Coronary artery fistula between the left anterior descending artery and pulmonary artery

Abdul Al-Douri, BSc^a, Ari Cedars, MD^b, and Dat Tran, MD^c

^aDepartment of Internal Medicine, Texas A&M College of Medicine, Bryan, Texas; ^bDivision of Cardiology, Baylor Heart and Vascular Hospital and Baylor University Medical Center, Dallas, Texas; ^cDepartment of Internal Medicine, Baylor University Medical Center, Dallas, Texas

ABSTRACT

Coronary artery fistula is a rare anomalous connection between a coronary artery and a cardiac chamber or major vessel. Here we present the case of a 36-year-old woman who presented with sharp, substernal chest pain, dyspnea, and dizziness. A fistula between her left anterior descending artery and pulmonary trunk was found. Herein, we describe our approach and review the epidemiology, pathophysiology, diagnosis, and treatment options.

KEYWORDS Congenital heart disease; coronary artery anomaly; coronary artery fistula; pulmonary artery

Congenital artery fistulas (CAFs) are very uncommon, seen in between 0.17% and 0.64% of asymptomatic children and adolescents.¹ The majority of persistent fistulous connections are small, with very limited flow, and are of no clinical consequence. Nevertheless, an awareness that fistulas may be present is important because in rare cases they may be associated with clinical disease including chest pain or heart failure in young adults.² Typically, fistulas are diagnosed by computed tomography angiogram, coronary angiogram, magnetic resonance imaging/angiogram, or transesophageal echocardiogram.³ Several complications may arise later in life due to these fistulas; therefore, closure of large CAFs is recommended in cases of symptoms or significant shunting.⁴

CASE PRESENTATION

A 36-year-old white woman with a past medical history of type 2 diabetes mellitus, depression, and obesity initially presented with syncope at home associated with chest pain, dyspnea, and dizziness. Electrocardiogram and cardiac markers were unremarkable. Nuclear stress testing suggested possible anterior wall ischemia. Coronary angiography revealed multiple fistulous connections between the mid-left anterior descending (LAD) coronary artery and the pulmonary trunk. As fistulous flow was believed to be insignificant, the patient was discharged home with nitroglycerin.

Due to continued chest pain, the patient returned to the hospital within a week. The pain was described as sharp, substernal, radiating to her back, left arm, and left side of her neck, and rated at a 7–8/10. The pain was relieved by

nitroglycerin for about 30 minutes but always recurred. The pain was provoked by both exertion and anxiety. Computed tomography angiogram showed a small, tortuous LAD-to-pulmonary artery fistula that was estimated to be about 3 mm in diameter (*Figure 1*).

After much discussion, the patient underwent attempted coiling of the fistula. Arterial access was obtained in the right common femoral artery and the coronary arteries were visualized. The procedure was complicated by extreme difficulty in engaging and stabilizing the catheter tip in the fistula from both the LAD and pulmonary arterial sides of the connection. Ultimately, the main fistulous tract was successfully coiled from the LAD side using a 3 mm × 2 mm tornado coil, with 2 to 3 very small residual LAD-to-pulmonary artery fistulas remaining. The distal aspect of the implanted tornado coil protruded into the LAD representing a risk for thrombosis. After multiple attempts to more securely implant the coil, an Alpine Rx 3.5 mm × 12 mm stent was placed to pin the protruding coil to the arterial wall in the LAD and decrease the risk of thrombosis.

Thereafter, the patient reported substantial improvement in her chest pain. She was started on daily aspirin 81 mg and clopidogrel 75 mg, in addition to as-needed nitroglycerin 0.4 mg and twice daily carvedilol 3.125 mg. At outpatient follow-up, she was chest pain free.

DISCUSSION

Congenital CAFs are rare congenital anomalies that involve an abnormal communication between a coronary artery and one of the cardiac vessels or chambers adjacent

Corresponding author: Ari Cedars, MD, Baylor Heart and Vascular Hospital and Baylor University Medical Center, 621 North Hall St., Suite 120, Dallas, TX 75226 (e-mail: acedars97@gmail.com)

Color versions of one or more of the figures in the article can be found online at www.tandfonline.com/ubmc.

Figure 1. Coronary angiography showing the patient's fistula between the left anterior descending and pulmonary artery.

to the heart. The fistulas are usually dilated and tortuous. They originate from the right coronary artery in approximately 52% of cases, LAD in approximately 30% of cases, and left circumflex artery in 18% of cases.⁵

Based on a study by Zamani et al,⁶ CAFs are usually asymptomatic during the first 2 decades of life, after which the frequency of both symptoms and complications increases. They reported that most cases of CAF with LAD origin, like that presented in this case, were first diagnosed in adulthood.⁶ Complications from CAF include steal mediated myocardial ischemia, thrombosis and embolism, high-output cardiac failure, atrial fibrillation, rupture, endocarditis/endarteritis, and arrhythmia.⁷ Patients most commonly present with symptoms of dyspnea or angina pectoris and occasionally arrhythmias. In patients with high-flow CAFs, a continuous precordial murmur may be auscultated. Steal phenomena may occur in previously asymptomatic patients possibly due to a small fistula increasing in size or secondary to changes in coronary physiology related to the onset of age-related vascular disease or endothelial dysfunction.⁸

The American College of Cardiology/American Heart Association guidelines for the management of adult congenital heart disease recommend closure of all large CAFs regardless of symptoms. Closure of small to moderate fistulae is recommended only in symptomatic patients (including those with angina pectoris, ischemia, arrhythmia, and unexplained contractile dysfunction thought to be attributable to the fistula). Closure is not recommended for

patients with small, asymptomatic fistulae. Follow-up with echocardiography every 3 to 5 years is encouraged for all patients with CAFs due to the possibility of small or moderate-sized fistulae enlarging over time.⁹ In certain borderline cases without significant shunting, medical therapy may be an option, and there have been reports of symptom resolution with optimal antianginal medical therapy. For patients meeting criteria for closure, the two primary treatment options are surgical ligation or catheter-based coil occlusion. Transcatheter closure has become the treatment of choice in patients without any additional cardiac disease. No formal clinical studies comparing the two options have been undertaken. Instead, only case reports comparing small samples of the two options exist.^{10–13} One such study, by Jama et al,¹⁴ examined the clinical and angiographic outcomes of a sample of 36 patients at the Mayo Clinic in Rochester, Minnesota, who had undergone catheter occlusion of CAF. Of the 36, 32 (89%) had no residual flow and 4 (11%) had trivial residual flow. Complications included two instances of device migration, one of coronary spasm, and one of coronary thrombosis. There were no deaths or long-term morbidity. It was concluded that transcatheter closure of CAF is an acceptable and safe alternative to surgery in most patients.

1. Kimbiris D, Iskandrian AS, Segal BL, Bemis CE. Anomalous aortic origin of coronary arteries. *Circulation*. 1978;58:606–615. doi:10.1161/01.CIR.58.4.606.
2. Neufeld HN, Schneeweiss A. *Coronary Artery Disease in Infants and Children*. Philadelphia, PA: Lea & Febiger; 1983.
3. Davis JA, Cecchin F, Jones TK, Portman MA. Major coronary artery anomalies in a pediatric population: incidence and clinical importance. *J Am Coll Cardiol*. 2001;37:593–597. doi:10.1016/S0735-1097(00)01136-0.
4. Frommelt PC, Frommelt MA, Tweddell JS, Jaquiss RD. Prospective echocardiographic diagnosis and surgical repair of anomalous origin of a coronary artery from the opposite sinus with an interarterial course. *J Am Coll Cardiol*. 2003;42:148–154. doi:10.1016/S0735-1097(03)00503-5.
5. Challoumas D, Pericleous A, Dimitrakaki IA, Danelatos C, Dimitrakakis G. Coronary arteriovenous fistulae: a review. *Int J Angiol*. 2014;23:1–10. doi:10.1055/s-0033-1349162.
6. Zamani H, Meragi M, Arabi Moghadam MY, Alizadeh B, Babazadeh K, Mokhtari-Esbaie F. Clinical presentation of coronary arteriovenous fistula according to age and anatomic orientation. *Caspian J Intern Med*. 2015;6(2):108–112.
7. Barsoum EA, Saiful FB, Asti D. Rare case of coronary to pulmonary vein fistula with coronary steal phenomenon. *World J Cardiol*. 2014;6:682–684. doi:10.4330/wjc.v6.i7.682.
8. Graham TP, Driscoll DJ, Gersony WM, Newburger JW, Rocchini A, Towbin JA. Task Force 2: congenital heart disease. *J Am Coll Cardiol*. 2005;45:1326–1333. doi:10.1016/j.jacc.2005.02.009.
9. Warnes CA, Williams RG, Bashore TM. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines for the Management of Adults with Congenital Heart Disease. *Circulation*. 2008;118:e714–e833. doi:10.1161/CIRCULATIONAHA.108.190690.
10. Cheng TO. Management of coronary artery fistulas: percutaneous transcatheter embolization versus surgical closure. *Catheter Cardiovasc Interv*. 1999;46(2):151–152. doi:10.1002/(SICI)1522-726X(199902)46:2<151::AID-CCD7>3.0.CO;2-G.
11. Reidy J, Anjos R, Qureshi S, Baker E, Tynan M. Transcatheter embolization in the treatment of coronary artery fistulas. *J Am Coll Cardiol*. 1991;18:187–192. doi:10.1016/S0735-1097(10)80239-6.

12. Perry S, Rome J, Keane J, Baim D, Lock J. Transcatheter closure of coronary artery fistulas. *J Am Coll Cardiol*. 1992;20:205–209. doi:10.1016/0735-1097(92)90160-O.
13. Armsby L, Keane J, Sherwood M, Forbess J, Perry S, Lock J. Management of coronary artery fistulae: patient selection and results of transcatheter closure. *J Am Coll Cardiol*. 2002;39:1026–1032. doi:10.1016/S0735-1097(02)01742-4.
14. Jama A, Barsoum M, Bjarnason H, Holmes DR, Rihal CS. Percutaneous closure of congenital coronary artery fistulae: results and angiographic follow-up. *JACC Cardiovasc Interv*. 2011;4:814–821. doi:10.1016/j.jcin.2011.03.014.