Diagnosis of Coronary Artery Fistulas: Clinical Aspects and Brief Review of the Literature

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Abstract

Keywords

- congestive heart failure
- fistulas
- cardiac catheterization
- angiography
- coronary arteries

An 86-year-old woman with history of hypertension, diabetes, hyperlipidemia, and coronary artery disease was admitted with new-onset congestive heart failure and pneumonia. She underwent coronary angiography after suffering a non-ST elevation myocardial infarction. There was severe coronary artery disease in addition to coronary artery–pulmonary fistulas involving proximal right coronary artery and a branch of left main coronary artery.

Coronary artery fistula (CAF) is an abnormal communication between one or more coronary arteries and great vessels or a cardiac chamber. We reviewed 15 cases of CAFs published in PubMed and studied the clinical features of CAFs.

Case Report

An 86-year-old woman with a history of hypertension, diabetes, coronary artery disease, and arrhythmias was presented to the emergency department with a productive malaise, fever, worsening shortness of breath, and diaphoresis. The patient was normotensive, tachycardic, hypoxic, with regular heart sounds without associated murmur with clinical evidence of congestive heart failure (distension of jugular veins and bilateral basal lung crackles).

A 12-lead electrocardiogram revealed sinus tachycardia with a heart rate of 124 beats per minute with new T-wave inversions in lead aVL, and ST depressions in leads V5 and V6. Beta natriuretic peptide was 546 pg/mL. Chest X-ray demonstrated diffuse pulmonary edema.

The patient initially was placed on a nitroglycerin drip; however, her blood pressure dropped substantially, and therefore, this medication had to be discontinued. The patient also received nebulizer treatments and oxygen that resulted in improvement of her clinical condition. She was admitted for management of congestive heart failure and pneumonia. She was started on medical treatment of pneumonia (Levofloxacin, Sagent Pharmaceuticals, Schaumburg, IL) and congestive heart

failure (furosemide, β -blockers, and angiotensin-converting enzyme inhibitor). Echocardiogram demonstrated multiple wall motion abnormalities, ejection fraction of 30 to 35% with almost global hypokinesis with akinetic areas in the mid septal, basal inferior, and inferoseptal areas, two-fourth diastolic dysfunction, mild mitral regurgitation, and mild pulmonary hypertension. Cardiac catheterization revealed severe triple vessel disease with left dominant system, and aneurysmal left main disease with a critical lesion of proximal circumflex with ulcerated plaque that was subsequently stented. The patient also had a 70% proximal left anterior descending (LAD) lesion and coronary artery to pulmonary artery fistulas originating from the left main coronary artery (► **Fig. 1**) and right coronary artery (► **Fig. 2**) with small shunt flow.

Discussion

Coronary artery fistula (CAF) is an abnormal communication between a coronary and a heart chamber or any segment of the systemic or pulmonary circulation. These are often grouped together as coronary arterial–venous fistulas. A coronary artery connection to the pulmonary

Fig. 1 Coronary angiogram showing a coronary-to-pulmonary artery fistula at the level of a left main coronary artery.

artery (CAPF) may also be grouped under this heading. They can be congenital or acquired. CAFs account for approximately 14% of all congenital cardiac defects. CAFs can also be acquired as a complication of chest trauma, coronary angioplasty, pacemaker placement, or bypass surgery.¹⁻³

We reviewed 15 cases of CAFs in the literature and studied the clinical features of this congenital anomaly. Females accounted for two-thirds of the cases, with a median age of 59 years (range, 40–79 years). Males accounted for one-third of the cases, with a median age of 58 years (range, 41–64 years). All major coronary arteries were involved: right coronary artery (37%), left coronary artery (33%), LAD artery (26%), and left circumflex artery (4%). One-third of these cases were associated with aneurysms. The most common presenting symptoms were as follows: chest pain (60%), dyspnea on exertion (46%), syncope (20%), palpitations (26%), and asymptomatic (13%). About 33% of cases had only 1 fistula, 40% had 2 fistulas, and 27% had 3 fistulas. Approximately, 40% were managed conservatively, 27% had coil embolization, 27% had surgical treatment, and 13% had catheter-mediated occlusion of the fistula.¹⁻¹⁶

Dodge-Khatami et al¹⁷ have classified CAFs as follows:

Type A: Proximal type CAFs with dilatation of coronary segment proximal to fistula.¹⁷

Type B: Distal type CAFs with coronary artery dilated over the entire length and terminating as a fistula in right side of heart (end-artery type).¹⁷

CAF is suspected when a continuous murmur with diastolic accentuation at the left sternal edge is heard on a routine clinical examination. Patients can present with signs of heart failure or angina, with diffuse apex beat with palpable or audible third heart sound (S3) gallop in a large fistula.^{5,7}

Most patients with CAF remain asymptomatic, but elderly patients can present with exertional chest pain and dyspnea, fatigue, congestive heart failure, palpitations, or arrhythmias.^{1,2,5,8}

Most fistulas are small and hemodynamically inconsequential. However, some can be large and lead to preferential blood flow from coronary circulation to low-pressure pulmonary circulation, resulting in pulmonary hypertension and coronary-steal-related chronic myocardial ischemia.^{2,9,10} Chronic coronary-steal leads to ischemic cardiomyopathy,

Fig. 2 Coronary angiogram showing a right coronary artery branch to pulmonary artery fistula.

myocardial infarction, congestive heart failure, arrhythmias, thrombosis, fistula rupture, and endocarditis.^{1,2,9,11}

Most CAFs are incidentally discovered on coronary angiograms. Selective coronary angiogram can define the origin of a fistula, the total number of fistulas, and the presence of an aneurysm. However, it is an invasive procedure that provides two-dimensional images that may not be able to clearly define fistula course and anatomical relationship with other adjacent structures.⁸ A newer technology, like multidetector row computed tomography, is a noninvasive tool recommended by the American Heart Association Committee to

Table 1 Suggested indications for surgery in patients with coronary artery fistulas¹

- Arteriovenous shunt more than 30%.
- Electrocardiographic evidence of ischemia.
- Advanced pulmonary hypertension.
- Advanced congestive heart failure.
- History of infective endocarditis.
- Risk of rupture resulting from aneurysmal change.
- Social influence of heart murmurs.^{3,5}

evaluate suspected or known CAF.^{12,13} Transthoracic echocardiography with color flow imaging can demonstrate a CAF noninvasively.¹³ Magnetic resonance angiography can also be used to determine the anatomic course of CAF.⁷

No consensus exists on the optimal management of CAFs. Management of these remains controversial, especially in asymptomatic patients. Various treatment modalities, like coil embolization, catheter-mediated stent occlusion, and surgical ligation are available.^{2,11} Small, asymptomatic CAFs can be managed conservatively, because they run a benign course and can close spontaneously.

Surgical closure is preferred in symptomatic or complicated fistulas but runs the risk of major cardiac surgery, myocardial infarction, or recurrence. **Table 1** lists suggested indications for surgery. Percutaneous transcatheter closure is the safe and preferred method for anatomic approachable vessels.^{2-4,8,14-16}

Conclusion

CAFs are an abnormal communication between one or more coronary arteries and the great vessels or a cardiac chamber. They can be congenital or acquired. They account

for approximately 14% of all congenital cardiac defects. They could also be acquired as a complication of chest trauma, coronary angioplasty, pacemaker placement or coronary artery bypass surgery. Approximately, 15 to 30% of these are CAPF. Usually, they are found during routine echocardiography or coronary angiography. Most fistulas are hemodynamically inconsequential. However, large fistulas can lead to pulmonary hypertension and coronary-steal-related myocardial ischemia. Chronic coronary steal can lead to complications such as congestive heart failure, arrhythmias, infective endocarditis, and fistula rupture. No consensus exists on the optimal management of these fistulas. Treatment remains controversial, especially in asymptomatic patients.

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