Coronary Artery Fistula From the Left Circumflex to the Coronary Sinus

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SUMMARY

A 27-year-old woman, who had received mitral valve repair for mitral regurgitation resulting from infective endocarditis, was admitted for a close examination of abnormal echocardiographic findings in the left atrium. Transthoracic echocardiography showed trivial mitral regurgitation with normal left ventricular contraction and dilatation of the coronary sinus. Auscultation revealed a grade 2 continuous murmur along the left sternal border. Transesophageal echocardiography demonstrated a marked dilatation of the coronary sinus just behind the posterior wall of the left atrium and turbulent blood flow in the dilated coronary sinus. Cardiac catheterization showed no significant step-up of oxygen saturation in the right heart and normal pulmonary artery pressure. Coronary angiography revealed a markedly dilated and tortuous circumflex coronary artery connected to the coronary sinus through a fistula. A left circumflex artery with a fistulous connection to the coronary sinus is extremely rare. (Int Heart J 2006; 47: 147-152)

Key words: Coronary artery fistula, Coronary artery aneurysm, Circumflex coronary artery, Coronary sinus

Coronary artery fistula (CAF) is a rare congenital anomaly with a reported incidence of 0.1% to 0.2% in the adult population referred for cardiac catheterization.1) Fistulous connection into a cardiac chamber or major vessel often causes a marked dilation of the donor coronary artery leading to aneurysm formation.2) Among various coronary artery anomalies, fistulous connection of the aneurysmal circumflex coronary artery to the coronary sinus has been previously reported as an extremely uncommon form.3-8)

In this paper, we describe a case of an aneurysmal circumflex artery connecting to the coronary sinus through a fistula, and discuss surgical management for patients with CAF, particularly for asymptomatic patients with a small left-to-right shunt.
CASE REPORT

A 27-year-old woman was admitted for a close examination of abnormal echocardiographic findings in the left atrium. The patient had suffered from infective endocarditis of the mitral valve during pregnancy and had undergone mitral valve repair 42 days after delivery for severe mitral valve regurgitation (MR) at the age of 23 years, but she had not received cardiac catheterization or coronary angiography before the operation because of the active stage of infection and her relatively young age. After successful mitral valve repair, she returned to her daily routine without any symptoms and one year later gave birth to another healthy infant. Transthoracic echocardiography performed at the clinic in the 4th postoperative year showed normal left ventricular contraction and dilatation of the coronary sinus that was not noticed before the operation. On admission, she did not have any complaints of chest pain or any signs of congestive heart failure. Auscultation of the chest revealed a grade 2 continuous murmur along the left sternal border. Chest X-rays showed no cardiomegaly or pulmonary congestion. Electrocardiography showed normal sinus rhythm and no specific changes in the ST segment and T wave. Transesophageal echocardiography confirmed a marked dilatation of the coronary sinus just behind the posterior wall of the left atrium (Figure 1). Doppler study showed trivial MR and turbulent blood flow in the dilated coronary sinus. Cardiac catheterization showed no significant step-up of oxygen saturation in the right heart and normal pulmonary artery pressure. Coronary angiography revealed a normal right coronary artery and left anterior descending artery. The markedly dilated and tortuous circumflex coronary artery, which was aneurysmal and connected to the coronary sinus through a fistula, was delineated (Figure 2). Based on these findings, a diagnosis of CAF from

Figure 1. A: A transthoracic echocardiogram showing an unidentified cystic mass in the left atrium. B: A transesophageal echocardiogram showing a marked dilatation of the coronary sinus just behind the posterior wall of the left atrium. AO indicates aorta; LA, left atrium; LV, left ventricle; and CS, coronary sinus.
the circumflex artery to the coronary sinus was made, and interventional therapy such as transcatheter embolization or surgical closure of the CAF with cardiopulmonary bypass (CPB) was recommended. However, the patient refused to receive any interventions, despite our proposal.

**DISCUSSION**

This case demonstrates a very rare pathology of an aneurysmal circumflex coronary artery connecting to the coronary sinus through a fistula. CAF is the most common coronary arterial malformation but is a rare cardiac anomaly. Similarly, a coronary artery aneurysm is also a rare form of coronary artery disease, and is characterized by an abnormal dilatation to a diameter of more than 1.5 fold that of the adjacent normal coronary artery. Although the majority of coronary artery aneurysms are due to coronary atherosclerosis, congenital fistulous connection of the coronary artery into a cardiac chamber or major vessel often causes a marked dilation of the donor coronary artery leading to aneurysm formation. Congenital fistulous connection of the coronary artery into a cardiac chamber or major vessel often causes a marked dilation of the donor coronary artery leading to aneurysm formation, as seen in our patient. In an angiographic study, aneurys-
mal dilatation of the coronary artery was found in 26% of 23 CAFs. In CAF, the right coronary artery is generally involved most frequently, followed by the circumflex coronary artery, and the most common drainage site is the right ventricle. On the other hand, the rarest drainage site is the coronary sinus. To our knowledge, only 8 cases of fistulous connection of an aneurysmal circumflex coronary artery to the coronary sinus have been previously described in the English literature (Table).3-8)

The optimal therapeutic strategy for CAF is not clearly established. Although it is evident that surgical intervention is necessary for symptomatic patients, those with complications, and those with a significant shunt (Qp/Qs > 2.0), the indications for surgical intervention in asymptomatic patients with no or only mild shunting are still controversial, because of the great variability in the natural history. It has been thought that most patients with CAF are asymptomatic. However, some studies have emphasized that the incidence of symptoms and complications increases with age, particularly after the age of 20.11,12) A review of 187 patients has shown that symptoms and fistula-related complications occurred in 55% and 35% of patients over 20 years, respectively, but in 9% and 11% of those under 20 years old.11) The most frequent symptoms and fistula-related complications were dyspnea on exertion, palpitations, congestive heart failure, myocardial infarction, infective endocarditis, and death.11) Besides these major complications, rupture of coronary artery aneurysms, a very rare but life-threatening complication of CAF, has also been reported in several cases.13-15) Based on these reports, it is recommended that most patients with CAF should be treated prior to the development of symptoms and major pathological changes in the heart, coronary arteries, and pulmonary circulation.7)

| Table. Reported Cases of Aneurysmal Circumflex Coronary Artery With Fistulous Connection to Coronary Sinus |
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| No. of patients | Author (Year) | Age (years) and sex of patients | Symptoms and complications | Treatment |
| 1 | Perry (1992) | 3.2/F | None | Catheter closure with coil |
| 2 | Perry (1992) | 7/F | None | Catheter closure with coil |
| 3 | Perry (1992) | 9 months/F | None | Catheter closure with umbrella |
| 4 | Phillips (1996) | 68/M | SOB, orthopnea | Operative closure of fistula with CPB* |
| 5 | Chamberlain (2001) | 58/M | IE | Operative closure of fistula with CPB |
| 6 | Rajs (2001) | 48/M | DOE, angina | Died (rupture of aneurysm) |
| 7 | Dogan (2002) | 40/F | SOB, fatigue | No surgical treatment (refused) |
| 8 | Kasravi (2004) | 31/M | IE | Operative closure of fistula with CPB |

*; Mitral valve replacement and tricuspid annuloplasty were simultaneously performed. CPB indicates cardiopulmonary bypass; DOE, dyspnea on exertion; IE, infective endocarditis; SOB, shortness of breath; F, female; and M, male.
Since the first successful ligation of a CAF, several techniques, such as direct proximal and distal ligation, tangential arteriorrhaphy without CPB, direct intracardiac closure of CAFs with CPB, and transcatheter embolization, have been proposed for eliminating these defects while preserving coronary blood flow. Among these techniques, direct intracardiac closure of CAFs with the aid of CPB has been most frequently used because the exact number and localization of fistulas are easily confirmed in the cardiac chamber. Moreover, recent studies have indicated that intracardiac closure of the distal end of CAFs under CPB reduces the likelihood of residual or recurrent fistulas. On the other hand, transcatheter embolization of a CAF has been recently reported as an alternative to surgical therapy. Although several complications such as T-wave inversion on an electrogram, embolization of the coil to the pulmonary artery, and transient arrhythmias were experienced in 30% of patients, complete closure of CAVF was obtained in 78% to 82% of patients by transcatheter embolization. However, a retrospective review to determine the comparative therapeutic efficacy of operation and coil embolization showed that coil embolization was possible, at most, in 6 (38%) of 16 patients and has indicated that the requirements for satisfactory coil embolization of a CAF include the ability to safely cannulate the branch coronary artery that supplies the fistula, the absence of large branch vessels that can be inadvertently embolized, the presence of a single, narrow restrictive drainage site into the cardiac chamber or vessel, and the absence of multiple fistulous connections. According to these results, transcatheter embolization is considered to be a reasonable alternative to standard surgical closure in a selected group of patients.

In conclusion, we have described a case of an aneurysmal circumflex artery connecting to the coronary sinus through a fistula, and emphasize that most patients with CAF should be treated prior to the development of symptoms and major pathological changes in the heart, coronary arteries, and pulmonary circulation by interventional therapy such as catheter embolization or surgical closure using CPB.

**REFERENCES**


