

Case Report

Incidental Diagnosis of a Large Coronary Fistula: Angiographic and Cardiac MRI Findings

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Coronary artery fistulas are rare anomalies of the coronary arteries that may sometimes be totally asymptomatic and can be found incidentally during coronary arteriography later in adult life. We report the case of a 75-year-old man with such a fistula and describe our diagnostic approach. In addition, we review the literature on the etiology, epidemiology, the diagnostic modalities and treatment of coronary fistulas.

Coronary artery fistulas are rare anomalies of the coronary arteries, which may be associated with other congenital heart disease or occur later in life as a result of accident. Here we describe the case of a patient in whom such a fistula was an incidental finding during coronary arteriography.

Case presentation

A 75-year-old man was referred to our institution for coronary angiography for evaluation of atypical chest pain with a positive treadmill stress test. Cardiac echocardiography had demonstrated normal left ventricular size and function, and borderline right ventricular size with normal systolic function.

Selective coronary arteriography of the left coronary artery demonstrated a tortuous and dilated vessel originating from the left main artery, coursing posteriorly and laterally and eventually attaching to the right heart (Figure 1). The right coronary artery had the usual anatomy. Mild atherosclerosis, without any significant stenoses, was noted in the coronary arteries. Right heart catheter-

ization demonstrated a marginal step-up of oxygen saturation in the right atrium, compared to the superior and inferior *vena cava* mixed venous blood, suggesting drainage of the anomalous coronary vessel into the right atrium. The pulmonary to systemic flow ratio (Qp/Qs) was calculated at 1.2:1, suggestive of a small shunt.

The patient subsequently underwent a cardiac magnetic resonance study to better define the anatomy and course of the anomalous vessel. The study confirmed the presence of a large anomalous vessel originating from the distal part of the left main coronary artery, coursing between the aorta and left atrium and ending at the juncture between the superior *vena cava* and right atrium (Figure 2). The pulmonary to systemic flow ratio (Qp/Qs), was measured at 1.3:1, comparable with the findings of cardiac catheterization.

As the patient's symptoms were rather atypical, a dobutamine stress echo test was performed to assess the clinical significance of the anomalous coronary fistula. No wall motion abnormalities were induced at a high dobutamine dose (40 µg/

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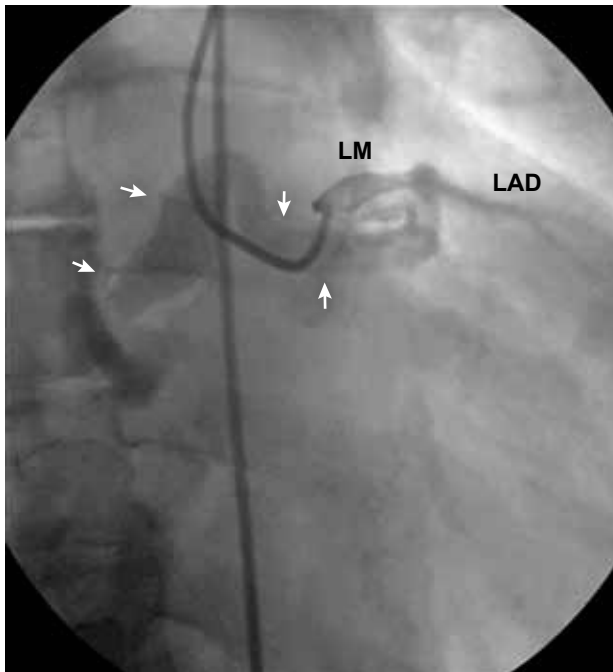


Figure 1. Right anterior oblique view of the left coronary artery. A large fistulous and tortuous channel (arrows) derives from the distal left main artery, initially following the course of the circumflex artery (proximally) and the proceeding to the receiving chamber in the right heart. LAD – left anterior descending artery; LM – left main coronary artery.

kg/min) and thus it was decided not to pursue surgical or interventional therapy for closure of the coronary fistula. At the 6-month follow-up visit the patient was stable without any significant changes in his clinical status.

Discussion

A coronary artery fistula is an abnormal communication between an epicardial coronary artery and a cardiac chamber, major vessel (*vena cava*, pulmonary vein or pulmonary artery), or other vascular structure (mediastinal vessels, coronary sinus).

In a study involving approximately 33,000 patients undergoing coronary arteriography, coronary artery fistulas were found in 0.1%.^{1,2} Fistulas from the right coronary artery are more common than from the left, and over 90 percent of fistulas drain into the venous circulation.^{1,3} Most coronary artery fistulas are single communications, but multiple fistulas have been identified.⁴ Coronary fistulas may be associated with other congenital heart disease or may occur later in life as a result of accident.⁵

The age at diagnosis varies widely. Many coronary artery fistulas are small and found incidentally during coronary arteriography, whereas others are identified as the cause of a continuous murmur, myocardial ischemia and angina, acute myocardial infarction, sudden death, congestive heart failure, endocarditis, stroke, arrhythmias, or superior *vena cava* syndrome. Some of the symptoms may be attributable to the amount of blood shunting from left to right and others to coronary hypoperfusion from the steal phenomenon. Myocardial ischemia has been documented in some patients with coronary artery fistulas who have no evidence of atherosclerosis.

Two-dimensional and color Doppler echocardiography may depict dilation of the affected coronary artery, although imaging of coronary arteries as well as the identification of the site of drainage may



Figure 2. Transverse images from a three-dimensional high-resolution magnetic resonance coronary angiography study with respiratory navigator gating and prospective slice correction. The study demonstrates an anomalous vessel (arrows) originating from the distal part of the left main coronary artery (panel A), coursing between the aorta and left atrium (panel B), and ending at the junction between the superior vena cava and the right atrium (panel C). Ao – aorta; LA – left atrium; SVC – superior *vena cava*; PA – pulmonary artery.

be difficult in adults.⁶ Magnetic resonance imaging helps in confirming the diagnosis, as the proximal coronary arteries and the entire length of the fistulous vessel can usually be well visualized.⁷⁻⁹ In older patients, stress myocardial perfusion imaging or stress echocardiography may be used to assess myocardial ischemia before and after treatment.¹⁰ Recently, multidetector row computed tomography (MDCT) cardiac imaging has provided excellent distal coronary artery and side branch imaging and can be used for the assessment of coronary anomalies.¹¹⁻¹³

The natural history of coronary artery fistulas is variable,³ with long periods of stability in some and sudden onset or gradual progression of symptoms in others. Spontaneous closure is rare but may occur in small fistulas. Small fistulous connections in the asymptomatic patient may be followed. Most lesions enlarge progressively and warrant repair, by either transcatheter or surgical techniques. Direct medical treatment for symptomatic relief can be used until investigations and operative repair can be performed. Endocarditis prophylaxis is advised in all patients. Surgical repair of a coronary artery fistula is recommended in patients who are symptomatic, those who have a major shunt and in those who have myocardial ischemia, pulmonary hypertension without Eisenmenger syndrome, or congestive heart failure. Other indications for coronary artery fistula closure are arrhythmias, infectious endocarditis and fistula rupture with cardiac tamponade. Controversy still persists as to whether fistula closure is justified in asymptomatic patients.

Current treatment options for coronary artery fistulas include surgical ligation alone (either with or without cardiopulmonary bypass), surgical ligation accompanied by coronary artery bypass surgery, and transcatheter closure.¹⁴⁻¹⁸ The introduction of minimally invasive percutaneous approaches has, however, broadened the indications for closure. Devices currently used for percutaneous approaches are Gianturco coils, detachable balloons, polyvinyl alcohol foams and plug devices. Intraluminal or percutaneous closure of coronary artery fistulas is associated with decreased mortality, morbidity and hospital stay compared with open surgery.

In our case the shunt was rather small, the ventricular size and function was normal, and there was no evidence of myocardial ischemia, as demonstrated by dobutamine stress echocardiography. Based on these findings, our patient was considered to have no definite indication for invasive intervention and indeed he is doing well on medical therapy.

In conclusion, coronary artery fistulas are rare anomalies of the coronary arteries that have traditionally been diagnosed with the use of echocardiography and angiography. New imaging modalities, such as MRI or MDCT, are very useful for the diagnosis of coronary artery fistulas. The choice between surgical and percutaneous treatment should be based upon the anatomical and functional characteristics of the fistula.

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