

A Giant Circumflex Coronary Artery Aneurysm with a Fistula into the Coronary Sinus: Case Report

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ABSTRACT

Coronary artery fistulas are a rare clinical entity in cardiac anomalies, and in a significant portion of patients, the fistulas are accompanied by coronary aneurysm formation. In this article, we present a giant circumflex coronary artery aneurysm with a fistula into the coronary sinus.

INTRODUCTION

Coronary arterial fistulas (CAFs), although rare, are one of the most common coronary artery anomalies (with an established incidence of 0.3-0.8%) and often present as congenital lesions [Lee 2012]. In rare cases, thoracic trauma, cardiac surgery, or coronary intervention are the other possible causes of CAFs. Coronary artery aneurysms may develop with an incidence of 19-20% of coronary arteriovenous fistula cases [Milici 2013]. In most cases, the fistula terminates in the right side of the heart. The endpoints of fistulas can be located in the superior vena cava, coronary sinus or pulmonary artery because of low pressures. The right coronary artery origin of fistulas (52%) is the most common form. Left anterior descending and circumflex artery origins are less common (17%). Moreover, circumflex artery–coronary sinus fistulas are even more rare.

Coronary arteriovenous fistulas in adult patients are asymptomatic. However, they may be present with heart failure, arrhythmia, stroke, or coronary artery rupture, or may cause myocardial ischemia due to stealing from the coronary artery [Lee 2012; Milici 2013]. These clinical symptoms may vary depending on the coronary artery anatomy and the flow rate of the shunt formed between the right and left sides of the heart.

Although there is no clear consensus regarding CAF treatment in the literature and surgical experience is limited, it is recommended that treatment by transcatheterization or

surgical closure should be considered at the onset of heart failure symptoms or in patients with left-to-right shunts. It is also important that the management of coronary fistulae be individualized based on the magnitude of the volume load on the heart and the presence or absence of myocardial ischemia or ventricular dysfunction.

In this report, we present a case of aneurysmal coronary arteriovenous fistula with an origin in the circumflex artery and developing into a coronary sinus fistula, which was treated surgically.

CASE REPORT

A 48-year-old woman presented to our institution with fatigue, palpitations, chest pain, and shortness of breath (NYHA class II) of six months' duration. Her past medical history was unremarkable except for well-controlled hypothyroidism. The clinical examination revealed that she was slightly distressed with tachypnea (without pallor or cyanosis) and a continuous heart murmur. Her vital signs were stable. An ECG showed atrial fibrillation. Chest X-ray displayed mild enlargement of the cardiac shadow. Transthoracic echocardiography (TTE) revealed an anomalous vessel-like structure

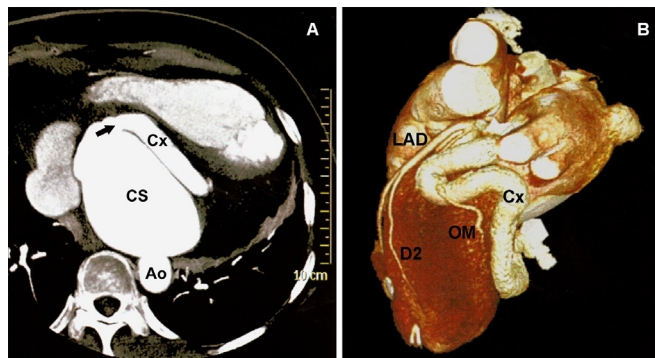


Figure 1. Coronary CT angiography of (A) giant circumflex coronary artery opening into the orifice of the coronary sinus and a dilated coronary sinus due to abnormal flow; (B) aneurysmatic left circumflex artery. Cx indicates circumflex artery; CS, coronary sinus; Ao, aorta; LAD, left anterior descending artery; D2, second diagonal artery; OM, obtuse marginal branch of the circumflex artery.

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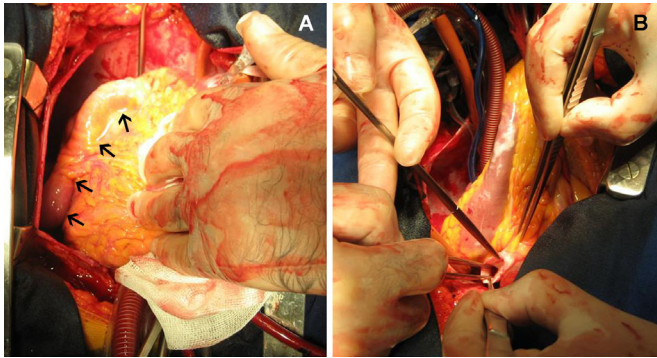


Figure 2. Operation image of (A) aneurysmal circumflex artery on the posterior wall of the heart; (B) arteriotomy performed to the distal segment of the circumflex artery. The forceps demonstrate the opening of the fistula into the coronary sinus.

around the coronary sinus, with a high flow ($Q_p:Q_s > 2$) and an enlarged coronary sinus. The left ventricular ejection fraction (LVEF) was 60%, with no wall or valvular abnormalities. Due to the unclear origin of this aneurysmatic vessel and the iatrogenic risk of rupture, we performed MDCT coronary angiography, which demonstrated a dilated left circumflex artery connecting to a giant coronary sinus (Figure 1).

In this case, surgical repair was preferred because of a symptomatic patient, a large CAF characterized by a hemodynamically significant shunt, significant aneurysm formation from the LCA (2 cm), and a giant coronary sinus.

With the patient under general anesthesia, the heart was approached through a median sternotomy. On the posterior aspect of the heart, the left circumflex artery (LCA) and the coronary sinus were observed to be extremely enlarged (Figure 2, A). Therefore, the heart was arrested via only antegrade cardioplegia administration, with digital compression of the fistula to prevent direct run-off into the coronary sinus (CS). The distal LCA was dissected and opened longitudinally. Both the fistula and the communication points of the LCA were identified from the inside (Figure 2, B). The fistula was closed using a 6/0 monofilament continuous suture and reinforced with autologous pericardium. The tortuous and aneurysmatic LCA was plicated with a 7/0 monofilament continuous suture, and then the CS opening was closed using the same technique.

Weaning from CPB was uneventful with a sinus rhythm, and the intraoperative TEE demonstrated good wall contractility of all segments without any ECG alternations. The postoperative course was also uneventful, and we observed no arrhythmias. The patient was discharged home in excellent condition on postoperative day 5. At the one-year follow-up, she was free from symptoms, and the repeat echocardiogram revealed a patent and normal-sized CS, with a 65% LVEF.

DISCUSSION

Coronary artery fistula in adults is a rare entity; according to Ogden's classification of congenital anomalies of the coronary arteries, CAFs are present in 0.002% of the general

population and represent 0.4% of all cardiac malformations [Ogden 1969; Dodge-Khatami 2000]. Although no guidelines have been developed to aid in managing these patients due to the rarity and anatomical variations of these fistulas, according to the angiographic classification by Sakakibara et al, types A and B exist, as follows: 1) type A – proximal type, proximal coronary segment dilated to the origin of the fistula, with the distal end normal; and 2) type B – distal type, coronary artery entirely dilated, terminating as a fistula into the right side of the heart [Sakakibara 1966].

Clinical symptoms of a congenital CAF depend on the underlying anatomy and on the size of the fistulous connection to the left or right side of the heart. Symptoms are variable and include chronic myocardial ischemia and angina, congestive heart failure, cardiomyopathy, myocardial infarction, pulmonary hypertension, infective endocarditis, arrhythmias and, rarely, rupture. CAFs can also be associated with atherosclerotic coronary artery disease in older patients or after previous cardiac interventional procedures.

The pathophysiology of a fistula is related to the connection site. In our case, fistula insertion into the right-sided heart chamber caused left-to-right shunting and right- and left-sided heart volume overload. Because of the large fistula and marked flow, myocardial ischemia due to a coronary steal phenomenon may occur. Other possible complications related to coronary artery fistulae are endarteritis, thrombosis within a fistula, fistula rupture, or pulmonary hypertension.

Aneurysmatic dilatation of the LCA may depend on the resistance of the fistulous connection at the site of fistula termination and increased wall tension due to high flow. According to the Poiseuille-Hagen law, the resistance is determined by the size, tortuosity, and length of the pathway. Consequently, blood follows the lower-resistance pathway through the fistula rather than flowing through the higher-resistance smaller arterioles and capillaries of the myocardium, thereby increasing pressure on the wall tension of the coronary artery. This fact may explain why CAFs tend to grow with age, because they are affected by long periods under high pressure. On the other hand, flow from the coronary artery to the coronary sinus occurs throughout the cardiac cycle. Thus, fistulas may cause diastolic runoff and draw blood away from the normal coronary pathway with a widened pulse pressure, and may result in coronary steal [Lowe 1981].

It is obvious that the clinical symptoms of ischemia or volume overload are the primary indications for closing a fistula. However, the treatment of asymptomatic lesions is controversial [Lowe 1981]. Some authors recommend early surgical intervention, whereas others recommend a more conservative approach. Although it is very uncommon, spontaneous closure of the fistula secondary to spontaneous thrombosis has also been reported [Zenooz 2009]. Consequently, this is the main reason for treatment considerations with respect to asymptomatic patients. Although there is no urgency to close CAFs immediately and although careful periodic evaluations with close follow-up are definitely useful, CAFs should be corrected because of potentially fatal complications and to prevent the occurrence of symptoms (especially in the pediatric population). Current treatment options for CAFs vary and include

surgical ligation alone, surgical ligation with coronary artery bypass surgery, or transcatheter closure [Chowdhury 2009].

Surgical closure can be performed using different techniques [Chowdhury 2009; Darwazah 2011; Nakayama 2011]. Proximal-type CAF (type A) can be treated by epicardial ligation maintaining normal flow, and distal-type CAF (type B) requires termination of the fistula flow by an intracardiac purse-string suture via cardiopulmonary bypass. Although the CAF with LCA aneurysm in our patient was of the distal type, we preferred to plicate the LCA to prevent turbulent flow, because of the aneurysm and to avoid the necessity of performing an obtuse marginal bypass.

Surgical treatment results are satisfactory, with a low mortality from 0-4% and morbidity from 10-15%. Due to the risk of postoperative recanalization, long-term follow-up is essential for persistent dilatation of the coronary artery and ostium, thrombus formation, calcification, arrhythmias, and myocardial infarction [Sherwood 1999].

Consequently, coronary arteriovenous fistulas are rare but also make treatment difficult when observed with coronary artery aneurysms. In cases that have a left-to-right shunt with high flow, it is not recommended to perform transcatheter embolization, and surgical treatment is required. Fistula ligation and resection of aneurysmal formation, while carrying the risk of myocardial infarction, creates the risk of rupture only during the late period of fistula closure. In these cases, the surgical treatment strategy is currently controversial.

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