New methods in diagnosis and therapy

Optimal treatment of coronary-to-pulmonary artery fistula: surgery, coil or stent graft?

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Abstract

We report a case of a 57-year-old man with typical angina due to a coronary artery-to-pulmonary artery fistula, which was evident on transthoracic and transesophageal echocardiography with color Doppler flow mapping. The diagnosis was confirmed by coronary angiography. The patient underwent surgical ligation of the fistula. However, repeated transesophageal echocardiography and coronary angiography revealed persistence of the fistula with significant left-to-right shunt. The orifice of the fistula was then obliterated by stent-graft implantation, which was proven successful by angiography and echocardiography.

Key words: coronary artery-to-pulmonary artery fistula, stent-graft implantation.

Introduction

Congenital coronary artery fistula (CAF) is a rare malformation that involves the coronary artery allowing blood outflow into the cardiac chamber, coronary sinus, vena cava, pulmonary artery or pulmonary vein. Dual coronary artery-to-pulmonary artery fistulas have also been described as an uncommon congenital anomaly [1, 2]. Hemodynamic consequences and clinical presentation depend on the size of the fistula and shunt volume. Potential complications of coronary artery fistulas include bacterial endocarditis, rupture of the fistula, myocardial ischemia due to coronary steal phenomenon and pulmonary hypertension as well as congestive heart failure, if there is a large left-to-right shunt [3].

Case report

A 57-year-old man was referred to the Department of Cardiology because of exacerbation of exertional chest pain associated with shortness of breath. Pertinent medical history included well-controlled hypertension. Physical examination revealed a grade 3/6 mild systolic ejection murmur at the left sternal border. Resting electrocardiogram showed left ventricular hypertrophy and left anterior hemiblock. Chest X-ray was normal. Exercise treadmill test performed according to the Bruce protocol was terminated at the level of 6 METs due to fatigue and shortness of breath with ischemic changes in leads II, III and aVF. Two-dimensional echocardiogram disclosed hypokinesis of the anterior wall with a slightly reduced ejection fraction of 53%, Qp/Qs ratio of 1.3 and abnormal diastolic turbulent flow detectable in the pulmonary trunk (Figure 1). Transesophageal echocardiogram revealed normal flow and morphology of proximal coronary arteries and prominent left anterior descending artery. Tortuous, dilated vascular structure of oval shape (13 mm x 16 mm) with detectable flow was visualized in modified upper esophageal short-axis view in the area between the left atrial appendage, pulmonary artery and aorta. This finding was highly suggestive of cavernous coronary fistula with suspected intraluminal thrombus. Color Doppler flow mapping demonstrated flow from this abnormal vessel into the pulmonary artery 1 cm above the pulmonary valve. Coronary angiography confirmed a large, tortuous, dilated coronary artery fistula originating from the 1st diagonal branch (I Dg) of the left anterior descending artery with a single opening into the pulmonary trunk. The patient underwent surgical ligation of the fistula without extracorporeal circulation (Figure 2). The patient’s postoperative course was uncomplicated and on the 15th day the patient was discharged. However, at a follow-up visit after 3 months the patient again complained of exertional chest pain associated with shortness of breath – symptoms which occurred 2–3 weeks after the surgical treatment. Systolic murmur was also present and small diastolic, turbulent flow was detectable in the pulmonary trunk by transesophageal echocardiography.
Fig. 1. Two-dimensional transthoracic echocardiogram (parasternal short axis view) with color Doppler flow mapping demonstrates turbulent flow (arrow) entering the pulmonary trunk from the coronary artery fistula (CAF).

PT – pulmonary trunk, Ao – ascending aorta

Fig. 2. Surgical ligation of the fistula. A – Ao – ascending aorta, PT – pulmonary trunk, X – dilated coronary artery fistula (diagonal branch) to pulmonary trunk. B – Arrow indicates triple purse-string suture closing vessels of the fistula

Fig. 3. Coronary angiography. A – LAO 90 – lateral view of diagonal branch/left coronary to pulmonary artery fistula (arrows); B – RAO 25 CAU 25 view of diagonal to pulmonary artery fistula emptying into the pulmonary artery; C – LAO 90 – early post-stenting lateral view, 99% narrowing of feeding vessel PA – pulmonary artery
coronary arteriography. It is generally believed that a symptomatic fistula has to be closed, either by surgical epicardial or endocardial ligation, or by an intravascular method with coil embolization or occluder systems. All these interventional methods are considered to be relatively safe, with a low peri-operative morbidity and mortality [9]. However, Saito et al. reported that simple ligation of the fistula may lead to late recanalization [10]. The majority of fistulas may be initially addressed by percutaneous techniques such as coil obliteration. The largest series of transcatheter closures of coronary artery fistulas have been published by Armsby et al. [11] and Trehan et al. [12]. However, in patients with difficult anatomy (sharp angulation of a tortuous and narrow feeding vessel) surgical treatment may be preferred. Coil embolization of the fistula ought to be made distally, far from the vessel orifice, to prevent the closure of normal coronary arteries or migration of the coil.

In our patient, echocardiography provided an initial suspicion of coronary artery fistula which was confirmed using invasive methods. Because of symptomatic ischemia surgical ligation of the fistula was performed. Percutaneous access to the feeding vessel was difficult (sharp angle) and safe coil delivery was problematic. Moreover, large size of the feeding vessel was another factor favoring surgical treatment as the first option. However, the surgical treatment was not effective. A few weeks following the surgery our patient developed recurrence of angina probably caused by recanalization. Therefore, our second choice was to obliterate the orifice of the fistula by stent-graft implantation. This was proven successful by echocardiography and angiography.

In conclusion, as a result of increasing experience and improved devices and techniques, the transcatheter closure of coronary artery fistulas is emerging as a successful therapeutic strategy. However, the preferred approach for any individual patient depends on the anatomy of the fistula (the size and the location of the feeding arteries) and the presence or absence of associated defects.

**References**

1. Vijayvergiya R, Bhadauria PS, Jeevan H, et al. Myocardial ischemia secondary to dual coronary artery fistulas draining into main pulmonary artery. Int J Cardiol 2010; 140: e30–e33.
2. Papadopoulos DP, Perakis A, Votreas V, Anagnostopoulos S. Bilateral fistulas: a rare cause of chest pain. Case report with literature review. Hellenic J Cardiol 2008; 49: 111–113.
3. Moukarbel GV, Nasallah AT. Coronary artery fistula draining into the pulmonary artery. Int J Cardiol 2005; 99: 493–494.
4. Angelini P. Coronary to pulmonary fistulae. Tex Heart Inst J 2000; 27: 327–329.
5. Angelini P. Questions on coronary fistulae and microfistulae. Tex Heart Inst J 2005; 32: 53–55.
6. Lipiec P, Peruga JZ, Krzeminska-Pakula M, et al. Right coronary arterio-to-right ventricle fistula complicating percutaneous transluminal angioplasty: case report and review of the literature. J Am Soc Echocardiogr 2004; 17: 280–283.
7. Brussee H, Gasser R. Images in clinical medicine. Fistula connecting the left main coronary artery with the right atrium in a marathon runner. N Engl Med 2002; 346: 904.
8. Moukarbel GV, Nasallah AT. Coronary artery fistula draining into the pulmonary artery. Int J Cardiol 2005; 99: 493–494.
9. Androulakis A, Chrysohoou C, Barbetseas J, et al. Arteriovenous connection between the aorta and the coronary sinus through a giant fistulous right coronary artery. Hellenic J Cardiol 2008; 49: 48–51.

10. Saito A, Shiono M, Yamamoto T, et al. Surgical treatment for innominate artery aneurysm with a coronary pulmonary artery fistula: a case report. Ann Thorac Cardiovasc Surg 2005; 11: 55–58.

11. Armsby LR, Keane JF, Sherwood MC, et al. Management of coronary artery fistulae. Patient selection and results of transcatheter closure. J Am Coll Cardiol 2002; 39: 1026–1032.

12. Trehan V, Yusuf J, Mukhopadhyay S, et al. Transcatheter closure of coronary artery fistulas. Indian Heart J 2004; 56: 132–139.