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 × 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 trans-

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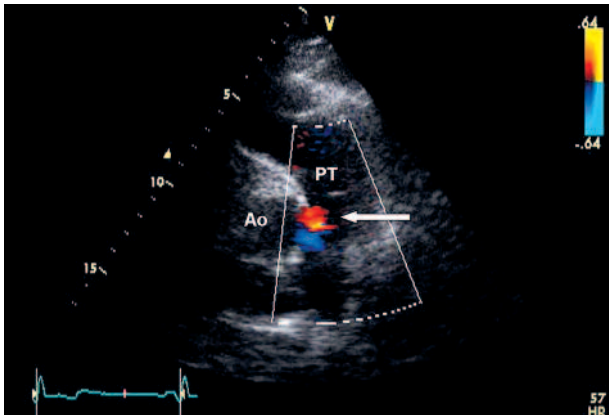

Fig. 1. Two-dimensional transthoracic echocardiogram (parasternal short axis view) with color Doppler flow mapping demonstrates turbulent flow (arrow) entering the pulmonary trunk from the coronary artery fistula (CAF)
PT – pulmonary trunk, Ao – ascending aorta

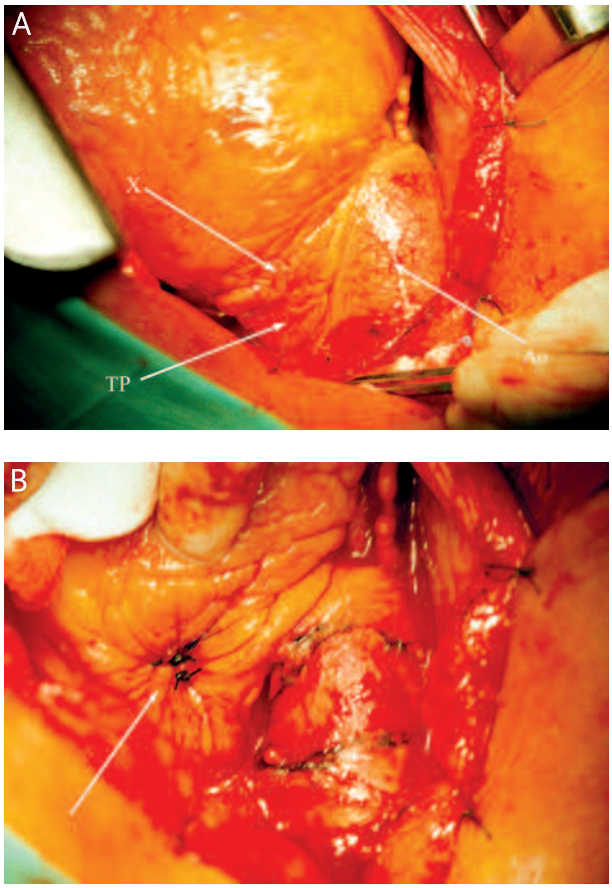

Fig. 2. Surgical ligation of the fistula. **A** – Ao – ascending aorta, PT – pulmonary trunk, X – dilated coronary artery fistula (diagonal branch) to pulmonary trunk. **B** – Arrow indicates triple purse-string suture closing vessels of the fistula

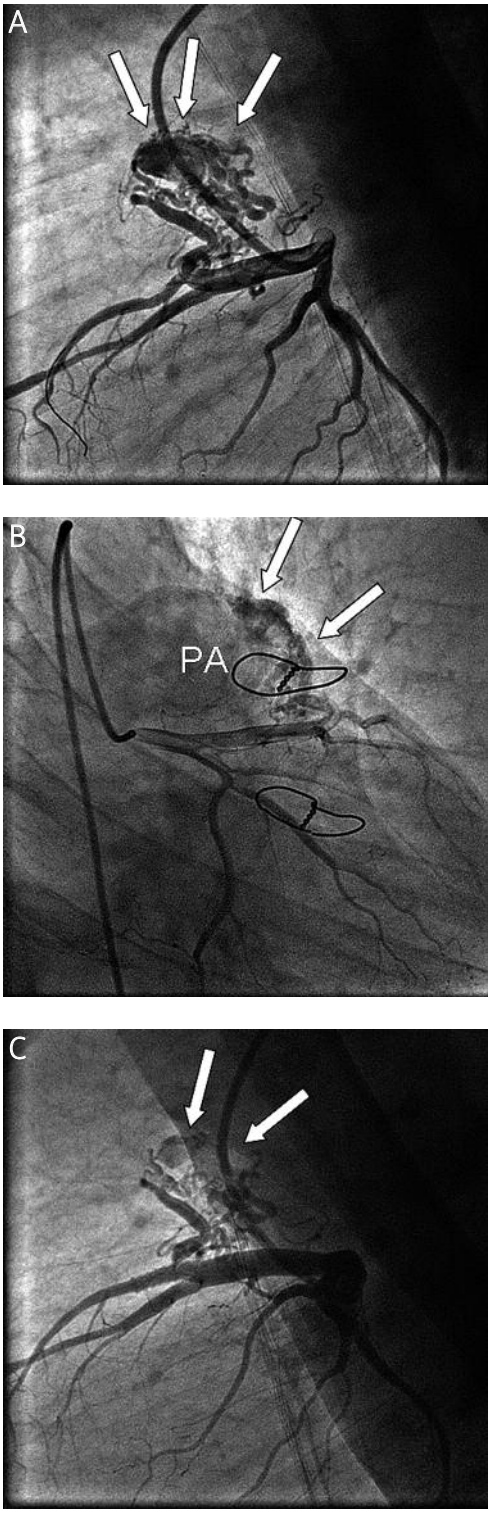

Fig. 3. Coronary angiography. **A** – LAO 90 – lateral view of diagonal branch/left coronary to pulmonary artery fistula (arrows); **B** – RAO 25 CAU 25 view of diagonal to pulmonary artery fistula emptying into the pulmonary artery; **C** – LAO 90 – early post-stenting lateral view, 99% narrowing of feeding vessel
PA – pulmonary artery

thoracic and transesophageal echocardiography. Nevertheless, the patient refused control coronary angiography and was stabilized on antianginal medication.

Four years after the surgery he was re-admitted to the Department of Cardiology because of exacerbation of anginal symptoms. Echocardiography and coronary angiography confirmed the presence of a coronary pulmonary fistula similar to the previously reported examination with significant shunt (Figure 3). The patient was then scheduled for transcatheter closure of the feeding vessel using a stent graft (Jostent GraftMaster 3.5 mm × 12 mm) which was implanted using femoral access into the I Dg artery, covering the orifice of the vessel feeding the fistula. Subtotal closure of the ostium was achieved with significant decrease of blood inflow into the fistula and minimal contrast passage into the pulmonary trunk. The postprocedural course was uncomplicated and the patient was discharged without any symptoms of angina or heart failure. The systolic murmur disappeared. One year after the procedure the patient was still asymptomatic and without cardiac murmur. The control echocardiographic study showed a minimal residual flow from the fistula into pulmonary artery flow with normal Qp/Qs.

Discussion

We report a case and echocardiographic presentation of a patient with typical angina due to a coronary artery-to-pulmonary artery fistula, with failed surgical treatment and a successful percutaneous approach.

A true fistula of the circulatory system is characterized by an ectatic vascular segment that exhibits continuous flow and connects two vascular territories governed by widely different hemodynamic environments with large pressure differences [4]. Coronary artery fistulas are uncommon congenital anomalies, present in 0.87% of coronary angiograms [5]. Coronary artery fistula can also occur as a complication of congenital heart surgery or following transcatheter interventions such as coronary angioplasty [6].

In 55% of cases congenital coronary fistulas originate from the right coronary artery or its branches; the left coronary artery is involved in 35% of cases, and both coronary arteries are involved in 5% of cases [7]. Typical drainage sites include the right ventricle and pulmonary trunk. 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 [8].

Diagnosis of fistulas is almost always incidental during 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 interventions are considered to be relatively safe, with a low perioperative 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 torturous 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.

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