

Coronary artery fistula connecting the left main coronary artery with the superior vena cava

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Abstract

Background: Coronary artery fistulas are rare congenital coronary artery abnormalities, with direct communication between a coronary artery and a cardiac chamber, great vessel or other structure.

Description of case: We report a case of a large coronary artery fistula connecting the left main coronary artery with the superior vena cava in a 70-year-old patient undergoing diagnostic coronary angiography for a non-ST-segment elevation myocardial infarction. The patient rejected closure of the fistula and remains asymptomatic on follow-up.

Conclusion: Angiographic recognition of coronary artery fistulas is important for the appropriate diagnosis and management of patients. Hippokratia 2015; 19 (2):186-188.

Keywords: Coronary artery fistula, coronary abnormalities, non-ST-segment elevation myocardial infarction, left main coronary artery, superior vena cava

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Introduction

Coronary artery fistulas (CAFs) are rare coronary artery abnormalities in which blood bypasses the myocardial capillary network and is shunted into a cardiac chamber, great vessel or other structure¹. They usually originate from the right coronary artery and terminate into the right ventricle or right atrium. We report a rare case of a CAF originating from the left main coronary artery and draining into the superior vena cava.

Case description

A 70-year-old man was referred for a diagnostic coronary angiography after a non-ST-segment elevation myocardial infarction, with the initial electrocardiogram revealing ST segment depression in the inferior leads and a pre-existing right bundle branch block. He had a 2-year history of arterial hypertension and megaloblastic anemia under treatment with monthly intramuscular injections of vitamin B12. On physical examination, the only remarkable finding was a continuous murmur, with a louder systolic component, at the upper right and left parasternal border.

The diagnostic coronary angiography revealed a dilated tortuous coronary artery fistula that was connecting the left main coronary artery with the superior vena cava and drained into the right atrium (Figure 1). The epicardial coronary vessels presented with no stenoses. The Qp/Qs ratio calculated from right heart catheterization was 0.968, very close to 1, which means that the left-to-right shunt was small. On coronary computed tomography an-

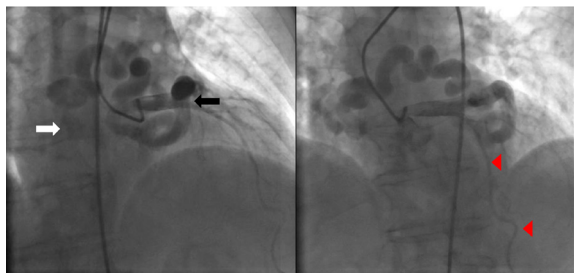


Figure 1: Diagnostic coronary angiography of the left coronary artery showing the large, tortuous fistula, originating from the left main (black arrow) and terminating through the superior vena cava into the right atrium (white arrow). The red arrowheads show the left anterior descending artery presenting with no stenosis.

giography the CAF followed a very tortuous pathway; was located between the aortic root and the left atrium, under the right branch of the pulmonary artery, formed a loop over a part of the left atrium, and finally drained into the superior vena cava prior to its entry site into the right atrium (Figure 2). The echocardiogram revealed mild aortic stenosis (V_{max} 2.88 m/sec), no segmental wall motion abnormalities, and a continuous turbulent flow pattern at the level of the interatrial septum and at the drainage site of the superior vena cava into the right atrium (Figure 3). The dimensions of the left and the right cardiac chambers were within normal limits.

We suggested a single photon emission computed to-

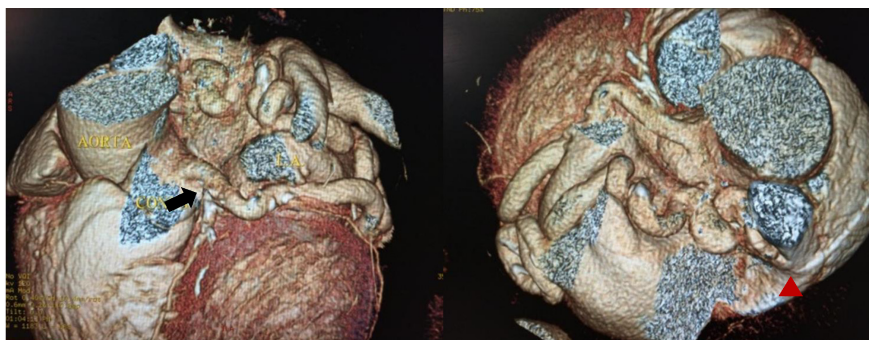


Figure 2: Coronary computed tomography angiography of the epicardial vessels showing the origin of the coronary artery fistula from the left main (black arrow), the very tortuous path over and around the left atrium, and the draining site into the superior vena cava (red arrowhead).

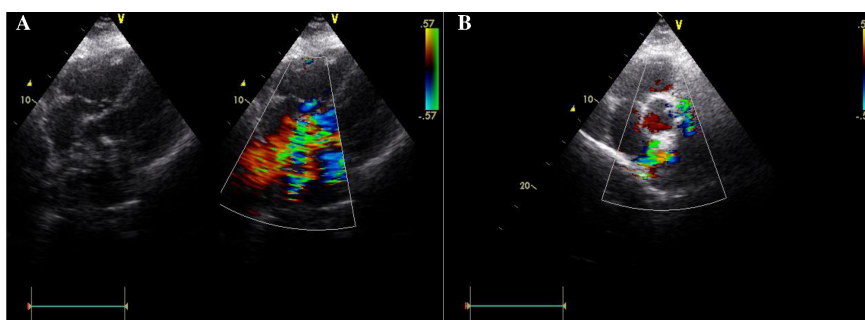


Figure 3: Transthoracic echocardiography. A turbulent flow pattern (“mosaic” color pattern) is seen at the drainage site of the superior vena cava into the right atrium on the subxiphoid view (A), and at the level of the interatrial septum on the parasternal short axis view (B).

mography to examine the possibility of ischemia during exertion, but the patient declined any further investigation and rejected the possibility of surgical or percutaneous closure of the CAF. He was dismissed with dual antiplatelet therapy for the acute coronary syndrome and instructions for good oral hygiene, but no antibiotic prophylaxis for infective endocarditis, based on current guidelines. On the six-month follow-up examination, the patient remains asymptomatic.

Discussion

CAFs are generally congenital in origin and account for 0.08% to 0.4% of all congenital heart diseases. They have been reported in 0.1% to 0.25% of patients undergoing coronary angiography^{2,3}. They are usually isolated, but they may be associated with other congenital cardiac anomalies, such as tetralogy of Fallot, atrial or ventricular septal defects, and persistent ductus arteriosus. They may be single or multiple⁴.

CAFs originate from the right coronary artery in about 55%, from the left coronary artery in about 35%, and from both coronary arteries in about 5% of the reported cases. The origin from the left main coronary artery is rare. Approximately 92% of CAFs drain into the right side of the heart, whereas only 8% of them terminate into the left side of the heart. More specifically, the majority of CAFs drain into the right ventricle (41%), into the right atrium

(26%), and into the pulmonary artery (17%). Termination into the superior vena cava is rare (1%)^{1,2}. There have been only a few cases reporting a CAF connecting the left main coronary artery with the superior vena cava⁴⁻⁸.

The majority of adult patients with CAFs are asymptomatic, but they can present with angina pectoris or myocardial infarction due to the coronary steal phenomenon. Indeed, CAFs are one of the causes of the rare non-atherosclerotic acute coronary syndrome in the adult population. Other causes include coronary vasculitis, dissection, congenital anomalies of the coronary arteries, fibromuscular dysplasia, drug abuse or iatrogenic complications⁹.

CAFs can also present with dyspnea due to the development of pulmonary hypertension and/or congestive heart failure. Other symptoms include arrhythmias, rupture or dissection of the CAF, stroke, and endocarditis^{1,5}. Transcatheter closure or surgical ligation of the fistula is the definitive treatment, especially in patients with large shunts, documented cardiac ischemia, congestive cardiac failure, or a pulmonary-to-systemic flow ratio greater than 1.5, but the majority of patients especially those with low-flow fistulas are managed medically^{2,10,11}. Close follow-up is mandatory.

Conflict of interest

The authors declare no conflict of interest.

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