

Surgical Repair of Congenital Coronary Artery Fistula With Giant Aneurysm

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ABSTRACT

A 67-year-old man with a 3-year history of dyspnea on exertion arrived to our institution with discontinuous palpitations and short breath. He was diagnosed with congenital coronary artery fistula (CAF). Angiography revealed 3 giant aneurysmal formations and coronary artery calcification. We report a case of successful repair of CAF with a giant coronary aneurysm by closing the orifice and resecting the aneurysm and reconstructing the left coronary artery. The surgical procedure included closure from within a vessel dilated by aneurysm and excision of the aneurysm. We were able to completely obliterate the fistula and preserve the normal blood flow through the coronary arteries post operation. The postoperative course was eventful, but the patient was discharged home. The patient was doing well at his 28-month follow-up visit.

INTRODUCTION

Coronary artery fistula (CAF) is a rare congenital anomaly with a prevalence of 0.27% to 0.4% in the population with congenital heart disease [Hashiyama 2017; Karazisi 2017]. In recent years, the detection rate of CAF is much higher, due to the widespread use of coronary angiography and the majority of patients received treatment in childhood. Only a few reports were found describing the surgical management in older patients. Surgical ligation of fistula followed by endocardial or epicardial closure remain the most common treatment methods. However, the long-term results were not found to be satisfactory, particularly in the patients with severely dilated coronary artery (CA). Abnormality of the CA morphology led to a series of complications post CAF closure. This includes thrombosis, myocardial infarction, and cardiac dysfunction [Shah 2016; Yim 2015; Wang 2015]. Also, we could not find any report or data suggesting a guideline for indication or strategy of CA reconstruction. Thus, in the present case report, we prove that repair of CAF with a giant coronary aneurysm was feasible and have made it accessible for reference for future practice.

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CASE REPORT

A 67-year-old man with a 3-year history of dyspnea on exertion arrived to our institution with discontinuous palpitations and short breath. The patient was in New York Heart Association functional class III on admission. Echocardiography revealed congenital CAF with a left ventricular cavity measuring 6.0/4.5 cm in size. The right atrium (RA) measured 5.8 cm, and the left ventricular ejection fraction (LVEF) was 43%. Multislice computed tomography imaging demonstrated that the CAF originated from the sinus node artery and drained into the RA. Orificium fistulae measured 14.2 mm in size. The left main coronary artery (LM) was remarkably dilated with a diameter of 22.5 mm. The left circumflex artery (LCX) was aneurysmal in its proximal segments, presenting a giant aneurysm (43*16 mm). The calcification in the CA was obvious, especially in the aneurysmal wall and dilated segments (Figure 1). Angiography further revealed 3 giant aneurysmal formations in the left

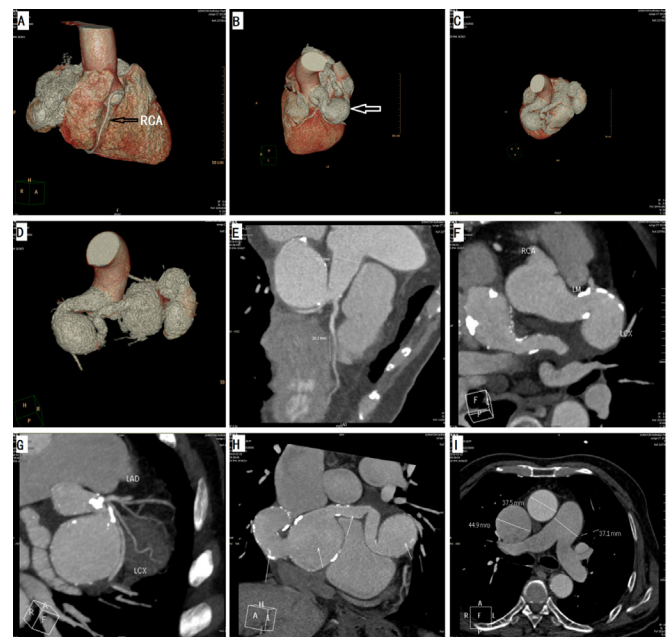


Figure 1. Multislice computed tomography (MSCT) imaging before the operation. A, The RCA was normal; B-D, Aneurysmal formations in LCA; E-H, Calcification of CA was obvious, especially in the aneurysmal wall and dilated segments; I, The superior vena cava was much widened (44.9 mm) because of fistulous vessel compression.

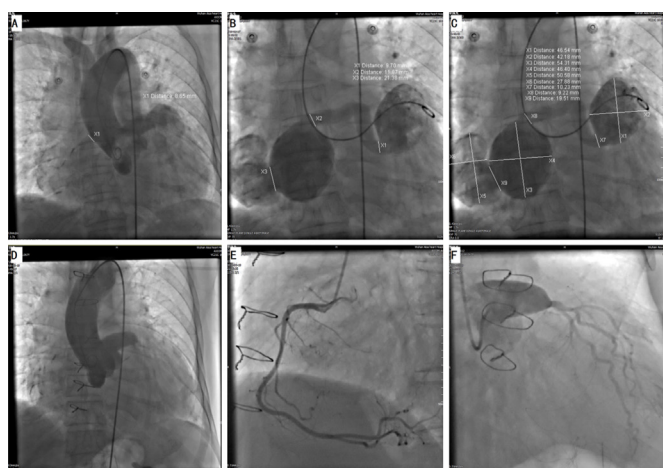


Figure 2. A-C, Preoperative angiography: 3 giant aneurysmal formations in LCA. D-F, Postoperative angiography: diameter of LM was much smaller than preoperative size. The coronary circulation of RCA and branch of LM was normal.

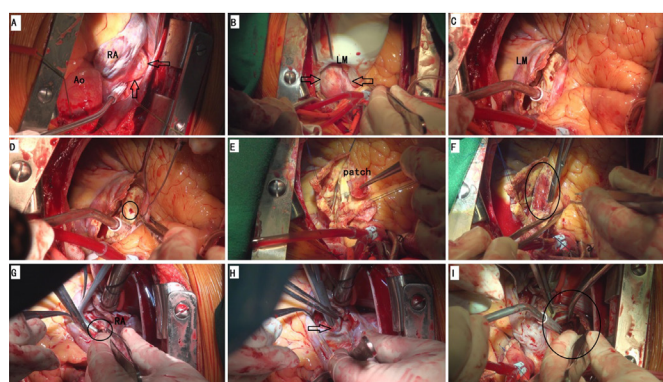


Figure 3. A, Formation of giant coronary aneurysmal; B, The LM was remarkably dilated with a diameter of 22.5 mm; C-D, LM was transected and the branches were explored; E-F, Orificium closure and reconstruction of LCA with autologous pericardial patch; G-H, Incision of the RA and probing the distal orificium; I, Fistulous vessel near interatrial groove was incised and a little thrombus was detected; C and I, Severely calcified aneurysmal wall.

coronary artery (LCA) measuring 47*37 mm, 55*46 mm and 49*35 mm, respectively (Figure 2). Written informed consent was obtained from the patient for publishing and using the images and description of his case for academic purposes.

The guidewire could not pass through the fistulous vessel, and hence the transcatheter closure was abandoned. The patient subsequently underwent an operation and the surgical exposure was facilitated by cardiopulmonary bypass (CPB) with median sternotomy. The patient signed informed consent and underwent the treatment procedures. Intraoperative findings revealed that the fistula arose in the anterior segment of LM. Later, the fistulous vessel traveled along the interatrial groove to reach superior vena cava (SVC). Finally, the fistula entered into the RA at the junction site with SVC. The diameter of the fistulous vessel ranged between 4 and 5 cm with the segment

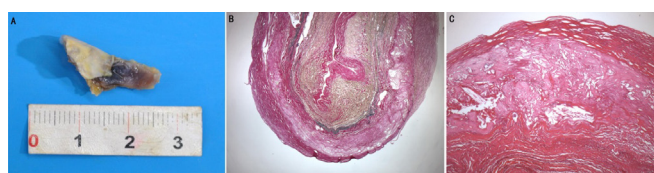


Figure 4. A-C, Staining of the fistulous vessel: atheromatous plaque and calcification in endarterium; tunica media was thin with several layers of elastic fiber; edema and hemorrhage in the tunica adventitia.

near the top wall of the LA measuring 1 cm. The surgical procedure includes endocardial closure of CAF and reconstruction of LCA. First, we exposed the fistula by transecting the proximal end and made an arteriotomy. Then we closed the orificium and separated the LM and LAD to the aorta side with an autologous pericardial patch. The proximal portion of the fistulous vessel was put aside. Finally, we made an incision in the RA and closed the distal orificium with a 4-0 prolene suture. Further, we incised the fistulous vessel near the interatrial groove to confirm that no other orificium was present (Figure 3). The entire surgical procedure was completed in 390 min that included 147 min of CPB and 99 mins of aortic cross-clamp time. (For detailed procedure refer to the supplement material). Postoperative transesophageal echocardiography revealed complete obliteration of CAF and EF of 52%. The electrocardiogram (ECG) indicated that there was no myocardial ischemia. The pathological results were recorded (Figure 4).

Hemostasis procedure consumed most of the operation time as bleeding was the major postoperative problem, due to the vessel walls' calcification. The volume of the chest tube drainage during the first 2 hours was 1500ml in the intensive care unit. We carried out thoracotomy for hemostasis, but did not find it effective. The patient was infused with 10 unit red cells, 4 unit platelets, 1300 mL plasma, and 14 unit cryoprecipitate, respectively. The chest tube drainage gradually decreased and stabilized on the third day post operation. However, several other complications occurred, including renal failure, severe infection, arrhythmia, hypoxic-ischemic encephalopathy, respiratory failure, and gastrointestinal dysfunction. Thus, the patient's postoperative course was eventful. We reexamined angiography, which showed normal coronary circulation and the diameter of LM was much smaller compared to the preoperative size (Figure 2). He was discharged home in approximately 4 weeks after the procedure and was doing very well at the 28-month follow-up visit. ECG taken at the time of follow-up revealed a reduction in the size of the left ventricle with an EF of 52%. The hepatic, renal and respiratory functions remained normal and no myocardial ischemia and thrombus incident occurred.

DISCUSSION

The first successful surgical treatment for CAF was reported in 1958. The goal of treatment is to obliterate the fistula completely and to preserve normal flow through the coronary arteries [Hou 2014]. The different surgical procedures that followed include independent distal ligation, proximal and

distal ligation, ligation and bypass grafting, endocardial or epicardial closure with or without CPB [Thakkar 2015; Zhang 2015]. Every procedure aimed to not obstruct coronary circulation. However, the optimal surgical technique was chosen, depending on the coronary vessels becoming aneurysmal, points of termination, and the origin of the fistula. Aneurysmal formation was detected in 14%~20% of the population with CAF, including both pediatric and adults [Gurkan 2015; Bolukcu 2017]. However, the incidence was found to be higher in adults than in children. Dealing with aneurysm still remains controversial to the best of our knowledge. Surgical ligation or endocardial closure frequently was the chosen treatment method in the past [Jamali 2016; Speedie 2016]. The majority of the clinics were prone to not deal with aneurysmal formations. The current study showed that the long-term outcome of CAF closure may not be satisfactory as we expected. The possible complications include residual leak, coronary dilatation, thrombosis and myocardial infarction [Welisch 2016]. Thrombosis is the common and severe complication that occurs after CAF closure. It is closely associated with severely dilated CA and aneurysm. In our present report, we treated an isolated CAF patient with 3 giant aneurysms by closing the orifice, resecting the aneurysms, and reconstructing the LCA. Thus, we were able to reduce the thrombosis risk to a greater extent and expect long-term prognosis to improve. The postoperative course was eventful, but the patient stabilized and was doing well at his follow-up visit. Further detailed researches are necessary to understand if revascularization outweighs the risk for other patients with the same condition.

CONCLUSION

Treating CAF with dilated CA or aneurysmal change surgically either by ligation or closure of fistula might lead to complications, such as thrombosis and myocardial infarction. Our study confirms that it is possible to resect the aneurysm and reconstruct the coronary artery to obtain an improved outcome in the long term. Postoperative hemorrhage being the primary problem, the technique of vascular anastomosis for calcified vessel still remains a challenge.

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