Coronary Artery Fistula between Left Coronary Artery and Coronary Sinus in Newborn

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Abstract

Aneurysmal circumflex coronary artery fistula connected to the coronary sinus is a rare clinical entity that usually remains asymptomatic until later in life. The timing of surgical treatment for asymptomatic patients is crucial. The decision to leave or exclude the aneurysmatic coronary artery following ligation of the fistula is controversial. Herein, we report the successful management of a coronary fistula between the circumflex artery and the coronary sinus without using cardiopulmonary bypass during the newborn period.

Keywords

► aneurysm
► pediatric
► off-pump surgery

Introduction

Congenital coronary artery fistula (CAF) is defined as an abnormal communication between the coronary artery and one of the cardiac chambers or vessels.1 Volume overload due to the fistulous connection may cause marked dilation of the coronary artery, which eventually leads to aneurysm formation.2 In the majority of the patients, the fistula originates from the right coronary artery (RCA). In 35% of the cases, left coronary descending (LAD) artery is affected. True CAF fistula with blood shunting into coronary sinus or its major branches is seldom seen. Moreover, aneurysmal circumflex (Cx) coronary artery with fistulous connection to the coronary sinus is a rare clinical condition.3

Herein, we present, an 8-day-old patient with congenital AV fistula between aneurysmatic Cx artery and coronary sinus requiring early surgical treatment due to congestive heart failure (CHF).

Case

An 8-day-old girl was admitted to the hospital with dyspnea and discomfort. The blood pressure was 70/28 mm Hg, heart rate was 170/min, and respiratory rate was 48/min. In her physical examination, the patient had tachypnea and 2/4 systolodiastolic murmur. There was cardiomegaly in her chest roentgenogram. On ECG, there were rhythm and non-specific ST changes. On echocardiography, VSD with diameter of 3.2 mm in the muscular trabecular area was detected. Although the pulmonary and systemic venous return was normal, there was turbulence in the coronary sinus which was dilated; therefore, angiography was planned. The coronary angiography revealed markedly dilated Cx artery (6.7 mm) connected to the coronary sinus (► Fig. 1). The LAD artery and the RCA were normal. The aortic and pulmonary artery pressures were 65/40 (mean 50) and 51/27 (mean 36) mm Hg, respectively. The Qp/Qs was 4.16. The patient was intubated on the 6th hour after angiography due to CHF. Despite high-dose inotropic treatment, the CHF persisted. Surgical closure of the fistula was planned.

Surgical Procedure

Following median sternotomy, the pericardium was opened. The heart was enlarged and arrhythmic. Bilateral pleural effusion was detected. When the right atrium was pulled toward the left shoulder, the fistula between the Cx artery and the coronary sinus was seen (► Fig. 2). The thrill on the fistula was palpated.

The aforementioned positioning of the heart did not deteriorate the hemodynamic status; hence, the procedure was performed on the beating heart. To avoid narrowing of the coronary sinus, two horizontal mattress sutures with pledgets were passed beneath the distal part of the Cx artery.
Both sutures were tightened with snare. The thrill disappeared. The central venous pressure dropped from 14 to 9 mm Hg. In the intraoperative transesophageal echocardiography (TEE) examination, the turbulence in the coronary sinus was seen to disappear. The left ventricular wall motion was normal. There was also no change in ECG, therefore the sutures were tightened. The loose pulmonary artery band was applied to reduce the increased blood flow which was VSD related. After pulmonary artery banding procedure, the pulmonary artery pressure proximal to the band was measured as 48/24 (mean 32) mm Hg whereas the pressure distal to the band was 39/21 (mean 28) mm Hg. There was no difference between the preoperative and postoperative ECGs. The postoperative course was uneventful. She was extubated at the postoperative 36th hour and was transferred to the ward on the 6th postoperative day. She was discharged on the 11th postoperative day. The control echocardiography at the first month was normal.

Comment

CAF between Cx artery and the coronary sinus is a rare clinical entity that can be complicated by CHF, endocarditis, myocardial infarction, distal embolization or rupture. Although most CAF patients are asymptomatic in the newborn period, they are generally diagnosed later in life with CHF. This is mainly the result of increased blood flow that is shunted from the coronary artery to the coronary sinus in the presence of a concomitant lesion such as VSD. These accompanying cardiac pathologies make early surgical treatment crucial. On the other hand, it is also claimed by some authors that untreated patients with CAF have benign clinical course. Operation is indicated only in symptomatic patients or patients with large shunts, since some of these fistulas are likely to develop CHF and have tendency toward development of infective endocarditis over time.

The goal of surgical treatment is complete obliteration of the fistula and preservation of normal blood flow through the coronary artery. Several techniques, such as direct proximal and distal ligation, tangential arteriography without CPB, direct intracardiac closure of CAFs with CPB, and transcatheter embolization, have been proposed. Direct intracardiac closure with the aid of CPB is an appropriate method since it provides precise localization of the fistula without giving damage to any of the cardiac structures. CAF fistula can be safely closed by suture ligation without CPB in the presence of an easily accessible site. If CAF fistula drains into the coronary sinus, the surgeon should make sure that the ostium of the coronary sinus is not narrowed.

When aneurysmatic dilatation of the coronary artery is present, exclusion of aneurysmatic dilatation of the coronary artery is required. Nakahira et al reported the rupture of aneurysmatic Cx artery draining into the left atria 6 weeks after ligation of CAF fistula. They recommended the exclusion
of aneurysm and revascularization of the remaining branches in patients with aneurismal Cx arteries connecting to the coronary sinus, when there were suitable branches for bypass. When there is no change in the wall motion of the left ventricle in TEE, exclusion of the aneurysm is performed without grafting of branches of the Cx artery. In our patient, excluding the aneurysm and bypassing the remaining Cx artery or its branches would be challenging since the patient was a newborn. On the other hand, the Cx coronary artery aneurysm size was not appropriate for plication or exclusion. Moreover, this patient’s condition was not suitable for cardiopulmonary bypass.

In the review of the patients with CAF fistula, Liberthson reported the mortality as 4%. However, in the presence of giant aneurysm requiring aneurysm excision and establishment of bypass grafting, the risk of ischemia and arrhythmia increases. Therefore, the mortality rate also increases. Transcatheter embolization of CAF has been recently reported as an alternative to surgical therapy. However, the success in obliterating the CAF fistula is low compared with surgical treatment.

To the best of our knowledge, this is the first case of CAF with CHF detected during the newborn period and undergoing successful fistula ligation without use of cardiopulmonary bypass.

Aneurysmal Cx coronary artery with fistulous connection to the coronary sinus is a rare clinical condition. Although there is a case reporting rupture of the aneurysmatic Cx artery after surgical repair of the CAF fistula, suture ligation without bypass grafting can be used in infants and patients who are unsuitable for CPB.

References