Case Report

Multiple Coronary to Pulmonary Arterial Fistulas with Coronary Steal Phenomenon

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Abstract

Coronary artery fistula is an abnormal communication between one or more coronary arteries and great vessels or a cardiac chamber. Majority of them are clinically silent, but rarely chest pain, myocardial ischaemia and heart failure may be the presenting features in the absence of obstructive coronary arterial disease. Here, we present a case of a 30-year-old woman with multiple coronary to pulmonary fistulas, diagnosed by coronary angiography and multidetector computed tomography angiography whose symptoms resolved with anti-ischemic medical treatment without any surgical intervention.

Introduction

Coronary artery fistulas are abnormal connections between coronary arteries and cardiac chambers or great vessels [1]. Majority of them are congenital [2], but may be acquired secondary to intravascular diagnostic or therapeutic procedures or even secondary to blunt or penetrating trauma [3-5]. Patients with coronary artery fistula may be asymptomatic or present with different symptoms such as angina pectoris, palpitations, syncope, congestive heart failure, and may even present with sudden cardiac death [6]. There are various imaging modalities to detect coronary artery fistulas such as echocardiography, coronary angiography and multidetector computed tomography angiography. The following case report describes a young patient presenting with angina symptoms, whose coronary angiography revealed an abnormal tortuous plexus of multiple fistulas from circumflex and right coronary arteries communicating with pulmonary trunk.

Case Presentation

A 30-year-old woman presented to the outpatient clinic with recurrent chest pain for the last 2 years. Recently, she had increased severity and frequency of angina symptoms. Physical examination was normal and family history was not suggestive of any cardiac illness. Blood pressure was also within normal limits (124/78 mm of Hg). Chest X rays, electrocardiography were normal. Transthoracic echocardiography revealed normal left ventricular ejection fraction with no wall motion abnormalities. Treadmill exercise test showed the depression of ST segments in lead II, III and aVF. Coronary angiography was performed but revealed no sign of atherosclerosis in the coronary arteries; however, abnormal tortuous communications were present originating from the distal branches of circumflex and right coronary arteries emptying into the main pulmonary artery (Figure 1, Video 1-3).
Figure 1: Coronary angiography revealing multiple coronary artery fistulas originating from the distal branches of circumflex (A) and right coronary arteries (B) emptying into the main pulmonary artery (Cx: Circumflex Coronary Artery, LAD: Left Anterior Descending Coronary Artery, RCA: Right Coronary Artery).

Video 1. Coronary angiography right anterior oblique caudal view revealing coronary artery fistula originating from the distal branches of circumflex coronary artery and emptying into the main pulmonary artery artery.
Video 2. Coronary angiography left anterior oblique view revealing two coronary artery fistulas originating from the distal branches of right coronary artery and emptying into the main pulmonary artery artery.

Video 3. Coronary angiography anteroposterior cranial view revealing two coronary artery fistulas originating from the distal branches of right coronary artery and emptying into the main pulmonary artery artery.
Additionally, multidetector computed tomography angiography was performed and showed small, tortuous side arteries from the circumflex and right coronary arteries and the exact terminations to the main pulmonary artery were confirmed. The best treatment method for the patient was discussed in the council of cardiology and cardiovascular surgery. Since the fistulas were multiple, surgical ligation was suggested however the patient refused surgery and was asymptomatic for 6 months under antianginal medical therapy including acetylsalicylic acid (100 mg/day) and metoprolol (50 mg/day).

**Discussion**

Coronary artery fistulas are abnormal communications between the coronary arteries and a chamber of the heart or any segment of systemic or pulmonary circulation close to the heart. They are typically congenital but may also be acquired following trauma or invasive cardiac procedures [2-5]. They have been reported in 0.05%-0.25% of patients undergoing coronary angiography, with an estimated prevalence of 0.002% in the general population [7]. In 55% of cases congenital coronary fistulas originate from the right coronary artery or its branches; the left coronary artery is involved in 35% of cases, and both coronary arteries are involved in 5% of cases. Typical drainage sites include the right ventricle and pulmonary trunk [8].

Patient with coronary artery fistulas are usually asymptomatic that the fistula may be accidentally detected by echocardiography and coronary angiography, but patients may have varied symptoms such as angina pectoris, palpitations, syncope and dyspnea. In some cases, physical examination may reveal a murmur if the flow is significant [6]. Hemodynamic consequences and clinical presentation depend on the size of the fistula and shunt volume. Potential complications of coronary artery fistulas include bacterial endocarditis, rupture of the fistula, myocardial ischemia due to coronary steal phenomenon and pulmonary hypertension as well as congestive heart failure, if there is a large left-to-right shunt [9].

Imaging plays an important role in the diagnosis and management of these fistulas. Coronary angiography is the traditional technique used in the evaluation of coronary fistulas, however, the identification of the exact site of drainage and relationship to adjacent structures is often difficult. Transthoracic and transesophageal echocardiography have a limited role in the evaluation of coronary fistulas, especially in demonstration of the anatomy and drainage. Cross sectional modalities such as computed tomography and magnetic resonance imaging with high spatial and temporal resolutions are the most useful imaging modalities in diagnosis and accurate characterization of cardiovascular fistulas. Multi-planar three dimensional reconstruction techniques such as volume rendering and surface shading enable exquisite demonstration of the anatomy of the fistulas, which is essential for surgical/interventional planning [10].

Small asymptomatic coronary artery fistulas are often treated with antiplatelet therapy and monitored for complications, without intervention. The main treatment options for symptomatic coronary fistulas are surgical ligation and transcatheter coil embolization which has become a safe and effective alternative to standard surgical closure [11,12]. Success of surgical closure has been detected 89.4%, 85.1%, 83.0%, and 83.0%, at discharge, 6 months, 1 year, and 5 years postsurgery, respectively [13]. A residual shunt smaller than 2-3 mm could be observed without further intervention close monitoring should be performed during the postprocedural follow-up period. Recent American College of Cardiology guidelines [14] recommends interventional management as a class I recommendation for large coronary artery fistulas, regardless of the symptoms, and small to moderate-size fistulas with symptoms, including myocardial ischemia, arrhythmia, ventricular dysfunction, and endarteritis. Fistulas with a large size, multiple communications, and adjacent vessels at risk may be contraindications to transcatheter closure.

Angina secondary to coronary artery fistula is managed according to the standard guidelines on the management of stable coronary artery disease. The treatment may include antiplatelets, beta-blockers, calcium channel blockers, trimetazidine, nitrates, and ranolazine [15]. In the present case, the preferred treatment was surgical ligation but the patient refused surgery and her symptoms were resolved with optimal anti-ischemic medical therapy.

**References**