Imaging Approach for Coronary-Cameral Fistula: A Relatively Rare Anomaly That May Be Accidentally Detected

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Abstract

Coronary-cameral fistulae (CCFs) are rare anomalies often detected incidentally during angiography or cardiac surgery (valve replacement or coronary artery bypass grafting). They represent about 0.2% to 0.4% of all cardiac malformations and 14% of all coronary anomalies. Here we present a brief review of various diagnostic approaches to CCFs.

Keywords: Coronary Artery Fistula (CAF), Imaging, Diagnosis

1. Background

Coronary-cameral fistulae (CCFs) are rare anomalies that are often picked up incidentally during angiography or such cardiac surgeries as valve replacement and coronary artery bypass grafting (1). CCFs represent approximately 0.2% to 0.4% of all cardiac malformations (2) and 14% of all coronary anomalies (3).

Generally isolated (80%), CCFs may also be associated with other congenital cardiac malformations (20%) such as patent ductus arteriosi, atrial septal defects, ventricular septal defects, and tetralogy of Fallot (4). CCFs can be single or multiple; they mostly originate from a coronary vessel and subsequently enter cardiac chambers or large vessels such as venae cavae, pulmonary arteries, and veins.

The origin of CCFs seems to be related to an aberration in normal embryonic development (5). These fistulae, especially if small, do not usually cause symptoms in the first 2 decades of life but over time, both the symptoms and any related complications tend to increase (6). Complications include embolism, heart failure, atrial fibrillation, rupture, endocarditis/endarteritis, and new-onset arrhythmias (7). A particular complication is a thrombosis within the fistula, which is albeit rare capable of causing acute myocardial infarctions, paroxysms of atrial fibrillation, or ventricular arrhythmias (8).

We read with interest a clinical case of an 82-year-old woman with a coronary fistula between the main left coronary artery and the pulmonary artery, bringing about spontaneous rupture and resultant massive hemopericardium and cardiac tamponade other rare but possible complications (9).

This underscores the importance of a precise and accurate diagnosis of these rare vascular anomalies. The main differential diagnosis is patent ductus arteriosi; it is, however, necessary to exclude other types of congenital arteriovenous shunts. In this scenario, coronary angiography is a precious diagnostic tool for the delineation of the coronary anatomy.

Some fistulae, especially if small, can close spontaneously. Nonetheless, once surgery with simple ligation of the fistula was the traditional method of treatment. Today, the gold standard is catheter closure with multiple types of devices (coils, etc.), which confers excellent results and precludes the consequence of cardiac surgery. Surgical or catheter-based closure is strongly recommended in symptomatic patients and in asymptomatic patients with high-flow shunting (not least in pediatric patients), while there are no clear indications of treatment of asymptomatic adult patients with insignificant shunting. When the patients become symptomatic, treatment is mandatory so as to prevent complications. The appearance of these symptoms depends on the size of the communication, amount of the blood that is drained, and resistance of the chamber into which the fistula drains (10).

Selecting an accurate strategy for the diagnostic imaging of CCFs remains a major challenge to the cardiologist. Indeed, the purpose of this manuscript is to propose an ap-
proach aimed at the early identification of the problem. Intrigued by the clinical case presented by Pouraliakbar et al. (11) concerning a young woman of 29 years, who presented with palpitations, shortness of breath on exertion, and atypical chest pain suggesting adult congenital anomalies, we decided to do a literature review to understand how individual clinical cases on CCFs (6, 12) have been addressed and what would constitute the most effective strategy for diagnostic imaging.

2. Clinical Features

Young patients (20 - 45 y) that do not have cardiovascular risk factors (smoking, diabetes mellitus, family history, dyslipidemia, sedentary lifestyle, and peripheral arterial disease) are likely to present with the following features:

- Vague symptoms: atypical chest pain, dyspnea on exertion, or palpitations
- Heart rate > 60 bpm
- Atypical systolic, diastolic, or continuous murmur (The continuous murmur tends to be crescendo-decrescendo in both systole and diastole, but louder in diastole.)

3. ECG

ECG may show volume overload, right or left ventricular hypertrophy, arrhythmias, or typical myocardial ischemia patterns. ECG is undoubtedly the 1st instrumental examination to be performed in patients with chest pain.

4. Chest X-ray

Chest X-ray is justified by the presence of dyspnea and is undertaken to exclude pulmonary disease or cardiomegaly.

5. Echocardiography

Echocardiography may show enlargement in the left or right chambers (often in the absence of valve disease) and defects in segmentary or global function. It may illustrate an abnormal flow with a tortuous course (e.g., from the left Valsalva sinus to the right atrium).

6. Transesophageal Echocardiography (TEE)

TEE is a valuable method in that it can obtain information about the origin of the fistula, draining points of the fistula, quantification of valve regurgitation, and efficacy of the eventual obliteration of the CCF. The coronary vessel involved will be dilated with an unusual and turbulent flow.

7. Multidetector Computed Tomography (MDCT)

MDCT is superior to echocardiography in overweight patients inasmuch as it confers excellent anatomical delineation by comparison with echocardiography. MDCT is capable of determining the presence or absence of obstruction and, thus, the likelihood of a coronary steal presentation. A contrast opacification into the receiving chamber/vessel is useful in confirming the CCF entry site and the patency of the shunt (15).

MDCT should be considered in case of negative or inconclusive TEE and treadmill tests, whereas coronary angiography is recommended when based on the previous examinations there is a high suspicion of CCF.

8. Coronary Angiography

Angiography is the main diagnostic technique for the precise diagnosis of CCFs. Cardiac catheterization provides the hemodynamic evaluation of the fistula and remains the modality of choice for defining coronary artery patterns for structure and flow. Moreover, it allows to program interventional closure with dedicated devices and provides the most detailed anatomy of the fistula yielding information about the size, course, origin, presence of any stenosis, and drainage site. Additionally, it helps to exclude various anomalies and defects, including but not limited to patent ductus arteriosi, ventricular septal defects with aortic incompetence, and arteriovenous fistulae in the lungs or chest wall. Furthermore, the exercise test and myocardial perfusion scan can be useful tools in the assessment of myocardial ischemia related to CCFs (14).

9. Magnetic Resonance Imaging (MRI)

MRI is an additional workup that may assist in the confirmation of the diagnosis. It may reveal the detailed anatomy of CCFs and can be used as an adjuvant noninvasive method in their investigation (15). However, its greatest limitation is the determination of the distal coronary course. As a result, this technique is less helpful in evaluating fistulae, coronary origins outside the normal sinuses,
and collateral vessels. Furthermore, visualization of the posterior descending branch is not easy. The goal of an efficient diagnostic process must be to arrive at a diagnosis quickly, avoiding also the waste of economic resources. In addition, only a correct and accurate diagnosis can direct toward the best therapeutic procedure for the patient. It is, therefore, necessary to opt for a diagnostic procedure so as to minimize the complications related to a late diagnosis especially of a clinical situation so misunderstood and rare as CCFs.

References


