Coronary artery fistulas: symptoms may not correlate to size. An emblematic case and literature review

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Introduction

Fistulous communication of coronary with pulmonary arteries in the adults is a common type of coronary artery fistula (CAF)[1–3]. In most reported cases, the fistula usually arises from the proximal left and/or right coronary arteries via the anterior conal branches that connect to the anterior wall of the main pulmonary artery. These anteriorly located abnormal communications are usually asymptomatic and are incidentally found during an angiography of the coronary arteries in 0.2–0.3% of the exams[4].

Case report

We are here presenting a case of a 73-year-old man, history of effort angina from youth, poor controlled blood pressure and family history of coronary artery disease. No history of previous significant chest wall trauma nor any invasive cardiac procedures were reported. The patient did not receive previous investigations for his angina. His medication consisted of acetilsalicilate, Ca-antagonist, angiotensin-converting-enzyme inhibitor, and digoxin for permanent atrial fibrillation.

The patient was admitted to Cardiac Surgery Unit for ascending aorta aneurysm, diagnosed accidentally. At admission the patient was asymptomatic and in good physical condition, with a blood pressure of 140/80 mmHg and a heart rate of 70 beats/min. A diastolic murmur related to aortic regurgitation was heard on cardiac auscultation. The electrocardiogram (ECG) showed atrial fibrillation at 70 bpm. Trans Thoracic Echocardiography (TTE) showed a good left ventricular global and segmental function with left ventricular ejection fraction (EF) of 60%, severe aortic regurgitation, and dilation of the aortic root (55 cm) and ascending aorta (56 cm).

Coronary angiography showed no significant coronary atherosclerotic lesions, left coronary artery ectasia (Figure 1A) and reveled a coronary artery fistula arising from proximal right coronary artery (RCA) and draining to main pulmonary artery (Figure 1 B).

According to the symptoms, despite not significant shunt entity at rest, we decided to close the fistula during ascending aorta replacement through Bentall-De Bono operation in theatre. During the isolation of the right sinus a vessel enriched from the right coronary artery to the main pulmonary artery was ligated (Figure 1C, D, E). The intraoperative course was uneventful and transesoephageal echocardiography showed no changes to the preoperative status. The patient was discharged after ten days and was asymptomatic for angina or dispnea without complications.

At one-year clinical follow-up the patient is still asymptomatic for effort angina with improved quality of life.

Discussion and literature review

Although coronary artery fistulas are infrequent, they are becoming increasingly important, as their management and treatment could prevent serious complications[5]. They represent the most common hemodynamically significant congenital defect of the coronary arteries, including 13-14% of angiographically found coronary artery anomalies[3,4,6].

We can distinguish types of coronary artery fistulas associated with other congenital cardiac malformations (eg. pulmonary atresia) or isolated forms[7,8]. The acquired forms may be secondary to infectious processes, dissecting thoracic aorta or traumatic or iatrogenic mechanism[9,10]. The majority of these fistulas arise from the right coronary artery (52-55% of cases) or the left anterior descending coronary artery (30-35% of cases), while the circumflex coronary artery is less frequently involved (5-18% of cases)[2,3,10,11]. Over 90% of the fistulas drain to low pressure structures, such as right-sided chambers (right ventricle 41%, right atrium 26%), pulmonary artery (17%), superior vena cava, and coronary sinus (7%)[6,11].

The clinical presentation depends on the severity of the left-to-right shunt and the majority of adult patients are usually asymptomatic[10], so that their real incidence is difficult to discern

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but coronary steal could induce ischemia in patients without atherosclerotic coronary artery disease[5].

Clinical presentations include dyspnea, angina, endocarditis, arrhythmias, stroke, myocardial ischemia or myocardial infarction. Myocardial ischemia/infarction can occur from decreased coronary blood flow distal to the fistula and has been documented in patients with coronary fistulas with no evidence of coronary atherosclerosis. They often assure a blood shunt of small magnitude which doesn't compromise myocardial blood flow. The majority of adult patients are usually asymptomatic. Unlike adults, a smaller percentage of pediatric patients tend to be asymptomatic[12,13]. Nevertheless, pediatric patients are usually identified due to electrocardiographic or chest X-ray abnormalities or for evaluation of a loud, superficial continuous cardiac murmur.

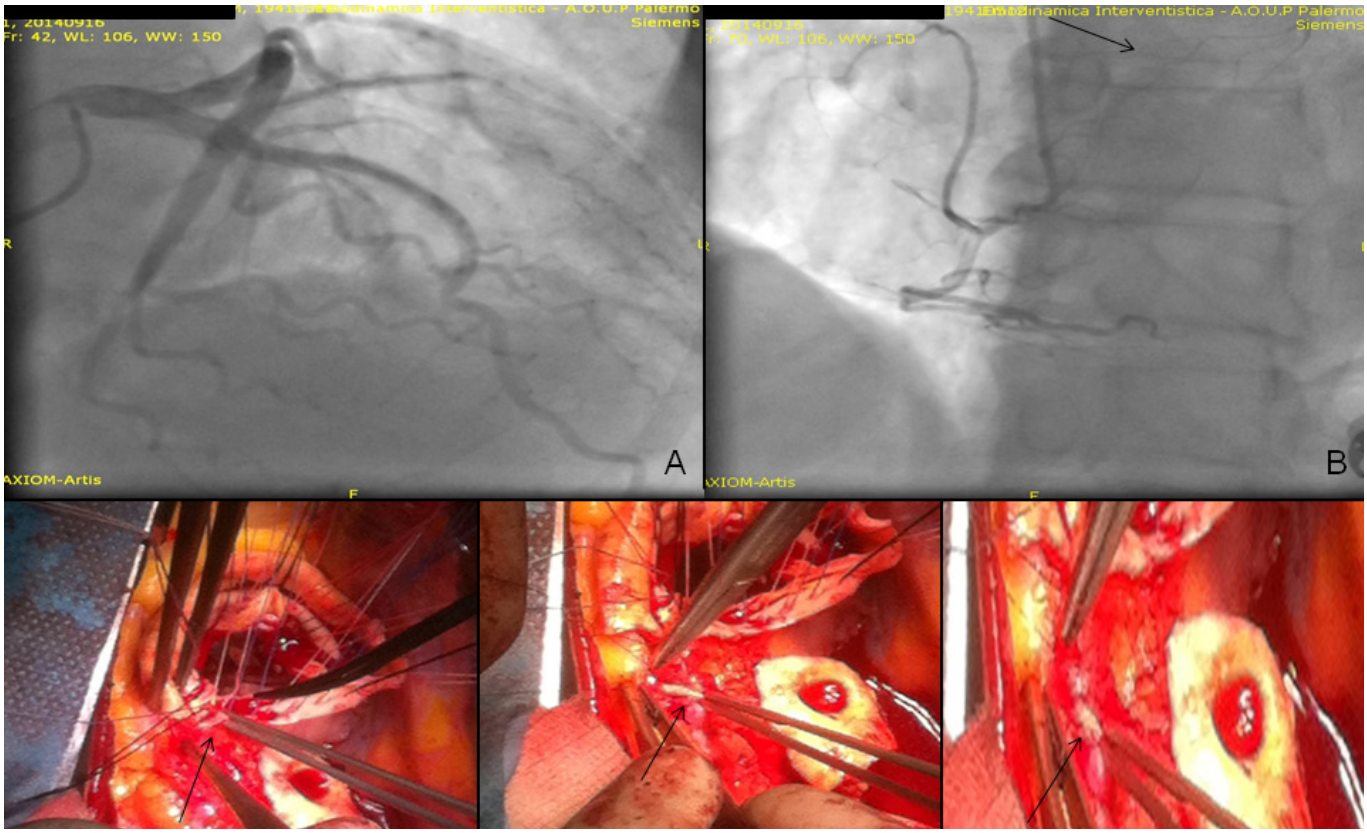
A continuous cardiac murmur may be the only clinical finding[6,10,14]. A presumptive diagnosis can occasionally be made upon hearing an atypical systolic, diastolic, or continuous murmur. In CAF the continuous murmur tends to be crescendo-decrescendo in both systole and diastole, but louder in diastole, in contrast, most of the other continuous murmurs reach their peak intensity at the time of the second sound. The place where the continuous murmur is loudest depends on where fistula enters in the heart. With entry into the right atrium the murmur is loudest along the sternal border, with entry into pulmonary artery it is loudest near the second intercostal space to the left of sternum, and with entry into the left ventricle the murmur is loudest near the apex.
Although diagnosis of coronary artery fistulas is based on angiography, non-invasive cardiac imaging techniques, such as 2D and color Doppler echocardiography, cardio-MRI and cardiac CT may be helpful[10,15,16].

Several angiographic classifications have been proposed. Coronary fistulas can be categorized into two different anatomical types: fistulas with one large channel or one or more small channels; fistulas composed of a plexiform network of vessels[5,17].

Congenital coronary to pulmonary fistula are rare. Their incidence ranges from 0.3 to 0.8% of patients who undergo a coronarography and the vast majority is asymptomatic. Their natural course is to develop with time, however a few case of spontaneous closure have been described. They usually become symptomatic on the 5th or 6th decade with sudden death, myocardial ischemia, pulmonary artery systolic hypertension, heart failure, arrhythmia, rupture or endocarditis[18,19].

Concerning the treatment, surgical or catheter-based closure is strongly recommended in symptomatic patients and in high flow-shunting asymptomatic patients, especially in pediatric; while there are no clear indications on treatment of asymptomatic adult patients with non-significant shunting[10]. When they become symptomatic, treatment is mandatory to prevent complications. The appearance of these symptoms depends on the size of the communication, the amount of blood that is drained and the resistance of the chamber into which the fistula drains. Potential complications in the presence of large left-to-right shunt are pulmonary hypertension and congestive heart failure; others include rupture or thrombosis of the fistula or associated arterial aneurysm or coronary steal phenomena[19].

Figure 1
A: Coronary angiography shows ectatic left coronary artery.
B: Coronary angiography shows coronary artery fistula (arrows) arises from RCA draining in pulmonary artery.
C, D, E: images during surgery confirm that a little coronary artery fistula (arrow) arising from RCA and draining to pulmonary trunk.



The guidelines of the '"American College of Cardiology/ American Heart Association" (ACC/AHA) indicate as Class I recommendation the percutaneous or surgical closure for large fistulas regardless of symptoms, while for small or medium size only in cases of myocardial ischemia, arrhythmia, ventricular dysfunction, ventricular dilatation or endarteritis[20]. Until a few years ago, the surgical treatment of coronary artery fistulas was the only option[21,22]. Since the 90s a possible alternative to surgical closure of the fistula is percutaneous management with the use of coils that promote thrombosis (coil embolization) or stents, but only in cases with favorable anatomy (e.g. origin vessel, tortuosity, caliber of the lumen, uniqueness of fistula, drainage site, etc.)[23, 24]. In the last years ithas been proposed the use of the "Amplatzer Duct Occluder" and "Amplatzer Septal Occluder" (AGA Medical Corporation, Golden Valley, MN), a braided-nitinol device for transcatheter patent ductus arteriosus (PDA) closure[25]. Surgery is still the first choice in case of cardiac malformations but coronary angiography is needed to determine the best therapeutic choice[24]. Medical therapy with anti-anginal drugs may improve the clinical and symptoms. Strongly recommended are antiplatelet agents and prophylaxis for bacterial endocarditis[10, 26, 27].

Conclusion

Coronary artery fistula should be considered and investigated in patients in which there are not clear causes of effort angina, especially in young patients. According to current opinion a symptomatic fistula should be closed, regardless of the size, with percutaneous coronary intervention or by surgery considering also if patient needs cardiac surgery for aortic valve replacement, as this case, or other congenital heart disease.

Statement of ethical publishing

The authors state that they abide by the statement of ethical publishing of the International Cardiovascular Forum Journal[28].

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The authors declare that there is no conflict of interest.

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