

A Rare Combination of Left Main Coronary Artery Fistula to Main Pulmonary Artery with Severe Left Main Coronary Artery Disease in a 69-Year-Old Man

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ABSTRACT

A 69-year-old man was referred to our hospital with a diagnosis of non-ST elevation myocardial infarction. A selective left coronary angiography revealed a fistula connecting the left main coronary artery with the pulmonary artery in addition to severe left main coronary artery disease. The patient subsequently underwent ligation of fistulae and coronary bypass grafting. The combination of a fistula and severe artery disease seen in this patient is unusual because fistulas originating from the left main coronary artery drain into the pulmonary artery in patients with severe left main coronary artery disease.

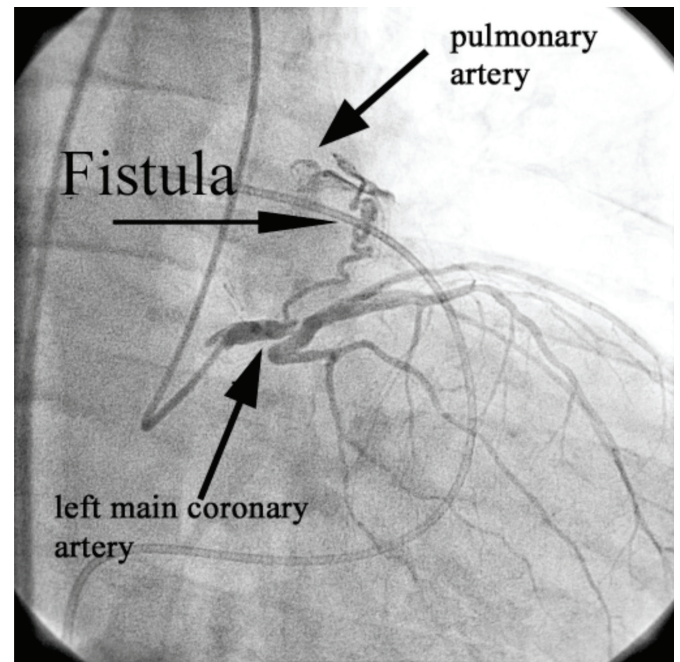
INTRODUCTION

Coronary artery fistula is defined as a direct communication of a coronary artery with a cardiac chamber, great vessel, or other vascular structure, bypassing the myocardial vascular bed [Shyam 1997]. Most fistulas drain into the right ventricle or pulmonary artery, although a variety of other drainage sites have been reported [Armsby 2002]. The incidence of this anomaly ranges from 0.2%–0.4% in selected series of congenital heart disease patients, and its incidence in the overall population is estimated to be about 0.002% [Liberthson 1979; Marci 1982; Vavurnakis 1995]. We present a case report of left main coronary artery-to-pulmonary artery fistula in a patient with severe left main coronary artery disease.

CASE REPORT

A 69-year-old man was admitted to our hospital because of recurrent episodes of chest pain that lasted for approximately 20 to 25 minutes and did not respond to the sublingual administration of nitrates. He was hospitalized with the diagnosis of non-ST segment elevation myocardial infarction. The patient had no history of cardiovascular disease. Electrocardiogram on admission revealed normal sinus rhythm. Cardiac

troponins and MB-CK were elevated to more than twice the upper limits of normal values. The patient's hemodynamic status was stable, with a blood pressure of 120/70 mm Hg and a heart rate of 88 beats/min. Physical examination was otherwise unremarkable. The patient was effectively treated with standard medical management based on pharmacotherapy with aspirin, clopidogrel, enoxaparine, β -blocker, ACE inhibitor, and statin. Echocardiography revealed hypokinesis of the anterior and apical walls, with decreased left ventricular ejection fraction reaching the level of 30%. Coronary angiography performed in an effort to more carefully delineate the anatomy confirmed the presence of a left main coronary-to-pulmonary artery fistula with severe left main coronary artery disease (Figure) and the normality of the remaining coronary anatomy. Ventriculography showed anterolateral and apical hypokinesia. Right-heart catheterization was performed.



Coronary angiogram (right anterior oblique view) showing a fistula between the left main coronary artery and the main pulmonary artery and severe left main coronary artery disease.

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Multiple blood samples obtained to calculate the magnitude of the left-to-right shunt documented a Qp/Qs of 1.1:1.

We discussed this case immediately with cardiothoracic surgeons and consensually decided to perform surgical revascularization and ligation of the fistula. Coronary artery bypass graft surgery was performed with a saphenous vein graft to the circumflex artery and a left internal mammary artery graft to the left anterior descending coronary artery. The fistula was ligated surgically.

DISCUSSION

Congenital coronary fistula is a rare heart defect that consists of a communication between a coronary artery and a cardiac chamber or pulmonary vessel. Most coronary artery fistulas drain into the right heart chamber or into the pulmonary artery [Fernandes 1992] and arise from the right coronary artery (50%); fewer than half arise from the circumflex artery, and 5% arise from both coronary arteries. The causative factors are unknown, but most fistulas are thought to originate as congenital anomalies or, less commonly, from injury during coronary intervention or a surgical procedure [Armsby 2002]. Because our patient had no history of coronary intervention or surgery and did not manifest any clinical or laboratory evidence of arteritis, we thought that the fistula was congenital in origin.

Potential complications of coronary artery fistulae include pulmonary hypertension and congestive heart failure if a large left-to-right shunt exists, bacterial endocarditis, rupture of the fistula, and myocardial ischemia due to coronary steal phenomenon [Brussee 1992].

Angiography is the traditional method used for definite diagnosis of coronary artery fistula. Transesophageal echocardiography and multislice computed tomography may provide more satisfactory images of the origin, course, and drainage site of the coronary artery fistula [Lin 1995; Shiga 2008]. In the patient we describe, this rare abnormality was discovered incidentally during routine coronary angiography.

Most coronary artery fistulas are asymptomatic, and conservative treatment has been recommended for asymptomatic patients [Athanasias 2002]. Clinically, patients with coronary fistula may present with dyspnea, symptoms of angina, or palpitations. There is general agreement that symptomatic patients should be treated. Current treatment options include careful observation, surgical ligation with or without cardiopulmonary bypass, ligation with bypass of the involved coronary artery, and transcatheter embolization [Lowe 1981]. Konno et al [1973] also suggested the following criteria regarding indications for surgery: a shunt rate $>30\%$, ischemic or hypertrophic changes indicated by electrocardiogram, anticipation of the progression of pulmonary hypertension or congestive heart failure, history of ischemia, morphological saccular-type aneurysm, and social reasons. Rittenhouse et al [1975] reported that the presence of a large shunt flow (Qp/Qs > 2.0) with hemodynamic changes and symptoms of heart failure indicate the need for surgery. Evaluation of shunt flow is important because the amount of shunt flow is a major factor in the decision to perform surgery. In our case the left-to-right shunt was insignificant (QP/QS = 1.1), and there was no significant step-up in oxygen saturation.

The fistula we found in our patient was small and without significant shunt circulation. We decided to ligate the fistula for 2 reasons. First, the patient required a surgical procedure involving cardiopulmonary bypass and aortic cross-clamp and cardioplegia for treatment of left main coronary artery disease. Second, we considered that this fistula increased coronary ischemia by decreasing the coronary perfusion pressure.

We have not seen another report in the literature of a patient with significant left main coronary artery disease and left main coronary artery-to-main pulmonary artery fistula associated with non-ST segment elevation myocardial infarction. In summary, in the rare cases in which left main coronary artery-to-pulmonary artery fistulas coexist with severe left main coronary artery disease, these fistulas can cause myocardial ischemia and should be treated surgically.

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