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CASE REPORT

BEGINNER

CLINICAL CASE

Coronary Artery Fistula

An Unexpected Cause of Heart Failure in a 58-Year-Old Woman

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ABSTRACT

Coronary artery fistula (CAF) is an abnormal connection between a coronary artery and a heart chamber resulting in left-to-right shunt. A large CAF was an unexpected cause of heart failure in a 58-year-old woman who underwent transcatheter closure of the CAF with improvement in symptoms but complicated by myocardial infarction. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2020;2:1192–5) © 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 58-year-old woman, with a heart murmur since childhood, presented with palpitations and shortness of breath. She had a 10-year history of paroxysmal atrial fibrillation, as well as progressive dyspnea on exertion, reduced exercise tolerance, and lower extremity edema. Systemic anticoagulation was not tolerated secondary to Von Willebrand disease (VWd). Examination was notable for 3/6 continuous murmur along the left sternal border and trace bilateral pitting edema to the ankles.

LEARNING OBJECTIVES

- To describe the pathophysiology, intervention indications, and post-closure complications related to CAF.
- To consider undiagnosed congenital heart disease in the differential diagnosis of an adult patient presenting with heart failure.

PAST MEDICAL HISTORY

Rheumatoid arthritis and hypercholesterolemia.

DIFFERENTIAL DIAGNOSIS

Myocardial ischemia, valvular heart disease, cardiomyopathy including dilated or tachycardia-induced and intracardiac shunt.

INVESTIGATIONS

Transthoracic echocardiography (TTE) demonstrated normal biventricular systolic function with 4-chamber dilation, no valve disease, and massively dilated coronary sinus (CS) and left main coronary artery. Color and spectral Doppler showed continuous high-velocity flow from the CS into the right atrium (**Figures 1A to 1D**). Coronary computed tomography angiogram confirmed a large coronary artery fistula (CAF) from the left circumflex coronary artery (LCX) draining into the CS (**Figure 1E**). Pharmacological nuclear stress test was negative for ischemia. Cardiac catheterization demonstrated a

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significant left-to-right shunt (Qp:Qs 2.5:1.0), elevated pulmonary artery pressure (50/21 mm Hg, mean 32 mm Hg), elevated capillary wedge pressure (mean 16 mm Hg), and normal pulmonary vascular resistance (1.6 WU).

MANAGEMENT

Given symptoms of heart failure attributed to a hemodynamically significant shunt and elevated pulmonary pressure, CAF closure was indicated. Transcatheter device closure using an Amplatzer vascular plug (AVP II, St. Jude Medical, Saint Paul, Minnesota) was performed with no residual shunting (**Figure 2**). Four months post-procedure, the patient was clinically well with resolution of symptoms, normalization of pulmonary artery pressures, and improvement in ventricular dilation. She was tolerating warfarin, with repeat laboratory testing confirming resolution of her VWd. Shortly thereafter, patient re-presented with acute chest pain, electrocardiogram changes, and elevated troponin-I level (peak 30 ng/ml) consistent with a non-ST-segment elevation myocardial infarction (MI). TTE showed new inferior wall hypokinesis and reduced ejection fraction to 45%. Coronary angiography showed no coronary thrombus or obstruction, thrombosed CAF with no residual flow, and a nondominant right coronary artery (**Figure 3**). The left main and LCX coronary arteries remained severely dilated with sluggish flow. We attributed this ischemic event to be thromboembolic into the distal LCX circulation, given no other obvious angiographic evidence of coronary obstruction. Chest pain resolved and troponin normalized, with observant management.

DISCUSSION

CAF is an abnormal connection between 1 or more coronary arteries and a chamber of the heart or major thoracic vessel, resulting in left-to-right shunting. Most CAF are congenital, have a similar prevalence in males and females, and account for approximately 0.1% of coronary anomalies (1). A majority are of no clinical consequence because the shunt volume is often hemodynamically insignificant. However, shunt volume may be of sufficient size to result in arrhythmia, symptoms of heart failure, pulmonary hypertension, and rarely, myocardial ischemia from a coronary steal phenomenon (2). Larger fistulas may be associated with marked and progressive dilation of the originating coronary artery that often does not resolve post-closure. Although CAF can be diagnosed in childhood, symptomatic CAF are more commonly encountered among adults (3).

Due to the rarity of large, hemodynamically significant CAF, our knowledge of the natural history, indication, and method for closure (i.e., surgical vs. transcatheter), and long-term outcome post-closure remains limited.

MI post-CAF closure is a known potential complication and may occur as a result of stagnant flow in the dilated originating coronary artery leading to thrombus formation and obstruction (4). Small single-center studies suggest older age at time of closure, distal origin of the CAF off the coronary artery, and

ABBREVIATIONS AND ACRONYMS

CAF = coronary artery fistula

CS = coronary sinus

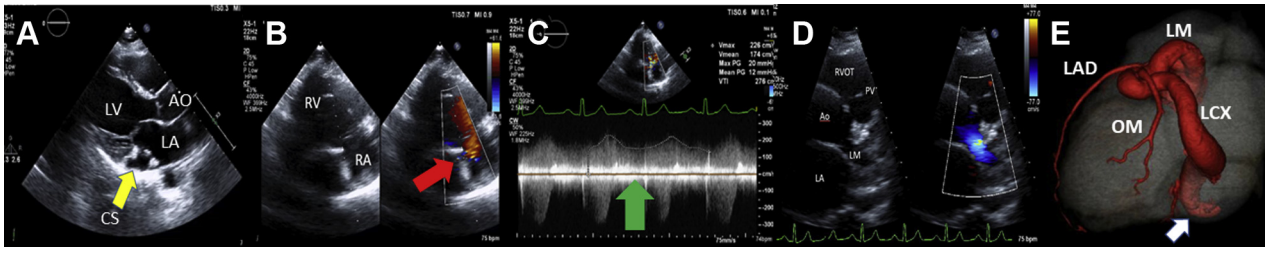
LCX = left circumflex coronary artery

MI = myocardial infarction

TTE = transthoracic echocardiography

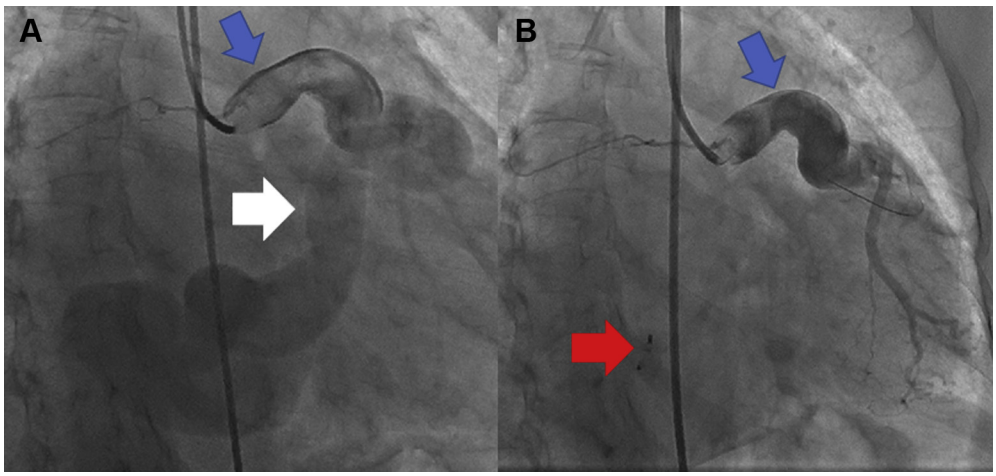
VWd = von Willebrand disease

FIGURE 1 CAF Pre-Closure TTE and CT Imaging



(A) Parasternal long-axis view showing dilated coronary sinus **(yellow arrow)**. **(B)** Right ventricle inflow view showing high velocity turbulent flow **(red arrow)** from the coronary sinus into right atrium. **(C)** Continuous-wave Doppler showing continuous high-velocity flow throughout the cardiac cycle from the coronary sinus into right atrium **(green arrow)**. **(D)** Short-axis view shows dilated left main coronary artery with laminar flow. **(E)** Coronary CT angiogram 3-dimensional volume rendering demonstrating a dilated LM and LCX with normal caliber LAD and OM branches. CAF connection between LCX and CS thought to be located distally in the LCX **(white arrow)** based on distal branches coming off the LCX. AO = ascending aorta; CAF = coronary artery fistula; CS = coronary sinus; CT = computed tomography; LA= left atrium; LAD = left anterior descending coronary artery; LCX = left circumflex coronary artery; LM = left main coronary artery; LV = left ventricle; OM = obtuse marginal coronary artery; PV = pulmonary valve; RA = right atrium; RV = right ventricle; RVOT = right ventricular outflow tract; TTE = transthoracic echocardiography.

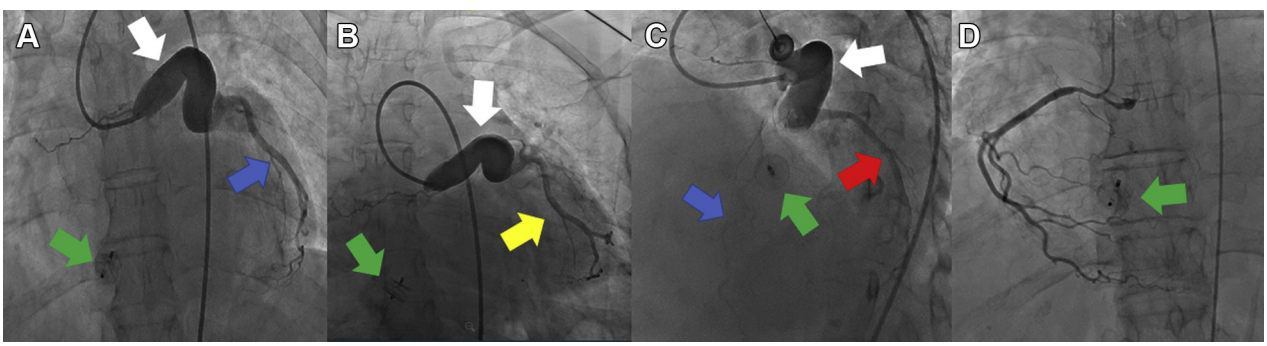
FIGURE 2 Intraprocedural Coronary Angiography During CAF Closure



Contrast injection into the left coronary system before **(A)** and after **(B)** coronary artery fistula (CAF) device closure **(red arrow)**. Note fistula contrast flow pattern before **(white arrow)** and after **(red arrow)** device placement, as the left main coronary artery remains dilated **(blue arrow)**.

CAF drainage into the CS have a higher association with post-closure MI, all of which were present in our patient (4,5). Timing of MI is variable and has been described both early and late after closure (4-6). Valente et al. (4) describe 76 cases of CAF, of which 64 underwent closure. Post-closure MI occurred in 7 patients, with 1 patient experiencing MI as far out as 4 years post-closure while on therapeutic warfarin. Traditional atherosclerotic risk factors, in addition to older age at time of closure and drainage into the CS, were associated with post-closure MI in this series. El-Sabawi et al. (6) described 45 patients that underwent transcatheter closure of CAF that resulted in 2 cases of post-closure MI thought to be secondary to stagnation of flow in the dilated coronary artery despite adequate anticoagulation. Gowda et al. (5) reported their experience with 16 CAF cases that underwent closure, with 3 experiencing MI (immediately, 9 months, and 42 years post-CAF closure, respectively), and the 2 latter MI events occurred despite anticoagulation. Management of post-closure MI was incompletely described in these series, with

FIGURE 3 Coronary Angiography Post-MI Diagnosis



(A and B) RAO view, left system injection showing the left main **(white arrow)**, LAD **(blue arrow)**, and the marginal artery **(yellow arrow)** with no filling defects. **(C)** LAO view, left system injection showing the LAD **(blue arrow)** and LCX systems **(red arrow)** with no filling defects. **(D)** LAO view, right injection showing the nondominant RCA with proximal spasm, but no filling defects. LAO = left anterior oblique; RCA = right coronary artery; RAO = right anterior oblique; other abbreviations as in Figure 1.

some managed conservatively and others undergoing bypass surgery. Optimal medical therapy to prevent an initial MI or recurrent MI post-CAF closure remains unknown. However, limited data advocate anticoagulation post-closure, especially if the originating coronary artery is dilated.

Despite anticoagulation and aspirin therapy, our patient experienced a post-CAF MI event. Given no angiographic evidence of intracoronary thrombus or obstruction, she was managed expectantly. To further reduce risk, clopidogrel and statin therapy were added to her regimen, and her international normalized ratio goal was increased to 3.0. Fortunately, she was found to have resolution of her VWd and was tolerating antithrombotic therapy. In retrospect, VWd was presumed to be acquired from shear stress related to high flow through the CAF into the CS, which was abolished post-CAF closure.

FOLLOW-UP

Currently, the patient is asymptomatic and has returned to an active lifestyle 8 months post-procedure. Current plan is to continue warfarin, aspirin, and clopidogrel indefinitely with continued aggressive cardiovascular risk factor modification, given expectation that her left main and LCX coronary arteries will remain dilated, associated with an undefined risk of recurrent MI.

CONCLUSIONS

Undiagnosed congenital heart disease should be considered in the differential diagnosis of an adult presenting with heart failure and murmur. CAF is a rare cause of heart failure, and closure is indicated when a hemodynamically significant shunt is present. Data are limited regarding long-term outcome and optimal medical management post-closure, but post-closure MI remains a concern.

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KEY WORDS chronic heart failure, congenital heart defect, coronary angiography, coronary vessel anomaly