

A coronary fistula diagnosed in the eighth decade of life: The utility of non-invasive methods in the selection of treatment approach

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Abstract

A 77-year-old woman was referred to our Department of Cardiology because of exacerbation of chest pain and decreased exercise intolerance. No acute ischemic electrocardiography changes were seen in an electrocardiogram recorded on admission. An exercise test was terminated at 7 METS because of shortness of breath without evidence of ischemia. The patient was referred for a coronary angiography which showed a coronary artery fistula filling from the left anterior descending (LAD) artery and resulting in a large inflow to the main pulmonary artery, without other significant coronary lesions. Transthoracic echocardiography showed a coronary artery fistula draining to the main pulmonary artery. Coronary steal was suspected and coronary flow reserve was evaluated in LAD, showing normal values for age. Due to the overall clinical picture, with the predominance of heart failure symptoms and the lack of significant abnormalities of flow reserve in LAD, medical therapy was selected. The patient remained free from cardiovascular symptoms at 6-month follow-up. (Cardiol J 2010; 17, 3: 299–302)

Key words: coronary fistula, ischemic heart disease, coronary reserve

Introduction

Coronary fistulae are congenital or acquired connections between coronary vessels and cardiac chambers or other vascular structures, such as vena cava, pulmonary artery, or pulmonary veins, resulting in blood by-passing the coronary bed. In 9% of cases, clinical symptoms due to a coronary fistula manifest before 20 years of age, and 19% of these are cardiovascular events due to the presence of a pathological left-to-right shunt [1]. With

age, the number of asymptomatic cases decreases and subjective complaints are reported by about 55% of patients older than 20 years. Complications of the fistula, including heart failure, pulmonary hypertension, coronary ischemia or even myocardial infarction, are seen in 63% of patients [1]. We present a case of a 77-year-old woman with typical angina and severe exercise intolerance who was found to have a fistula between the left anterior descending (LAD) artery and the pulmonary artery.

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Figure 1. Angiography of the left coronary artery (CAU 30, RAO 30) showing the lumen of a fistula (arrow) between the segment 6 of the left anterior descending artery and the pulmonary artery.

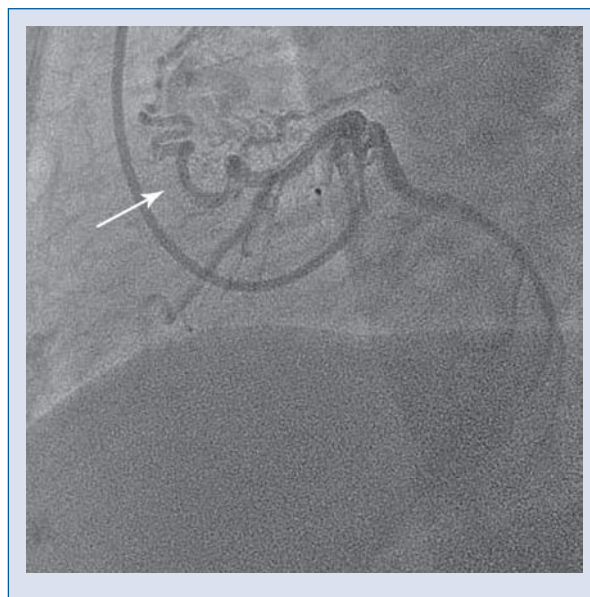


Figure 2. Angiography of the left coronary artery (LAO 90), with the visible lumen of a fistula (arrow) between the proximal left anterior descending artery and the pulmonary artery.

Case description

A 77-year-old woman with a history of smoking for 30 years was admitted to our Department of Cardiology due to exacerbation of retrosternal chest pain and decreased exercise intolerance, consistent with the Canadian Cardiovascular Society (CCS) class II/III symptoms.

Her past medical history included hypertension treated medically for 25 years, hypercholesterolemia (total cholesterol 238 mg/dL, LDL cholesterol 150 mg/dL), left-sided hydronephrosis, hypothyroidism treated with thyroid hormone supplementation (recent TSH level was 7.22 μ U/mL), and essential thrombocytosis with the platelet count of 754,000/ mm^3 . Admission electrocardiogram showed normal sinus rhythm of 70 bpm and left anterior hemiblock, with no evidence of acute myocardial ischemia. Treadmill exercise test was terminated at the workload of 7 METS due to dyspnea without typical angina. Heart rate at the peak workload was 117 bpm, corresponding to 80% of the age-predicted maximum heart rate (the exercise test was performed while the patient continued to take the previously prescribed beta-adrenergic blocker). No ST segment changes were seen suggestive of myocardial ischemia. The patient was then referred for a coronary angiography which showed a coronary artery fistula (Fig. 1, 2) filling from LAD, with a diameter larger than that of

LAD itself, resulting in a large inflow of the contrast agent to the main pulmonary artery, and a visual suggestion of impaired filling of LAD. In addition, atherosclerotic plaques were seen in the proximal segment of the right coronary artery but none of these lesions resulted in significantly impaired perfusion of the coronary bed.

Transthoracic echocardiography showed normal left ventricular systolic function, with left ventricular ejection fraction of 66%, mild enlargement of both atria, and a mild, hemodynamically insignificant aortic insufficiency. A diastolic flow with peak velocity 0.8 m/s was recorded in the pulmonary artery (Figs. 3, 4), consistent with a coronary fistula draining to the pulmonary artery and a hemodynamically insignificant shunt (no increase in pulmonary to systemic flow ratio [Qp/Qs] was seen).

Due to an angiographic suggestion of LAD ischemia related to a coronary steal resulting from blood shunting to the pulmonary artery, coronary flow reserve was evaluated in LAD using transthoracic echocardiography (Fig. 5). After intravenous administration of 12 mg of adenosine, an increase in maximal diastolic blood velocity from 22 to 55 cm/s was recorded in LAD, corresponding to a coronary reserve of 2.5 and suggesting no significant coronary steal from LAD through the fistula. Due to the overall clinical picture, with the predominance of heart failure symptoms and the lack of sig-

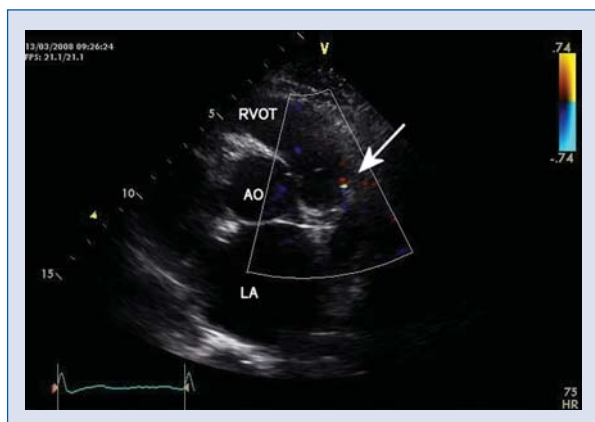


Figure 3. Transthoracic echocardiography, modified parasternal short-axis view with color Doppler flow mapping: a turbulent flow is seen in the pulmonary artery just over the pulmonary valve (arrow); Ao — ascending aorta; LA — left atrium; RVOT — right ventricular outflow tract.

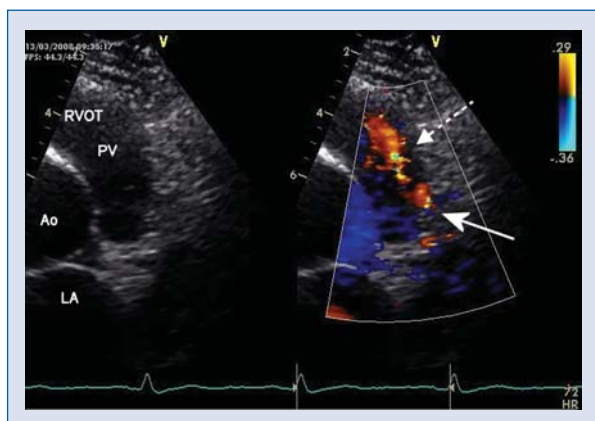


Figure 4. Transthoracic echocardiography, modified parasternal short-axis view: the orifice of the fistula is seen with a turbulent inflow to the pulmonary artery (solid arrow) and a pulmonary regurgitation (dotted arrow); Ao — ascending aorta; LA — left atrium; PV — pulmonary valve; RVOT — right ventricular outflow tract.

nificant abnormalities of flow reserve in LAD, medical therapy was considered appropriate for the patient. She was prescribed acetylsalicylic acid (150 mg/day), amlodipine (5 mg/day), levothyroxine (50 µg/day), metoprolol (12.5 mg bid) and indapamide (1.5 mg/day), and discharged home in a good clinical condition. The patient remained stable and free from worsening of cardiovascular symptoms during a 6-month follow-up.

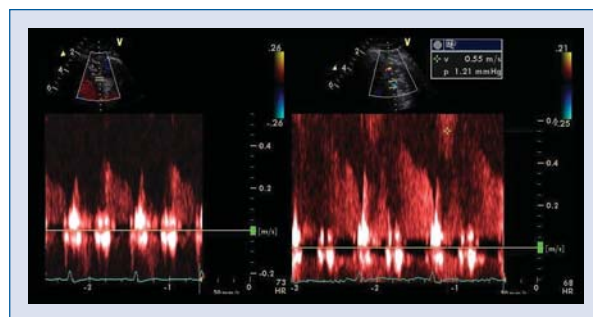


Figure 5. Transthoracic echocardiography, blood flow velocity measurement in the distal segment of the left anterior descending artery using pulsed wave Doppler at baseline (left panel) and following adenosine administration (right panel): 2.5-fold increase in blood flow velocity from 22 to 55 cm/s was recorded.

Discussion

Coronary fistulae are a form of congenital anomaly of coronary arteries or, more rarely, are acquired conditions, usually iatrogenic (e.g. developing as complications of coronary angioplasty or myocardial biopsy). Congenital coronary fistulae were seen in about 0.1–0.2% patients undergoing selective coronary angioplasty [2, 3].

In more than 50% of cases, coronary fistulae are abnormal communications of branches of the right coronary artery, and the remaining arise from branches of the left coronary artery. A communication between coronary arteries and the right ventricle was noted in 41% of cases, with the right atrium in 26% of cases, with the pulmonary artery in 17% of cases, with the left ventricle in 3% of cases, and with a vena cava in 1% of cases [1].

In 1947, Bjork and Crafoord performed the first successful closure of a fistula and the technique introduced by these authors remained the most commonly used one until 1983, when the first percutaneous closure was performed [2]. Percutaneous closure using vascular stents, coils, and occluders is associated with a lower procedural risk and thus became the preferred treatment method [4].

The clinical picture of a coronary fistula is largely related to the location and size of an abnormal communication. If the shunt is significant, pulmonary hypertension, heart failure, bacterial endocarditis, or myocardial ischemia due to coronary steal may ensue. “Childhood” type fistulae, usually of a large diameter and leading to a significant

dilatation of the coronary artery, diagnosed by the presence of a murmur or symptoms typical for a shunt lesion, should be differentiated from “incidental”, small fistulae, usually draining to the pulmonary artery. With the widespread use of coronary angiography and high sensitivity of transthoracic doppler echocardiography, the latter are currently more often diagnosed as an unexpected finding, commonly in adults undergoing routine coronary angiography. Blood flow recorded in these small fistulae is usually diastolic in nature, with a velocity below 1 m/s (unless the orifice of the fistula is small and restrictive), and must be distinguished from a similar spectrum of pulmonic regurgitation, although the velocity of the latter flow is usually higher. In this regard, it is important to define precisely the anatomic localization of the fistula as related to pulmonary valve leaflets in the short-axis parasternal view.

It should be stressed that a diagnosis of a coronary fistula in an adult patient is often a therapeutic dilemma, as the functional consequences of such lesions may be difficult to predict. Our case illustrates a practical approach to the evaluation of coronary fistula and selection of an appropriate treatment in a female patient with a coronary fistula diagnosed at the age of 77 years. In this particular patient, we used echocardiography for noninvasive visualization of the fistula and evaluation of relevant coronary artery flow reserve. These findings, showing lack of a significant impairment of coronary flow reserve, resulted in a decision to choose medical treatment.

Visualization of coronary fistulae using transthoracic echocardiography in adult patients has been reported previously [5]. However, the evaluation of coronary reserve using transthoracic echocardiography to assess physiology of a coronary fistula has been mostly described in the pediatric literature (e.g. in patients with Kawasaki disease), while myocardial perfusion scintigraphy, intracardiac ultrasonography, multidetector computed tomography, or even magnetic resonance imaging are more commonly used diagnostic modalities in the adults.

Although coronary angiography remains the preferred diagnostic tool to evaluate the anatomy of coronary fistulae, this invasive procedure carries an increased risk and thus noninvasive imaging

using magnetic resonance, multidetector computed tomography, and echocardiography should be preferentially used for further diagnostic and follow-up evaluations [6]. Echocardiographic evaluation, including assessment of coronary flow reserve, not only delineates the anatomy of the fistula but is also helpful in the determination of its functional significance. This may be assessed using information regarding blood flow through the fistula, Qp/Qs, and the possible contribution of the fistula in anginal symptoms due to coronary steal, giving important diagnostic clues in the evaluation of the risk of complications and the choice of the optimal treatment approach (surgical or percutaneous closure of large, hemodynamically significant fistulae, or medical treatment of nonsignificant lesions).

In summary, our case highlights the role of the evaluation of coronary flow reserve in such patients using transthoracic echocardiography, a diagnostic tool that is widely available in routine clinical practice.

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