Symptomatic bilateral coronary artery fistula to pulmonary artery in elderly patient

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ABSTRACT

We report the imaging findings of an uncommon coronary vascular termination anomaly, with fistula to the pulmonary artery.

This 70 year old female patient presented unstable angina, showing a coronary artery fistula depicted in coronary angiogram from the left coronary to the pulmonary artery, with no significant atherosclerotic pathology.

Due to development of ventricular tachycardia in stress echocardiogram examination, she was proposed for coronary fistula closure.

Coronary CT was performed for procedure planning and allowed the identification of a second unsuspected fistula from the right coronary to the right pulmonary artery.

Congenital coronary anomalies are a possible cause of symptomatic coronary pathology in patients of any age. In older patients, coronary artery fistulas are rare, especially when symptomatic. Adequately performed CT examinations, using its post processing capabilities, with 3D and MIP reconstructions are invaluable in delineating coronary anatomy, essential for further treatment planning.

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1. Introduction

Coronary anomalies are found in about 1% of routine autopsy examinations, and in 4%–15% of young individuals who experience sudden death [1]. These may be systematized as abnormalities of number, origin, course, termination, or structure of the coronary arteries. A coronary artery fistula (CAF) is classified as an abnormality in the termination of a coronary artery, ending in any of the four heart chambers or any of the large blood vessels (arteries or veins). Most have congenital origin. Other cases are related to trauma, infection, or iatrogenic damage [2].

The embryological basis of coronary-pulmonary fistula is explained by Hackensellner’s inversion-persistence hypothesis. This hypothesis states that among the six branches of the truncus, only two branches starting in the aortic sinus continue to form coronary artery, and the rest involute. When some of this branches of the pulmonary sinus do not involute and remains connected to a branch from the aortic sinus, a fistula is formed [3].

2. Clinical case

A 70 year old female patient was admitted due to retrosternal pain, dyspnea and fatigue for increasingly smaller efforts. The patient was obese (BMI:31; 1.58 cm, 81 kg), with previous diagnosis of hypertension and hypercholesteremia.

Due to clinical suspicion of coronary pathology, she was submitted to invasive coronary angiogram, which showed a fistula from the anterior descending artery to the pulmonary artery (Fig. 1A), with no other significant changes in the coronary arteries, having been discharged for outpatient investigation with the diagnosis of acute coronary syndrome of a non-atherosclerotic cause. She was submitted to a stress echocardiogram (dobutamin). During stress echocardiogram, she developed extreme angor with ventricular tachycardia, with no electrocardiographic signs of ischemia nor latter increase of serum myocardial necrosis markers. She was admitted to an intensive care unit for monitoring. After the cur-
rent episode, closure of the coronary fistula was considered and a Cardiac CT was requested.

A contrast enhanced coronary computed tomography was performed in a 64-slices device, with retrospective cardiac gating (Fig. 1). A coronary fistula was identified between the medial wall of the proximal anterior descending anterior coronary artery, with 2 mm caliber, with an tortuous path toward the main pulmonary artery. A second fistula was depicted in the right coronary artery, with a caliber of about 0.9 mm, coursing towards the right branch of the pulmonary artery (Fig. 2). No significant atherosclerotic plaques were seen.

3. Discussion

The estimated prevalence of CAF is 0.002% in the general population, representing only 0.4% of all congenital heart defects [4]. Despite being rare, there are two facts worth considering: firstly is that CAF are the coronary anomaly more often implicated in changes of hemodynamic parameters [5]. Secondly is that it has been reported that if untreated, hemodynamically significant fistulae may result in clinical symptoms or sequelae in approximately 19 percent of patients under the age of 20 and 63 percent of those over the age of 20 [6].

The use of MDCT for the detection of CAF has been confirmed to be reliable, with reported sensitivity of 100% even with the older 16-slice devices [7].

The prevalence of detection in patients undergoing coronary CT has recently been reported to be 0.9% [8] which is about a two to fourfold increase regarding the 0.1–0.2% prevalence referred in population submitted to invasive coronarography [9]. These discrepancies may be explained by the volumetric nature of CT, better characterization of mediastinal anatomy [2], as well as the lower rate of success in the difficult cannulation of anomalous arteries (31–55%) in angiography [10].

Regarding the technical protocol, standard coronary CT has been used. Saboo et al. [2] refer not using nitrates in order to assess eventual coronary aneurisms. Essential points to report are adequate description of the origin, degree of complexity, drainage sites, presence of an aneurysm (1.5× dilation comparing to adjacent vessels [8]), congenital or acquired anomalies, relation to adjacent structures.

Both right and left coronaries may be involved [10], with some recent series reporting high prevalence of bilateral fistula (up to 50% of CAF cases) [3,8].

Classically referred drainage sites are the right ventricle (41%), the right atrium (26%), the pulmonary artery (17%), the coronary sinus (7%), and, in less than 10% of cases, the left atrium and left ventricle, based on conventional angiographs [1]. However, other series have reported a higher prevalence of fistulae to pulmonary arteries (as high as 76.8%) [8].

The treatment of patients with CAF depends on the size and anatomic features of the fistula, the presence of symptoms, the patient’s age, and the presence of other cardiovascular diseases. Symptomatic fistula may be occluded by percutaneous intervention or by surgical ligation. The best approach to asymptomatic fistula, however, remains controversial [4].
4. Conclusion

Symptomatic CAF in older patients are rarely reported. Despite this, in any age coronary abnormalities may be an important cause of acute coronary syndromes, particularly in patients with no atherosclerotic changes. Patients with low risk of atherosclerotic pathology (or absence of these) and with evidence of ischemia may benefit from coronary CT for detecting coronary abnormalities [11].

CT’s post processing capabilities, tri-dimensional and maximal intensity projection reconstructions are invaluable in delineating coronary anatomy, essential for further treatment planning. These characteristics have also been associated with the higher prevalence of detected CAF in CT series comparing to the expected from previous surgical or angiographic series [2,3].

Awareness from radiologists and cardiologists is needed, as the prevalence of CAF may be higher than once thought and CAF may be a cause for cardiac symptoms.

Conflict of interest

No conflict of interest

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