Journal of Cardiology (2009) **53**, 467—472

ELSEVIER

JOURNAL of CARDIOLOGY
Official Journal of the Japanese College of Cardiology
www.elsevier.com/locate/jjcc

CASE REPORT

Coronary artery fistula: A case series with review of the literature

Manjunath G. Raju (MD)[a], Sandeep K. Goyal (MD)[b,*], Sujeeth R. Punnam (MD)[c], Dinesh O. Shah (MD)[c], George F. Smith (MD)[a], George S. Abela (MD, FACC)[c]

[a] *Department of Family Medicine, Michigan State University, East Lansing, MI 48824, USA*
[b] *Department of Medicine, Boston University School of Medicine, One Boston Medical Center, Boston, MA 02118, USA*
[c] *Division of Cardiology, Michigan State University, East Lansing, MI 48824, USA*

Received 8 August 2008; received in revised form 27 September 2008; accepted 30 September 2008
Available online 20 November 2008

KEYWORDS
Coronary circulation;
Fistula;
Angina-pectoris;
Coil embolization;
Interventional cardiology

Summary Coronary artery fistula (CAF) is an anomalous connection between a coronary artery and a major vessel or cardiac chamber. Most of the coronary fistulas are discovered incidentally during angiographic evaluation for coronary vascular disorder. The management of CAF is complicated and recommendations are based on anecdotal cases or very small retrospective series. We present three cases of CAF, two of which were symptomatic due to hemodynamically significant coronary steal phenomenon. They underwent successful transcatheter coil embolization, leading to resolution of their symptoms. Percutaneous closure offers a safe and effective way for the management of symptomatic patients. CAFs are rare cardiac anomalies but can give rise to a variety of symptoms because of their hemodynamic consequences or complications. They should be part of cardiac differential diagnosis particularly in patients without other risk factors. Correction of CAF is indicated if the patients are symptomatic or if other secondary complications develop.

Introduction

Coronary artery fistula (CAF) is defined as an anomalous connection between a coronary artery and a major vessel or cardiac chamber. It is an uncommon form of congenital heart disease. Most of the coronary anomalies are found incidentally during angiographic evaluation for coronary vascu-

* Corresponding author. Tel.: +1 517 990 4496.
E-mail addresses: drmgraju@gmail.com (M.G. Raju), sandeep_mamc@rediffmail.com (S.K. Goyal), punnam@msu.edu (S.R. Punnam), dshah@gmail.com (D.O. Shah), smith@msu.edu (G.F. Smith), abela@msu.edu (G.S. Abela).

0914-5087/$ — see front matter
doi:10.1016/j.jjcc.2008.09.009

lar disorder. The majority of these fistulas arise from the left anterior descending artery or from the right coronary artery. Most of these patients are asymptomatic, but heart failure, angina, myocardial infarction, endocarditis, and dyspnea have been reported in some cases. The management is complicated and recommendations are based on anecdotal cases of very small retrospective series. We present three cases of CAF, two of which were symptomatic due to hemodynamically significant coronary steal phenomenon.

Case 1

A 36-year-old Caucasian male with a history of hypertension and hyperlipidemia presented to the emergency room with recurrent episodes of chest pain. His chest pain had some typical and atypical features of angina. Family history was significant for premature coronary artery disease in the father at the age of 38 years.

His vital signs were stable and physical examination was unremarkable. Electrocardiography showed sinus rhythm with no pathological Q waves, ST segment, or T wave changes. Serial cardiac enzymes were negative for myocardial damage. Echocardiogram showed normal left ventricular function with ejection fraction (EF) of 65% and mild concentric left ventricular hypertrophy. Dobutamine Cardiolite stress test was performed which revealed 1 mm ST segment depression in inferior leads on the EKG and imaging part of the stress test showed mild reversible inferior ischemia. Cardiac catheterization was performed which revealed a small fistula communicating from the left anterior descending artery to the pulmonary artery (Fig. 1). There was no significant atherosclerotic disease noticed in the coronary arteries and the right heart catheterization detected pulmonary artery pressure of 23/9 mmHg. Pulmonary to systemic blood flow ratio (Qp/Qs) was 1, which excluded the possibility of significant left to right shunt. Since the anterior wall did not show any ischemic changes on the stress test, coronary steal was not suspected from the fistula. No acute intervention was deemed necessary at this point and clinical follow-up was recommended. He was asymptomatic at a 3-month follow-up.

Case 2

A 48-year-old Caucasian male with a history of hypertension, type 2 diabetes mellitus, hyperlipidemia, obesity, and smoking was evaluated in the clinic for left precordial pain. He denied any history of dyspnea, palpitation, dizziness, or syncopal episode. His physical examination was unremarkable. Electrocardiogram showed sinus rhythm with first-degree AV block and Q waves in leads V1—V2 (old antero-septal infarction pattern). Echocardiogram showed normal left ventricular function with an EF of 60%. Exercise Cardiolite stress test showed reversible antero-septal ischemia.

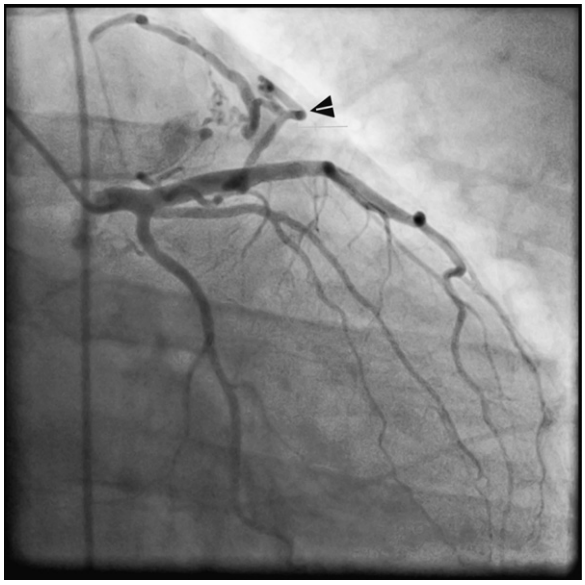

Figure 1 Small fistula connecting left anterior descending artery to pulmonary artery.

Cardiac catheterization did not show any atherosclerotic disease but was remarkable for a large CAF originating from the mid-left anterior descending artery to the pulmonary artery (Fig. 2). In view of the abnormal stress test showing anterior ischemia, we suspected coronary steal. Pulmonary artery pressure was 45/16 mmHg and Qp/Qs ratio was 1.4, indicating significant left to right shunt. He was referred to a tertiary care center specializing in coil embolization of the CAF. The fistula was sealed with two 4 mm $\times$ 2 mm and one 3.3 mm $\times$ 2 mm tornado coils by utilizing a 0.014 universal wire and a renegade catheter via the left anterior descending artery (Fig. 3). The patient remained chest pain-free after the procedure. A follow-up exercise Cardiolite stress test performed 6 months after the procedure was negative for inducible ischemia.

Case 3

A 47-year-old male with a history of hypertension, hyperlipidemia, and chronic obstructive lung

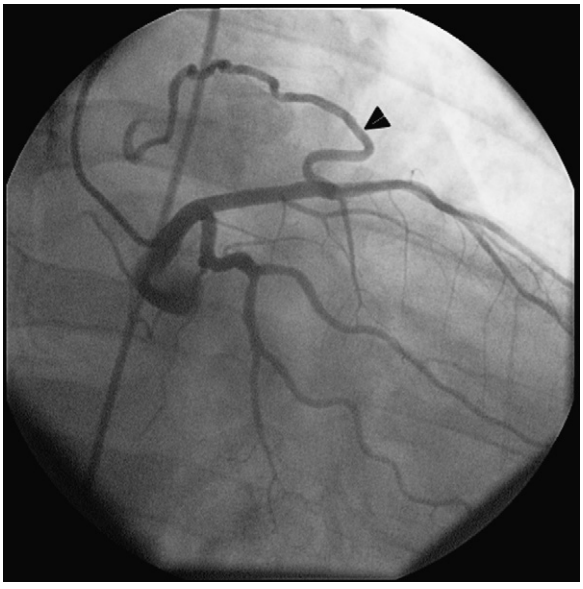

Figure 2 Fistula connecting mid-left anterior descending artery to pulmonary artery.

disease was evaluated for recurrent episodes of chest pain. Vital signs were stable and physical examination was benign. Electrocardiography showed sinus rhythm with Q waves in lead III and aVF. Echocardiogram showed moderate concentric left ventricular hypertrophy with EF of 67%. Exercise Cardiolite stress test revealed a normal study.

Since he continued to have recurrent chest pain, cardiac catheterization was performed, which showed an arteriovenous fistula connecting the conus branch of the right coronary artery to the main pulmonary artery (Fig. 4). Right heart catheterization revealed elevated pulmonary pressure of 40/14 mmHg. Qp/Qs ratio was calculated to be 1.3. He was referred to a tertiary care center for consideration of coil embolization. The conus branch of the right coronary artery was selectively cannulated with a renegade coil embolization delivery catheter over a 0.014 agility guidewire.

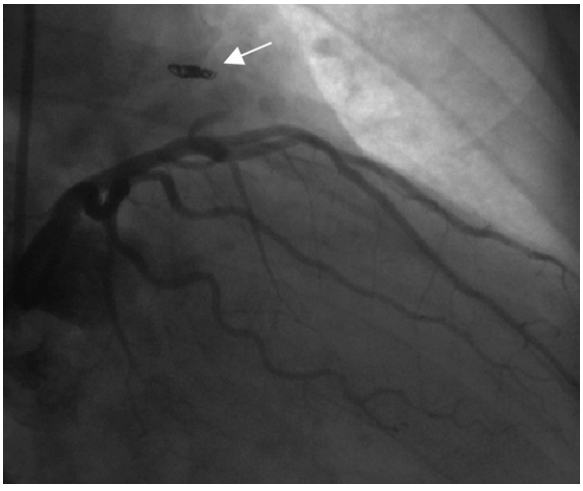

Figure 3 Post-coil embolization: coil sealing the fistula, compare with Fig. 2.

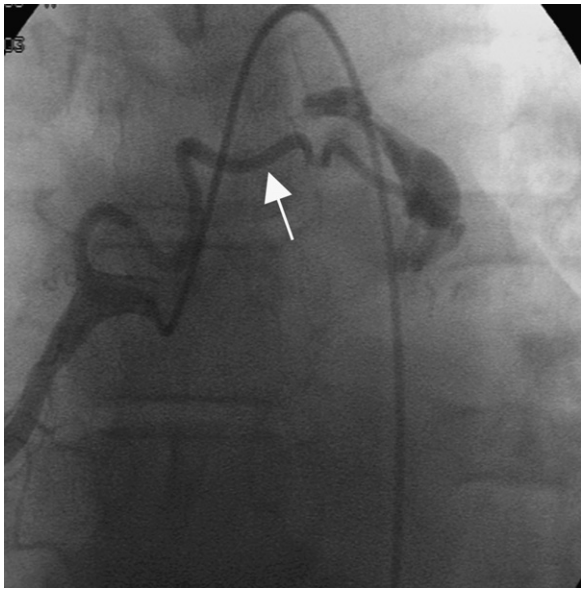

Figure 4 Fistula arising from the conus branch of the right coronary artery.

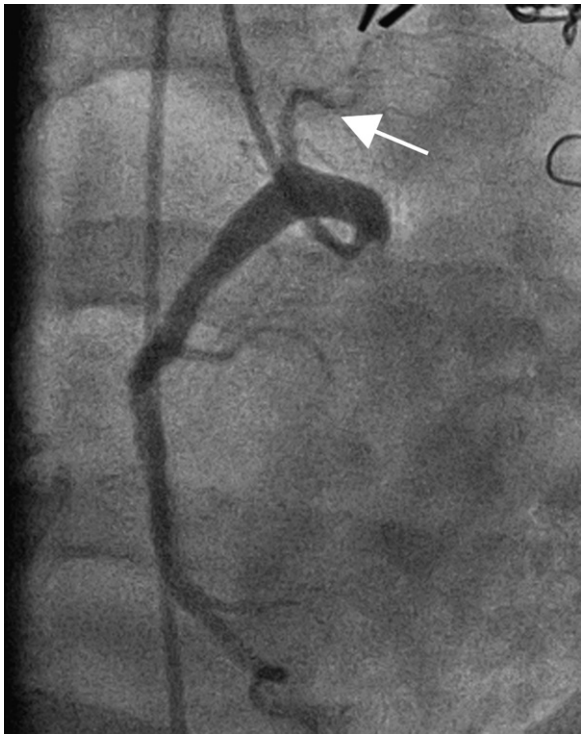

Figure 5 Obliterated fistula post-intervention, compare with Fig. 4.

Three Boston scientific complex helical coils were introduced into the distal part of the coronary AV fistula. This was followed by a 1.6 mm × 2 mm tornado micro-coil embolization and injection of 2 cm^3 diluted polyvinyl alcohol particles into the feeding AV fistula. Finally 100% occlusion of the fistula was achieved (Fig. 5). Patient remained chest pain-free at a 3-month follow-up. A follow-up exercise Cardiolite performed 7 months after the procedure was negative for inducible ischemia.

Discussion

The incidence of coronary anomalies varies between 0.6% and 1.5% of patients undergoing invasive cardiovascular imaging [1,2]. CAF was first described in 1841 and it is defined as an anomalous connection between a coronary artery and a major vessel or cardiac chamber [3]. Its incidence in the general population is about 0.002%. The incidence of coronary pulmonary fistulas was similarly reported only to be 0.1% in a study of 11 000 patients undergoing cardiac catheterization [4].

The majority of CAFs are congenital and not gender specific. Of the congenital fistulas, two major groups are identified: solitary CAFs or coronary artery-left ventricular multiple micro-fistulas (CA-LVMMFs). Contrary to the CA-LVMMFs, the solitary group may also have an acquired etiology, due to chest trauma or iatrogenic causes. Twenty percent of people with congenital CAFs have other concomitant cardiac anomalies, such as aortic and pulmonary atresia and patent ductus arteriosus.

Anatomic characteristics of fistulas vary between different studies. Earlier studies report right coronary system as the most frequent site of origin [5], while more recent studies suggest that the left system may be the more common site [6]. Most common sites of drainage include right ventricle [5] and pulmonary artery [6]. Left-sided chambers of the heart are less likely to be the draining site [5,6].

Clinical presentation depends on factors such as the age of patient, amount of shunting, development of cardiac ischemia, and resistance of recipient vessel or chamber. The majority of adult patients are asymptomatic and the lesion is detected on physical examination as a murmur or as an incidental finding during coronary angiography for an unrelated diagnosis. Among symptomatic patients, chest pain is the most common presenting symptom. In a large series of 51 patients with CAFs, angina pectoris occurred in 57% of cases [6] and was often present in the absence of underlying coronary artery disease. Ischemia is related to coronary steal, whereby blood flow is shifted away from the distal coronary vascular bed [7,8]. Other symptoms have also been reported including congestive heart failure, pulmonary hypertension, rupture [9], or thrombosis of fistula or an associated arterial aneurysm.

The classic physical finding described in patients with CAF is a soft, continuous murmur [10] that tends to be crescendo decrescendo in both systole and diastole but louder in diastole. In contrast, most of the other continuous murmurs reach their peak intensity at the time of the second sound. The location on the chest wall where the murmur is the loudest depends on where the fistula enters the heart. Sudden appearance of a pre-existing murmur after myocardial biopsy can provide a clue to the presence of iatrogenic CAF.

The natural history of fistulas is variable, with a long period of stability in some and a sudden onset or a gradual progression of symptoms in others. Spontaneous closure of CAFs is uncommon. Fifty percent of patients with large or multiple fistulas develop symptoms. Complications include bacterial endocarditis, thrombosis, rupture, premature atherosclerosis, pulmonary hypertension, and myocardial ischemia or infarction.

Coronary angiography remains the gold standard for diagnosis. Angiography helps define the artery of origin, recipient vessel, or chamber and the site of communication. Noninvasive techniques such as transthoracic echocardiography, transesophageal echocardiography and magnetic resonance imaging are becoming increasingly popular for diagnosis and follow-up of CAFs. Combined two-dimensional and pulsed Doppler echocardiography demonstrates a dilated coronary artery, turbulent flow in the fistula and the recipient chamber [11]. Transthoracic color Doppler echocardiography with a high frequency transducer and a low Nyquist limit allows multiple coronary arteries—left ventricular micro-fistulae to be visualized. Transesophageal echocardiography is used intraoperatively to identify the precise location of the site of drainage of the fistula, which could not be accurately revealed with preoperative coronary arteriography [12]. Magnetic resonance imaging and multidetector computed tomography have also become the alternative methods to evaluate the anatomy, flow, and function of CAF [11,13,14].

A variety of approaches can be used in the management of CAFs. Onset of symptoms such as chest pain and dyspnea is the primary indication for closure of fistula. Management strategy for asymptomatic fistulas remains controversial. But it

is prudent to reserve invasive strategy for asymptomatic fistulas when pulmonary to systemic flow ratio exceeds 1.5:1 or when aneurysmal degeneration occurs, which can lead to mural thrombosis and hazardous rupture or when side branch obstruction threatens to occur [15].

Bjork and Crafoord first reported surgical ligation in 1947 as an effective method of fistula closure. Its excellent long-term efficacy and safety [16] made it the procedure of choice and it remained so until 1983 [17] when the first percutaneous closure of fistula was reported. Catheter-based interventional techniques have become the procedure of choice in the current era, if technically feasible. A variety of materials have been used, including Gianturco coils, covered stainless-steel coils, detachable balloons, coaxial embolization with platinum micro-coils, double-umbrella devices, the Gianturco Grifka vascular occlusion device, and Amplatzer duct occluder. The selection of occlusion device is based on the anatomic features of the fistula. Coils have been used in small CAFs, and double-umbrella devices have been used in large CAFs. This procedure avoids the need for surgical intervention, cardiopulmonary bypass, and median sternotomy. The most frequent complication associated with catheter-based closure is the embolization of the occlusion device, and in one study this complication occurred in 7 (~17%) out of 40 patients. Surgical closure of the fistula is recommended in cases with extreme tortuosity of fistulous tract and/or aneurysm formation [18]. In two of our patients, catheter-based closure using controlled-release coil embolization resolved the CAF and the patient's symptoms.

We presented three cases of CAFs, two of which were symptomatic due to hemodynamically significant coronary steal phenomenon. They underwent successful transcatheter coil embolization, leading to the resolution of their symptoms. Percutaneous closure offers a safe and effective way for the management of symptomatic patients. The long-term patient outcome after fistula occlusion remains unknown, but intermediate-term results reveal persistent coronary artery dilatation in many of these patients [19]. Therefore, patients who have undergone coil occlusion of coronary fistulae require a close follow-up; and in certain cases, the use of antiplatelet therapy or low-dose anticoagulation may be warranted [19].

All symptomatic patients should undergo closure of medium to large CAFs. Asymptomatic or small coronary fistulas do not need to be treated; however, if these patients develop endocarditis or a long-term follow-up is not feasible, they should be treated [20].

Conclusions

CAFs are rare cardiac anomalies, but they should always be part of differential diagnosis of symptoms of chest pain and dyspnea, especially in patients without significant risk factors for acquired cardiac disease. They can give rise to a variety of symptoms because of their hemodynamic consequences or complications. Correction of CAF is indicated if the patients are symptomatic or if other secondary complications develop.

References

[1] Kardos A, Babai L, Rudas L, Gaal T, Horvath T, Talosi L, et al. Epidemiology of congenital coronary artery anomalies: a coronary arteriography study on a central European population. Cathet Cardiovasc Diagn 1997;42(3):270—5.
[2] Malouf JF, Edwards WD, Tajik AJ, Seward JB. Functional anatomy of the heart. In: Fuster VAR, editor. Hurst's the heart. 11th ed. New York: McGraw-Hill; 2004.
[3] Fernandes ED, Kadivar H, Hallman GL, Reul GJ, Ott DA, Cooley DA. Congenital malformations of the coronary arteries: the Texas Heart Institute experience. Ann Thorac Surg 1992;54(4):732—40.
[4] Said SA, Landman GH. Coronary-pulmonary fistula: long-term follow-up in operated and non-operated patients. Int J Cardiol 1990;27(2):203—10.
[5] Hobbs RE, Millit HD, Raghavan PV, Moodie DS, Sheldon WC. Coronary artery fistulae: a 10-year review. Cleve Clin Q 1982;49(4):191—7.
[6] Said SA, van der Werf T. Dutch survey of coronary artery fistulas in adults: congenital solitary fistulas. Int J Cardiol 2006;106(3):323—32.
[7] Gowda RM, Vasavada BC, Khan IA. Coronary artery fistulas: clinical and therapeutic considerations. Int J Cardiol 2006;107(1):7—10.
[8] Umana E, Massey CV, Painter JA. Myocardial ischemia secondary to a large coronary-pulmonary fistula—a case report. Angiology 2002;53(3):353—7.
[9] Iwasawa Y, Kitamura Y, Higuma K, Ono F, Imoto K, Kimura K. Cardiac tamponade due to rupture of coronary artery fistulas with a giant aneurysm containing a free floating ball thrombus: a case report. J Cardiol 2007;50(1):71—6.
[10] Haller Jr JA, Little JA. Diagnosis and surgical correction of congenital coronary artery-coronary sinus fistula. Circulation 1963;27:939—42.
[11] Hong GR, Choi SH, Kang SM, Lee MH, Rim SJ, Jang YS, et al. Multiple coronary artery-left ventricular microfistulae in a patient with apical hypertrophic cardiomyopathy: a demonstration by transthoracic color Doppler echocardiography. Yonsei Med J 2003;44(4):710—4.
[12] Iida R, Yamamoto T, Suzuki T, Saeki S, Ogawa S. The usefulness of intraoperative transesophageal echocardiography to identify the site of drainage of coronary artery fistula. Anesth Analg 2005;101(2):330—1.
[13] Lessick J, Kumar G, Beyar R, Lorber A, Engel A. Anomalous origin of a posterior descending artery from the right pulmonary artery: report of a rare case diagnosed by multidetector computed tomography angiography. J Comput Assist Tomogr 2004;28(6):857—9.

[14] Parga JR, Ikari NM, Bustamante LN, Rochitte CE, de Avila LF, Oliveira SA. Case report: MRI evaluation of congenital coronary artery fistulae. Br J Radiol 2004;77(918): 508—11.
[15] Angelini P. Coronary artery anomalies—current clinical issues: definitions, classification, incidence, clinical relevance, and treatment guidelines. Tex Heart Inst J 2002;29(4):271—8.
[16] Kamiya H, Yasuda T, Nagamine H, Sakakibara N, Nishida S, Kawasuji M, et al. Surgical treatment of congenital coronary artery fistulas: 27 years' experience and a review of the literature. J Card Surg 2002;17(2):173—7.
[17] Reidy JF, Sowton E, Ross DN. Transcatheter occlusion of coronary to bronchial anastomosis by detachable balloon combined with coronary angioplasty at same procedure. Br Heart J 1983;49(3):284—7.
[18] Armsby LR, Keane JF, Sherwood MC, Forbess JM, Perry SB, Lock JE. Management of coronary artery fistulae. Patient selection and results of transcatheter closure. J Am Coll Cardiol 2002;39(6):1026—32.
[19] Levin DC, Fellows KE, Abrams HL. Hemodynamically significant primary anomalies of the coronary arteries. Angiographic aspects. Circulation 1978;58(1): 25—34.
[20] McMahon CJ, Nihill MR, Kovalchin JP, Mullins CE, Grifka RG. Coronary artery fistula. Management and intermediate-term outcome after transcatheter coil occlusion. Tex Heart Inst J 2001;28(1):21—5.