Surgical Treatment of a Coronary Artery Fistula With Concomitant Saccular Coronary Artery Aneurysm

— A Case Report —

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An extremely rare case of a coronary artery fistula with a concomitant saccular aneurysm is presented. A 65-year-old woman, who had a history of chest bruising 5 years earlier, suffered from chest pain, which was diagnosed as being due to left coronary artery-pulmonary artery fistulae concomitant with a giant saccular coronary artery aneurysm. Suture closure of the afferent coronary artery to the aneurysm, aneurysmorrhaphy, and transpulmonary closure of coronary artery-pulmonary artery fistulae were performed. The postoperative course was uneventful and the patient was well at 3 months after the operation. Because the risk of surgery appears to be less than the potential development of fatal complications, it is recommended for the treatment of coronary artery fistula with a concomitant saccular aneurysm. (Jpn Circ J 1999; 63: 809–812)

Key Words: Coronary artery aneurysm; Coronary artery fistula; Saccular aneurysm

Coronary artery fistula (CAF), first described by Krause in 1865, is characterized by the involved coronary artery having a normal origin from the aorta, but with a fistulous communication with the atria or ventricles, or with the pulmonary artery (PA), coronary sinus, or superior vena cava. Although CAF has been considered a relatively rare disease, it is being recognized with increasing frequency because of the widespread use of selective coronary arteriography. However, a case of CAF with a concomitant saccular coronary artery aneurysm is quite rare and we report the successful surgical treatment of such a case.

Case Report

Clinical Data
A 65-year-old woman, who had a history of chest bruising 5 years earlier, developed an angina-like chest pain on effort. On physical examination, her heart rate was 68 beats/min and blood pressure was 136/86 mmHg. A high-pitched, continuous murmur of Levine 1/6 was heard from the third intercostal space to the left sternal border. A chest X-ray showed a bulge on the left cardiac border beside the left PA (Fig 1), which a routine chest X-ray taken 3 years earlier did not show. An electrocardiogram showed normal sinus rhythm and no significant ST-T changes. Additionally, no ST-T change was found at the maximum heart rate of 140 beats/min during treadmill exercise testing. An echocardiogram demonstrated a mass, with a maximum diameter of 5 cm and having a septum, located at the left side of the PA. Computed tomography (CT) showed a non-enhanced mass with a maximum diameter of 5.2 cm on the left side of the PA. The proximal left coronary artery was significantly dilated (Fig 2). Magnetic resonance imaging demonstrated a double-contrasted mass with a maximum diameter of 5.4 cm on the left side of the PA (Fig 3). Cardiac catheter data revealed normal pressure values in the right and left heart and no left–right shunt. Selective coronary arteriography demonstrated a contorted CAF with a large saccular aneurysm, which originated from the dilated left anterior descending (LAD) artery (Fig 4). The left coronary artery proximal to the CAF was significantly dilated. Minor fistulae from the right coronary artery were also shown. All the fistulae drained into the main PA.

From these data, the patient was diagnosed as having multiple CAFs with a concomitant large saccular aneu-
rysm, which was considered to have progressively enlarged during the preceding 3 years, and we decided to treat it surgically.

**Operation**

The heart was exposed through a median sternotomy. No thrill was found on any site of the heart surface. A 5.5×5.7 cm coronary aneurysm was located on the left side of the right heart outflow and PA. Although a contorted CAF was easily observed on the heart surface between the LAD artery and the coronary aneurysm, the precise origin could not be confirmed. Therefore, we elected not to cut the fistula at the origin, but instead to cut the connection from inside the aneurysm. Furthermore, because many CAF to the PA had been revealed by the coronary angiography, treatment of these CAF was performed within the draining chamber of the PA to avoid a possible residual shunt.

Following establishment of the cardiopulmonary bypass, the aneurysm was opened with the heart beating. The aneurysm was filled with both old and fresh thrombi, which were removed, and the orifice of a fill-in artery was closed with 4-0 prolene. The aorta was then cross-clamped and cardiac arrest was obtained by antegrade cold blood cardioplegia. We used additional cardioplegic solution to confirm that no remaining artery opened into the aneurysm. Excess aneurysm wall was excised and aneurysmorrhaphy was performed. The CAF draining chamber of the PA was opened and the openings of the fistulae were exposed by instilling cardioplegic solution. A total of 5 orifices (anterior wall of the main PA trunk, 2; posterior wall of the PA, 2; anterior wall of the PA bifurcation, 1) were all closed from within the PA vessel using 4-0 prolene. Because the fistula through the coronary aneurysm had already been cut by this time, all the connections to the PA were considered to have originated from either the right coronary or the left coronary artery, even though the latter was not associated with the coronary aneurysm. The PA wall was closed and weaning from the cardiopulmonary bypass was accomplished without difficulty. Total aortic cross-clamp time was 53 min and the extracorporeal circulation time was 87 min.

**Postoperative Course**

The postoperative course was uneventful. A postoperative CT showed a non-enhanced, low attenuation area in the left side of the PA, where the coronary aneurysm had been. Postoperative coronary angiography demonstrated no CAF. The patient was discharged on postoperative day 22. Pathological examination of the aneurysm showed collagen replacement of intima and media, and degenerative and inflammatory changes in the adventitia wall. The patient was well at 3 months after the operation.

**Discussion**

The incidence of CAF is 0.2% of congenital cardiac anomalies and 0.2–1% of all selective coronary arteriography cases. Although it used to be considered a rare disease, this condition is being diagnosed more often because selective coronary arteriography has become more widespread. The fistula originates from the right coronary artery in 55% of cases, left coronary artery in 35%; both coronary arteries in 5%, and a single coronary artery in 3%. Most of the fistulas’ recipient chambers are in the right heart: right ventricle, 42.5%; right atrium, 34%; pulmonary artery, 15%; left atrium, 5% and left ventricle, 3.5%. Lowe...
et al also reported similar data? The causes of CAF are considered to be congenital, such as the growth of the pulmonary artery pericardial ring or residual myocardial sinusoids.\textsuperscript{2,3} The fistula artery is often dilated diffusely\textsuperscript{9,10} but formation of a saccular aneurysm is quite rare.\textsuperscript{11–15} Generally, the cause of a coronary aneurysm is due to various acquired factors; for example, atherosclerosis, either rheumatic, traumatic, or mycotic.\textsuperscript{16} However, the main causes of coronary aneurysms concomitant with CAF are suspected inflammation, injury, or stenosis with atherosclerotic change.\textsuperscript{4} In the present case, we speculate that the congenital CAF were already extant and the coronary aneurysm was secondary, probably due to the previous chest bruising. Unfortunately the pathological findings could not prove the relationship between the chest bruising and the aneurysm.

It is commonly stated that most patients with CAF are asymptomatic.\textsuperscript{2–5} Congestive heart failure, dyspnea, fatigue,\textsuperscript{17–19} angina or chest pain due to acute myocardial ischemia\textsuperscript{10,19} and bacterial endocarditis\textsuperscript{21} have been reported as symptoms of CAF. Additionally, rupture of an aneurysm,\textsuperscript{11,12,15}16 and pulmonary hypertension\textsuperscript{17,18} have been reported rarely. The ‘coronary steal phenomenon’, which leads to coronary dysfunction, is reported in 3–7% of cases.\textsuperscript{4,6,11,12,22} In the present case, the patient complained of chest pain, which might have been associated with the enlargement of the aneurysm because no myocardial ischemic change was seen on the electrocardiogram.

Regarding the surgical indications, Konno et al.\textsuperscript{3} suggested the following criteria: (1) shunt rate >30%, (2) ischemic or loaded change on ECG, (3) progression of the pulmonary hypertension or congestive heart failure is anticipated, (4) history of ischememia, (5) morphological saccular type aneurysm, and (6) social reasons (eg, loss of employment opportunity due to existing heart murmur). With current open heart surgery techniques, the surgical risk in most cases appears to be considerably less than the potential development of serious and fatal complications.\textsuperscript{4} Furthermore, the reports of recovery from coronary dilatation and contortion after surgery support the early and aggressive employment of surgical treatment.\textsuperscript{24} In the present case, the patient complained of chest pain, which might have been associated with the enlargement of the aneurysm because no myocardial ischemic change was seen on the electrocardiogram.

The purpose of the operation is to close the shunt, maintain adequate coronary blood flow, correct concomitant cardiovascular lesions and prevent any life-threatening complications. Coronary ligation, selective fistula ligation, tangential arteriography,\textsuperscript{25} Symbas’s method,\textsuperscript{26} fistula closure from the intracardiac space,\textsuperscript{27} aneurysmorrhaphy or arteriorrhaphy\textsuperscript{28} and coronary artery bypass\textsuperscript{28} have been reported as suitable surgical procedures. Because the dissection of the CAF in the present case was difficult without damaging the normal coronary supply to the heart, we performed suture closure of the afferent coronary artery to the aneurysm, aneurysmorrhaphy, and transpulmonary closure of coronary artery-PA fistulae. When the draining chamber is the right atrium or PA, as in the present case, fistula closure from the draining chamber is a reasonable method because the fistula opening is clearly detected. However, where identification of the CAF opening is difficult, this method may leave residual CAF. Although the operation can be done without cardiopulmonary bypass, we believe it is preferable in most instances. Myocardial preservation during the cross-clamping period is extremely important. Fortunately, we were able to obtain cardioplegia arrest by using cardioplegia solution and by quickly closing off the shunts of coronary flow. However, in situations where an antegrade cardioplegia does not provide adequate myocardial protection, retrograde cardioplegia should be employed.

**Conclusion**

We surgically treated a case of coronary artery-PA fistulae concomitant with a giant saccular coronary aneurysm in a patient who had a history of chest bruise. We performed suture closure of the afferent coronary artery to the aneurysm, aneurysmorrhaphy, and transpulmonary closure of the coronary artery-PA fistulae. Postoperative CT and coronary angiography showed no communication of coronary-PA or coronary artery aneurysm. The patient remains well 3 months after the operation.

Since the surgical risk appears to be less than the potential development of fatal complications, surgical treatment is recommended.

**References**


