institution (Fundación Cardioinfantil, Bogotá, Colombia) for further work-up and management. His past medical history included a chronic diaphragmatic hernia with subsequent rightward deviation of the mediastinal structures as a sequela of multiple thoracoabdominal stabs wounds

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he received 30 years earlier. He had no previous history of hypertension, diabetes, obesity, dyslipidemia, or sleep apnea. Despite adherence to pharmacotherapy, his symptoms were progressively worsening, and he was repeatedly hospitalized for HF decompensation.

DIFFERENTIAL DIAGNOSIS

He was referred to our institution with a presumptive diagnosis of HFpEF and AF. However, additional previous clinical records and studies were unavailable. The first time we saw him, signs and symptoms of volume overload and an arrhythmic pulse were present. In this setting, the initial differential diagnosis was directed toward acute decompensation secondary to an uncontrolled ventricular response. In addition, cardiac and vascular causes, such as acute coronary syndrome or pulmonary embolism, were considered. The finding that the patient was hospitalized for multiple decompensations in that year raised the suspicion of an undetected cause. Previous transthoracic echocardiograms performed outside our institution failed to identify a specific cause of his symptoms.

INVESTIGATIONS

An electrocardiogram showed AF with controlled ventricular response and no acute signs of ischemia. High-sensitivity troponin I concentrations were 0.03 ng/ml on presentation and 0.032 ng/ml 4 h later (normal <0.026 ng/ml). The N-terminal pro-B-type natriuretic peptide plasma level was 448 pg/ml (previously 530 pg/ml).

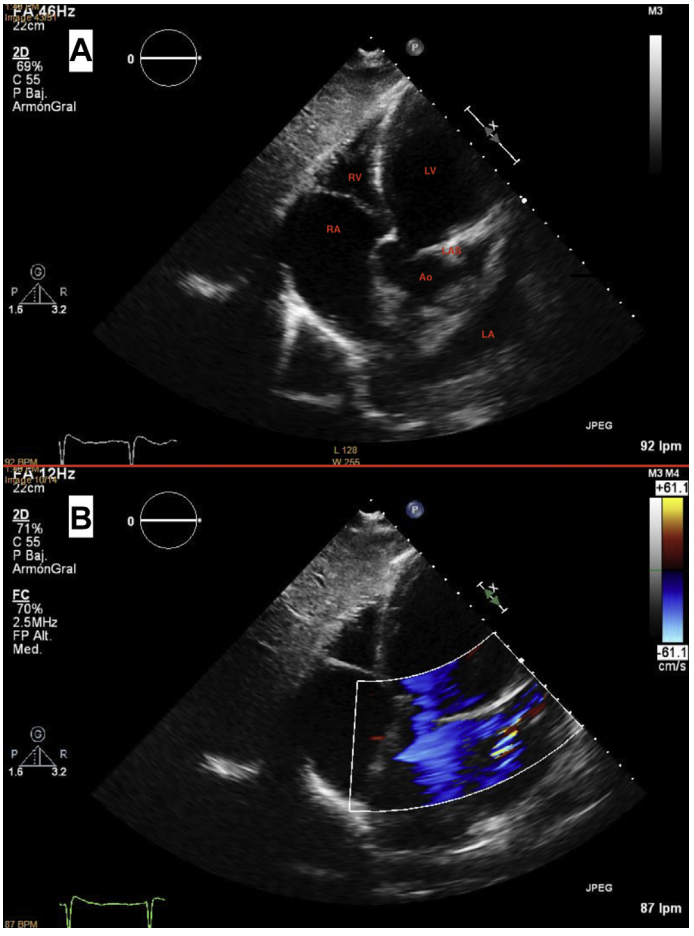
A transthoracic echocardiogram revealed left atrial and ventricular dilation, eccentric left ventricular hypertrophy, and a left ventricular ejection fraction of 55% (Figure 1A, Table 1). Color Doppler imaging showed a previously unnoticed turbulent flow from the left main coronary trunk into a significantly dilated left atrium, thus raising the suspicion of a coronary artery fistula (CAF) (Figure 1B, Video 1). Coronary angiography revealed a large fistula originating from the left main trunk to the left atrium with no anterograde flow to the left anterior descending artery (LAD) or circumflex artery (CxA). The right coronary artery had multiple collateral vessels that provided partial flow to the LAD and CxA territories, thereby preventing generalized ischemia of the left ventricle (Video 2). Right-sided cardiac catheterization showed a normal cardiac index and pulmonary-systemic flow ratio (Q_p/Q_s ratio) (Table 2).

Cardiac magnetic resonance (CMR) could not be performed because the patient had claustrophobia and insufficient heart rate control for high-quality image acquisitions. As an alternative, we performed computed tomography (CT) (Figures 2A, 2B, 3A, and 3B). Although functional characterization with CT is not reliable, it provided sufficient anatomic information for surgical planning. Three-dimensional cardiac scanning showed a markedly dilated left main trunk, a dilated and tortuous proximal portion of the first diagonal artery, and an additional fistula arising from the CxA draining into the left atrium. In addition, the first and second diagonal branches of the LAD and the obtuse marginal artery had a small caliber and were

ABBREVIATIONS AND ACRONYMS

AFib = atrial fibrillation
CAF = coronary artery fistulas
CMR = cardiac magnetic resonance
CT = computed tomography
CxA = circumflex artery
HF = heart failure
HFpEF = heart failure with preserved ejection fraction
LAD = left anterior descending artery

FIGURE 1 Transthoracic Echocardiography: Apical 5-Chamber View



(A) Left atrial and ventricular dilation and eccentric left ventricular hypertrophy. **(B)** Color Doppler image showing turbulent flow from the left main coronary trunk into a significant dilated left atrium. 2D = 2-dimensional; Ao = aorta; LA = left atrium; LAS = left aortic sinus; LV = left ventricle; RA = right atrium; RV = right ventricle.

TABLE 1 Echocardiographic Findings

LVEF, %	55
LV internal dimension in diastole, mm	60
LVEDVI, ml/m^2	58
LAVI, ml/m^2	39
Caval index	Minimal collapse (28%)
PASP, mm Hg	32
RWT	0.41
GLS	Not available (not systematically measured)

GLS = global longitudinal strain; LAVI = left atrial volume index; LV = left ventricle; LVEDI = left ventricular end-diastolic volume index; LVEF = left ventricular ejection fraction; RWT = relative wall thickness.

TABLE 2 Right-Sided Heart Catheterization

Cardiac output index, l/min/m^2	2.11
Q_P/Q_S	0.98
RA pressure, mm Hg	4
Mean PAP, mm Hg	10
PCWP, mm Hg	9
Mixed venous O_2S, %	67
RA O_2S, %	66.6
Pulmonary artery O_2S, %	63.6
LV O_2S, %	95.8

LV = left ventricle; O_2S = oxygen saturation; PAP = pulmonary artery pressure; PCWP = pulmonary capillary wedge pressure; Q_P/Q_S = pulmonary-systemic shunt ratio; RA = right atrium.

poorly opacified, suggesting no anterograde flow from the left main trunk. We considered performing myocardial perfusion imaging using single-photon emission CT to characterize the flow distribution of the left ventricle, depict ischemia, and assess tissue viability; however, we could not perform it because of the presence of AF and atrial flutter with a rapid ventricular rate.

MANAGEMENT

We considered that the cause of the patient's HF and progressive symptoms was ischemic heart disease secondary to coronary steal syndrome. Even though previous records were not available, and the patient did not provide a clear description of the traumatic event, we hypothesized that these fistulas appeared as a consequence of a coronary laceration secondary to the stab wounds he had sustained in the thoracoabdominal region.

The patient underwent surgery uneventfully. Surgical findings confirmed a dilated left main trunk, 1 fistula between the left main trunk and the left atrium, and 1 fistula between the CxA and the left atrium. We achieved successful repair of the coronary defects with ligation and revascularization of the LAD and CxA.

DISCUSSION

CAFs are anomalous connections between 1 or more of the coronary arteries and a cardiac chamber or great vessel (1). They comprise one-half of all coronary anomalies and have a prevalence of approximately 0.002% in the general population. Most fistulas have a congenital origin; however, a few are acquired, usually as a consequence of iatrogenic injury after cardiac surgery, endomyocardial biopsy, or coronary angioplasty (2), and less commonly as a result of trauma or infection or as a complication of Kawasaki disease. Complications appear later in life and include HF secondary to volume overload, myocardial ischemia secondary to coronary steal syndrome, arrhythmias, and, thrombosis.

CAFs remain a diagnostic and therapeutic challenge, mainly because their low prevalence decreases the clinician's level of suspicion. When the fistula drains into any of the left chambers, it causes volume overload without increasing pulmonary blood flow. In transthoracic echocardiography, this can be seen as dilation of the chambers involved or those located downstream. In the long term, this phenomenon can lead to congestive HF and rarely to myocardial ischemia secondary to severely decreased flow in the coronary circulation downstream of the fistula.

Few cases of CAFs in patients presenting after chest stab wounds have been reported, and most were diagnosed months to years after the initial injury. Trauma-related CAFs appear more often in the right coronary vessels because injury to the left coronary artery usually results in early death. Most of these fistulas drain into right-sided heart structures (i.e., right ventricle, right atrium, pulmonary artery), and only a few drain into the left chambers. To our knowledge, this is the first case of a CAF arising from the CxA and draining into the left atrium.

In our patient, symptoms of HF were attributed to coronary steal syndrome. Importantly, the confluence of late-acquired, multiple CAFs arising from the left main trunk and the CxA, as a consequence of traumatic injury to the coronary circulation, is a rare event. A previous history of multiple stab wounds in the chest and abdomen let us conclude a possible causal relationship between both events. However, the exact pathophysiological mechanism by which these fistulas appeared after the penetrating chest injury is not clear. Color Doppler echocardiography is an inexpensive initial approach, proving sufficient for the diagnosis of CAFs. Coronary angiography is the

gold standard for imaging the coronary arteries and was once the primary diagnostic technique for CAFs (1). Although it delineates the origin and course of CAFs, identification of distal drainage sites may be difficult given the contrast dilution in the usually lower-pressure chambers (3). Moreover, the utility of coronary angiography for surgical planning is limited because it poorly characterizes the heart's anatomy and the relationship of the fistula with the adjacent structures.

Under such conditions, noninvasive methods such as CMR and contrast-enhanced CT can complement coronary angiography and provide a better approach for surgical planning (4). CMR provides the best functional and anatomic characterization of the heart and adjacent structures. However, its relatively low availability and intrinsically demanding technical requirements decrease its widespread use. CT is a good alternative in this setting because its ability to detect CAFs has increased as a result of better sensitivity of volumetric data acquisition (5). Moreover, 3-dimensional rendered images provide an excellent overview of cardiac and vascular anatomy, which has proven to be useful for pre-operative planning (3).

The management strategy for CAFs needs to be individualized. In rare instances, the fistula may close spontaneously without further surgical interventions; however, in view of the morbidity of complications that can arise in untreated fistulas, closure may be justified even in asymptomatic patients (5).

FOLLOW-UP

The patient's symptoms significantly improved, and cardiac rehabilitation was initiated. He was prescribed anticoagulant therapy and a beta-blocker. He has not required further hospitalizations for acute decompensation.

CONCLUSIONS

The diagnosis of CAFs remains challenging because of its indolent course and nonspecific symptoms. The presence of progressive and unresponsive HF should raise the suspicion of an undetected and uncorrected cause.

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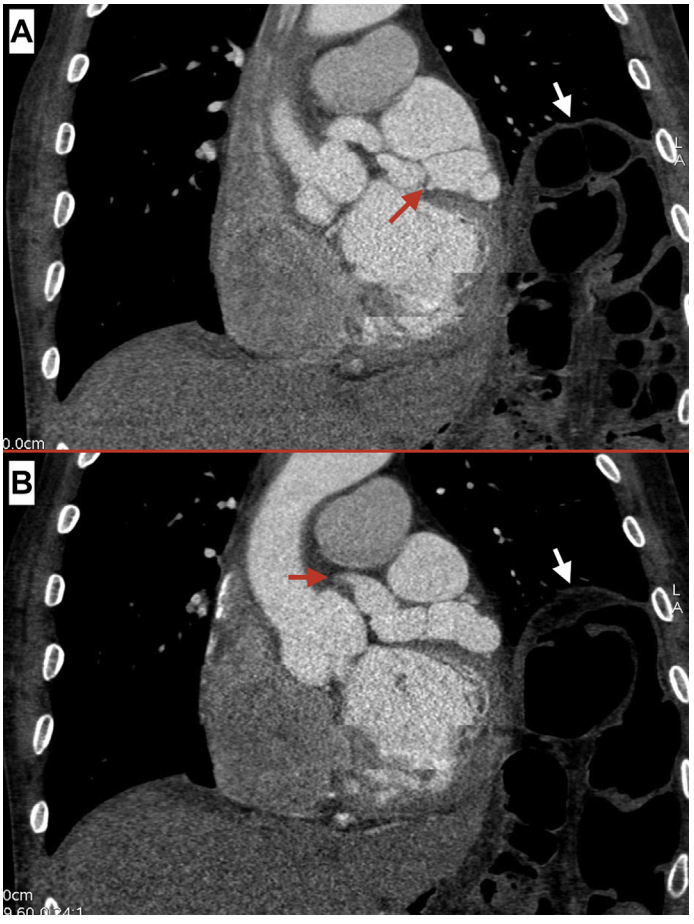
FIGURE 2 Computed Tomography: Coronal View



(A and B) Course of the coronary fistulas **(red arrow).** A huge diaphragmatic hernia is also shown **(white arrow).**

AUTHOR DISCLOSURES

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

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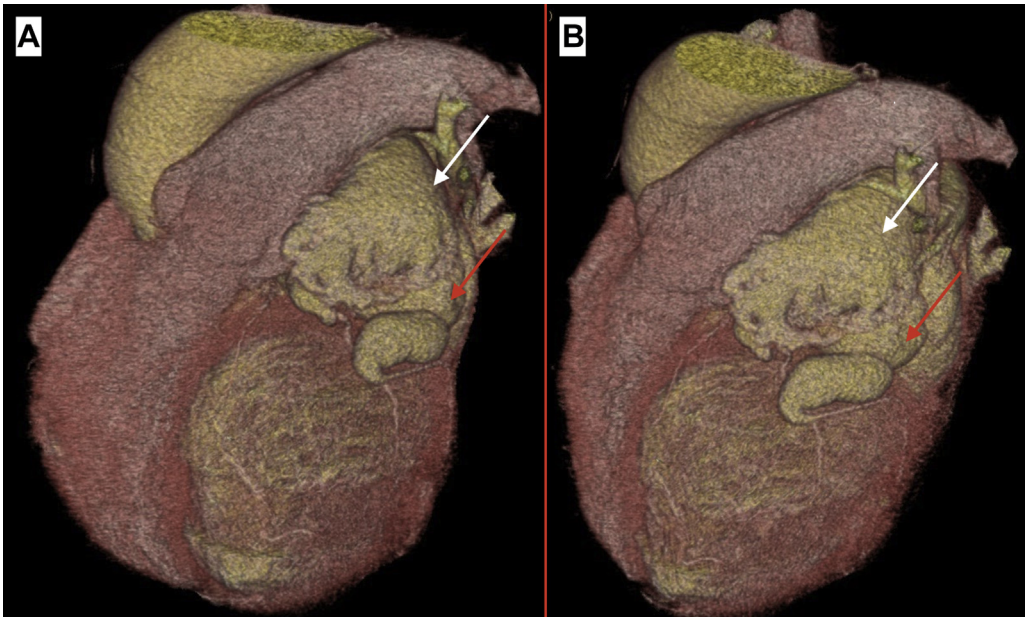
FIGURE 3 3-Dimensional Computed Tomography: Rendered Images



(A and B) Markedly dilated left main trunk and a dilated and tortuous proximal portion of the first diagonal artery. **Red arrow** = coronary fistula; **white arrow** = dilated left atrium.

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KEY WORDS arterioarterial fistulas, coronary artery fistulas, coronary steal syndrome, heart failure, stab wounds

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