

Successful repair of a coronary artery to pulmonary artery fistula with saccular artery aneurysm and critical stenosis of the left anterior descending coronary artery

Sandro Gelsomino, Giovanni Rubattu*, Pier Franco Terrosu, Luca Cossu*, Francesco Orrù*, Guglielmo Barbosa*

Department of Cardiovascular Sciences, S. Maria della Misericordia General Hospital, Udine, *Cardiac Surgery and Cardiology, Nuovo S. Annunziata Civic Hospital, Sassari, Italy

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We report an infrequent case of an adult patient with a coronary artery to pulmonary artery fistula associated with a coronary artery aneurysm, a critical stenosis of the left anterior descending coronary artery (LAD) and a LAD dissection located distally to the stenosis. The fistula was successfully closed with direct sutures by opening the aneurysm under complete cardiopulmonary bypass. The excess aneurysm wall was excised and aneurysmorrhaphy was performed. Closure of the distal opening of the fistula was carried out without pulmonary arteriotomy and the operation was completed with a coronary artery bypass graft on the LAD with the left internal mammary artery used as "free graft".

Postoperative angiographic evaluation demonstrated a normal artery distribution and the patient was asymptomatic without recurrence at 6 months after the operation.

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Address:

Dr. Sandro Gelsomino
Cardiologia
Azienda Ospedaliera
Careggi
Viale Morgagni, 85
50134 Firenze

Coronary arterio-venous fistula (CAF) is a relatively rare cardiac anomaly, characterized by a direct communication between a coronary artery having a normal origin from the aorta and a cardiac chamber or a large thoracic vessel¹. Its occurrence in patients undergoing selective coronary angiography ranges between 0.2 and 1%^{2,3}. The fistulous connection may indifferently enter any of the four cardiac chambers as well as the pulmonary artery, the coronary sinus or the superior vena cava². Nonetheless, most fistulas drain into the right cardiovascular structures (right ventricle 42.5%, right atrium 34%, pulmonary artery 15%)⁴. The presence of symptoms, complications and a significant left-to-right shunt are currently the main indications for CAF closure⁵ and the treatment of choice (transcatheter closure vs surgical therapy) is still subject of debate⁶. In addition, the management of asymptomatic patients with a non-significant shunt is still a matter of discussion⁵.

We report a successful surgical repair of a coronary artery to pulmonary artery fistula (CA-PAF) in a rare association with a coronary artery aneurysm and a critical stenosis of the left anterior descending coronary artery (LAD).

Case report

A 46-year-old woman was hospitalized on an emergency basis because of anginal-like chest pain with dyspnea and syncope. The patient's history did not include any episode of cardiac symptoms and cardiac auscultation on admission did not reveal the presence of any murmur. On physical examination she was conscious with a heart rate of 65 b/min and a systolic blood pressure of 110 mmHg. The chest X-ray was within normal limits. A continuous heart murmur (Levine 2/6) was audible at the third intercostal space. An electrocardiogram showed significant ST-T changes at rest (ST-segment depression and T-wave inversion) suggesting acute myocardial ischemia and a Q-wave in leads V₄-V₆, suggestive of myocardial infarction. Echocardiography showed a hypertrophic, non-dilated left ventricle, with hypokinesis of the apex of the heart and with a normal systolic function (left ventricular ejection fraction 46%). Cardiac catheterization studies confirmed the presence of a left-to-right shunt with a pulmonary-to-systemic flow ratio of 1.3:1. Selective left coronary angiography revealed the presence of a fis-

tula, which, from the LAD, entered the right heart through the pulmonary artery. A large sacciform coronary artery aneurysm originated from the fistula close to its origin. Furthermore, a critical stenosis of the LAD and an intimal flap were detected distally to the fistula (Fig. 1).

The heart was exposed through a median sternotomy. Using complete cardiopulmonary bypass, cardiac normothermic arrest was obtained by means of antegrade and retrograde blood cardioplegia. On opening the pericardium a coronary artery aneurysm, measuring 35×45 mm, was identified in the proximal segment of the LAD (Fig. 2).

The aneurysm was opened (Fig. 3) and the small orifice of the LAD was identified inside its proximal segment and closed using 5-0 monofilament sutures. An intimal rupture (flap) was detected distally to the aneurysm. The excess aneurysm wall was excised and

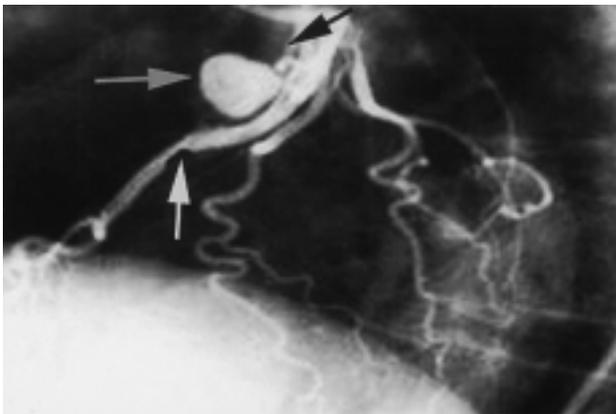


Figure 1. Selective left coronary angiography revealed the presence of a fistula that, from the left anterior descending coronary artery, entered the right heart through the pulmonary artery (black arrow). A large sacciform coronary artery aneurysm originated from the fistula close to its origin (gray arrow). Furthermore, a critical stenosis of the left anterior descending coronary artery was detected below the fistula (white arrow). Finally, an intimal rupture (flap) of the left anterior descending coronary artery was revealed distal to the stenosis.

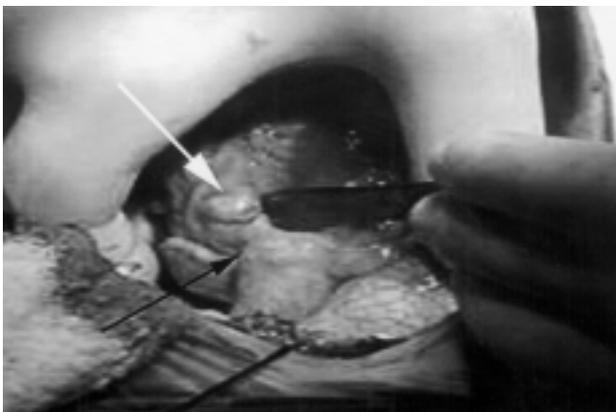


Figure 2. The coronary artery fistula with a saccular aneurysm, originating from the left anterior descending coronary artery (white arrow) and draining into the main pulmonary artery (black arrow).

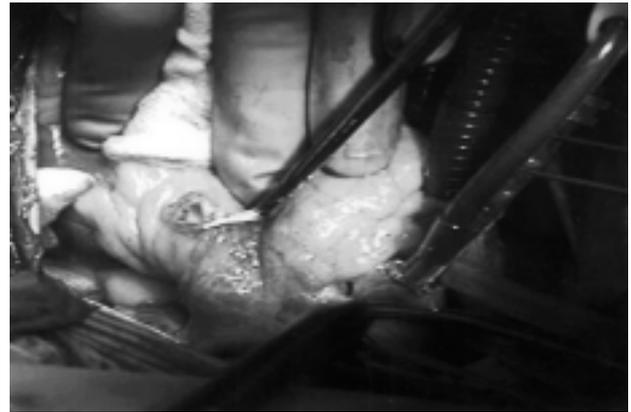


Figure 3. The coronary artery aneurysm was opened.

aneurysmorrhaphy was performed. Epicardial closure of the distal opening of the CA-PAF was performed using 5-0 monofilament sutures (Fig. 4). A distal anastomosis to a “free graft” left internal mammary artery was done in the LAD, distally to the dissection. The proximal anastomosis to the aorta was constructed while the heart was beating. Surgery was completed according to standard technique. The postoperative course was uneventful. At postoperative angiography the graft was patent and no aneurysm or abnormal communication related to the LAD was found, as confirmed by echocardiography. The patient was discharged on postoperative day 8. She was asymptomatic without recurrence at 6 months after the operation.

Discussion

CAF is a rare lesion and it was first described by Krause⁷ in 1865. Its etiology is most frequently congenital⁸; however, it may be secondary to cardiac trauma⁹, chest irradiation¹⁰, coronary angioplasty¹¹ or congenital heart surgery¹².



Figure 4. Epicardial closure of the distal opening of the fistula was performed using 5-0 monofilament sutures.

In 50-55% of cases fistulas originate from the right coronary artery or its branches; the left coronary artery is the involved vessel in about 35% of cases and in 55% both coronary arteries are the site of the CAF¹. In addition, more than 87% of fistulas open into the right heart chambers or their connecting vessels¹⁻⁴.

Most patients, especially those with small fistulas, are asymptomatic and the CAF is discovered casually during a coronary angiography performed for other reasons.

The most common symptoms include dyspnea, fatigue and chest pain or angina which, in the absence of a concomitant coronary artery disease, seems to be due to a "coronary steal phenomenon"¹³ secondary to an increased blood flow over the systemic to pulmonary fistula, which, in different studies, is reported to occur in 3-7% of cases¹⁴. In addition, intimal damage may occur because of an increased flow in the artery supplying the fistula⁵. Complications such as pulmonary artery hypertension, congestive heart failure, arrhythmias, bacterial endocarditis and CAF rupture with cardiac tamponade, have been rarely reported^{15,16}. Björck and Crafoord¹⁷ described the first successful surgical closure of a CAF in 1947. Later, Swan et al.¹⁸ reported the closure of a CAF using cardiopulmonary bypass in 1959. In more recent years, different techniques have been proposed: selective external CAF ligation without cardiopulmonary bypass (epicardial approach), direct suture of the fistulous ostium from within the involved heart chamber (endocardial approach), coronary ligation, Symbas's technique and coronary artery bypass^{11,19,20}. Furthermore, since 1983, transcatheter closure of the CAF has been advocated as a minimally invasive alternative to surgery²¹ and different methods of transcatheter closure have been proposed. These include detachable balloons, stainless steel and platinum coils, the Amplatz occluder, the patent ductus arteriosus umbrella, alcohol or foam injection and covered stents, the latter reserved for those cases of coronary fistulas associated with coronary stenosis. In a recent study Armsby et al.⁶, comparing their results with those reported in the literature, found transcatheter and surgical closure to have a similar early effectiveness, morbidity and mortality.

We report a successful closure of a CA-PAF in association with a coronary artery aneurysm, a critical stenosis of the LAD and an LAD dissection located distally to the stenosis. The association of a CAF with a coronary artery aneurysm is infrequent⁶ and its rupture is rare². The concomitance of a CAF and coronary atherosclerosis has been previously described²² and the location of the stenosis may play an important pathophysiological role; in fact, in case of a fistula distal to the stenosis, a right-to-left shunt occurs, the entity of which depends on the drop in pressure distal to the lesion. In this case the shunt may work as a bypass thus protecting the myocardium from infarction²³.

The fistula was closed by means of a direct suture under complete cardiopulmonary bypass. The excess aneurysm wall was excised and aneurysmorrhaphy was

performed. Suture of the distal opening of the fistula was performed without pulmonary arteriotomy and the operation was completed with a coronary artery bypass graft on the LAD using the left internal mammary artery as "free graft". The anastomosis was performed distally to the intimal flap. The operation was successful, the postoperative course was uneventful and at 6 months of follow-up the patient was well.

Regardless of the method of repair chosen, its main purpose is closure of the shunt with the aim of maintaining an adequate coronary blood flow and preventing complications. The presence of symptoms (ischemia, ECG changes), complications (congestive heart failure, pulmonary hypertension) and a significant left-to-right shunt (> 30%) are currently the main indications for CAF closure²⁴. In asymptomatic patients the indication is still debated; some authors suggest an early closure because of the progressive nature of the lesion, its possible complications and the low mortality rate of both the surgical and percutaneous techniques^{6,25}. For others, instead, in asymptomatic patients with a non-significant shunt the indication for CAF closure is not clear²⁴; in addition, spontaneous closure of the CAF, although infrequent, has been described in the literature²⁶.

In our opinion, the method of closure to be chosen depends on several factors: the anatomy of the fistula (which should be promptly assessed at angiography), its pathological presentation, the presence of concomitant cardiac defects and/or coronary atherosclerosis, the expertise of the cardiologists and surgeons. We believe that surgery should be preferred in case of concomitant significant coronary artery disease or when the CAF is associated with a complex heart disease requiring surgery. Furthermore, surgery is also advisable when the anatomic characteristics of the fistula make transcatheter closure inappropriate (i.e. complex anatomy, distal location, large fistula, adjacent vessel at risk). Finally, when the CAF is associated with a coronary artery aneurysm, its closure associated with aneurysm ligation is recommended, being the risk of surgery in this case, less than the potential development of fatal complications after transcatheter closure (i.e. perforation, mobilization of thrombi). Conversely, in older patients, in subjects with a proximal fistula or in the absence of concomitant cardiac diseases requiring surgery, the different methods of transcatheter closure may be employed.

In conclusion, the surgical option is indeed indicated whenever the percutaneous procedure is unfeasible or not contraindicated.

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