Dual Coronary Artery Fistula from Left Anterior Descending and Right Coronary Artery to Pulmonary Trunk in a Patient with Myocardial Infarction—A Case Report

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Abstract

Coronary artery fistulas (CAF) are precapillary communications between a coronary artery and a cardiac chamber or vessel. CAF have been described as the most common hemodynamically significant congenital coronary anomal. However, it remains a relatively uncommon clinical problem. Coronary fistulas originates slightly more common from the right than from the left coronary artery, but the bilateral fistulas—those that originate from both coronary arteries—accounts for only 5% of total cases. These bilateral fistulas have a unique tendency to terminate in the pulmonary artery. More than half of the bilateral and only 17% of unilateral fistulas, terminates in this manner [1]. CAF are believed to be embryological remnants of sinusoidal connections between the lumens of the primitive tubular heart. This was first described by Maude Abbott in 1908 [2]. These fistulas are usually discovered incidentally upon coronary angiography [3]. Their incidence in the overall population is reported about 0.002% and constitutes 0.13% of congenital cardiac lesion, however, they are found in 0.05% to 0.25% of patients who undergo coronary angiography. The most common site of drainage is the right ventricle seen in 41% of patients. Congenital CAFs usually result from abnormal embryological development of the myocardial vascular system. Acquired CAFs are seen after trauma, endovascular procedures like coronary angiography, endomyocardial biopsy etc or cardiac transplantation. True fistulas of the circulatory system are characterized by an ectatic vascular segment that shows aberrant flow connecting two vascular territories governed by large pressure differences. We report a case of double coronary to pulmonary artery fistula discovered during emergent coronary angiography for acute inferior wall ST-elevation myocardial infarction (STEMI) in a patient with no prior cardiac symptoms.

Keywords

Myocardial Infarction, Coronary Artery Fistula, Pulmonary Artery

1. Case

A 48-year-old man presented with acute onset chest pain in emergency. Electrocardiogram revealed ST segment elevation of >2 mm in leads II, III, aVF. Blood for troponin T was positive. The patient was diagnosed as a case of ST segment elevation inferior wall myocardial infarction. Dual Antiplatelets were started with other supporting measures and the patient was transferred immediately to the catheterization laboratory. Coronary angiography showed significant stenoses of the left anterior descending (LAD), left circumflex (LCX) arteries and borderline lesion in mid right coronary artery (RCA). Moreover, it revealed the presence of 2 large coronary artery fistulas (CAF), one originating from distal part of left circumflex artery (Figure 1) and the other from proximal part of right coronary artery (Figure 2), both draining into the pulmonary trunk at 2 different, but close, entry points.
The CABG was carried out on pump. No complications were observed in the post-operative phase and the patient was discharged 10 days after surgery to join a rehabilitation program.

2. Discussion

The incidence of coronary artery fistula in coronary angiogram has been reported to be 0.2%. Coronary artery fistula (CAF) originates from the right coronary artery in 15% of cases, from the left coronary artery in 53%, from both coronary arteries in 7% [4]. Fistula between coronary artery and pulmonary artery is a rare congenital anomaly first described by Krause in 1865. First successful surgical treatment of CAF was described by Fell and colleagues in 1958 [5]. Coronary artery fistula (CAF) can be congenital or acquired. The majority of CAFs reported are of the congenital form. Acquired CAF has rarely been discussed. There is no race or sex predilection for CAF. Bilateral coronary artery-pulmonary artery fistulas, however, are a distinct entity. In contrast with unilateral coronary artery fistulas, of which only 17% terminate in the pulmonary artery, 56% of the reported bilateral fistulas have their termination in pulmonary artery [4]. Most of the fistulas of coronary artery arise by failure of intramyocardial sinusoids to obliterate, so bilateral involvement requires independent developmental errors at two separate sites. However, fistulas to the pulmonary artery probably arise by supernumerary implantation of developing coronary arteries into pulmonary arterial portion of the embryonic truncus arteriosus. This bilateral involvement is thus facilitated by proximity of both coronary arteries to the truncus. Anomalous origin of the left or right coronary artery from the pulmonary artery can be explained by this defect in embryologic coronary implantation [6]. Acquired coronary to pulmonary artery fistulas are most often due to complications after coronary artery bypass surgery (CABG), particularly when the internal mammary artery is used and also after cardiac transplantation. It may also result after percutaneous coronary interventions (PCI). One of the mechanisms proposed for pathogenesis of the acquired coronary to pulmonary artery fistulas is because of neovascularization under the control of specific organizing proteins. The cause of the coronary fistula in our patient is uncertain because it could be congenital or acquired. Most of the patients are asymptomatic and are usually discovered as incidental findings during cardiac catheterization and are seen on one of every 500 - 1000 coronary angiograms [6]. Congenital CAF are commonly associated with obstructive coronary artery disease. Patient is usually asymptomatic. Patient may present with symptoms such as fatigue and dyspnea. Cardiomyopathy, congestive heart failure, atrial fibrillation can occur as late findings in cases of coronary artery fistulas [7]. Other complications related to coronary fistulas include myocardial ischemia from a steal phenomenon whereby competitive flow occurs and also from embolization due to thrombus formation within aneurysmal segments. Clinical signs uncommonly found in an adult patient with CAF include a continuous murmur heard along the left sternal border and cardiomegaly with enlargement of the right chambers [8]. Location of murmur at the apex and accentuation of the diastolic component are important characteristic findings of coronary artery fistulae. Selection of therapeutic option in fistulas is controversial. If the fistulas are small or does not cause ischemia follow up can be the alternative strategy. If ischemia occurs secondary to fistula or is symptomatic, transcatheter closure of coronary artery fistula or surgery can be the choice for treatment. In appropriate cases, trans-catheter closure can be the alternative to surgery. However, surgical repair may be indicated in cases of coronary artery fistulas with additional complex heart disease which requires surgery. The main indications for closure of CAF are clinical symptoms, particularly heart failure and myocardial ischemia. In asymptomatic patients with high-flow shunting closure is done to prevent occurrence of undesirable complications. Surgical intervention is also recommended in patients who are at risk for future complications such as bacterial endocarditis, thrombosis,
distal embolization, aneurysm, dissection, rupture, pulmonary hypertension, premature atherosclerosis, myocardial ischemia [9]. Currently, percutaneous treatment is proposed as the first choice because it is less radical and entails a shorter period of hospitalization [10]. Surgery is reserved for cases of multiple fistulae, those affecting large branches during embolization of coils, or when the fistulous connection is narrow, restrictive and draining into a cardiac chamber [11]. Our patient presented with inferior wall myocardial infarction with significant lesions in LAD, LCX and along with dual coronary artery fistula originating from distal part of left anterior descending artery and proximal part of right coronary artery. Patient was taken for coronary artery bypass grafting followed by ligation of fistula.

References


Abbreviations

CAF, Coronary Artery Fistulas;
LAD, Left Anterior Descending;
LCA, Left Circumflex Artery;
RCA, Right Coronary Artery;
STEMI, ST-Elevation Myocardial Infarction;
CABG, Coronary Artery Bypass Surgery.
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