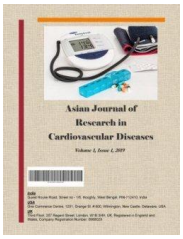

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Surgical Treatment of a Coronary Artery Fistula that has Opened into the Pulmonary Artery

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Author's contribution

The sole author designed, analysed, interpreted and prepared the manuscript.

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Case Report

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ABSTRACT

Introduction: Coronary artery fistula (CAF) is a rare cardiac malformation of congenital, traumatic or iatrogenic origin, and is an abnormal connection between the coronary arteries and the cardiac chambers or the great vessels. The incidence in the general population is estimated to be approximately 0.002%. CAF may be asymptomatic or complicated by congestive heart failure, myocardial ischemia, infective endocarditis, fistula rupture or thrombosis.
Objective: Coronary artery fistulas are very rare malformations, and they need to be kept in mind as a possible cause of myocardial ischemia in patients who present with fatigue, dyspnea, and chest pain.
Case Presentation: A symptomatic patient with CAF and fistulous aneurysm between the left anterior descending artery and the pulmonary artery, treated successfully with cardiopulmonary bypass on arrested heart is presented.
Discussion: Small fistulas can close spontaneously, but larger and symptomatic fistulas may require closure by surgical or transcatheter methods. Surgical treatment should be considered in patients who are symptomatic, have significant aneurysmal enlargement, and in whom transcatheter occlusion methods have failed.
Conclusion: In patients with CAF, early surgery is a good option to avoid potential complications, and surgical closure can be performed with very low morbidity and mortality rates.

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Keywords: Coronary artery fistula; coronary aneurysm; coronary angiography; cardiopulmonary bypass.

1. INTRODUCTION

Coronary artery fistula (CAF) is an abnormal connection between the coronary arteries and the cardiac cavities or great vessels such as the vena cava, the pulmonary artery (PA), and the pulmonary vein. It is a rare cardiac malformation identified incidentally in less than 1% of routine cardiac angiography series, and may have a congenital, traumatic or iatrogenic origin [1]. While previous studies have reported that the right coronary arteries (RCA) account for approximately 55% of CAF cases [2,3], some authors have stated that the left anterior descending artery (LAD) is responsible for most cases of CAF [4,5]**.** The fistulas most commonly open into the right ventricle, right atrium, PA, and coronary sinus, which are low-pressure chambers. In a previous report, the PA was reported to be the most common site of drainage, seen in 86% of the patients [5].

Coexistence of CAF and large aneurysms is rare. It has been reported that 14% to 26% of patients with CAF have coronary artery aneurysms. CAF may be asymptomatic or may be complicated by congestive heart failure, myocardial ischemia, myocardial infarction (MI), or even sudden death. Small fistulas can close spontaneously, but larger and symptomatic fistulas require closure by open surgical or transcatheter techniques [4,6,7]. Treatment options for CAF vary depending on the individual. Surgical treatment should be considered in patients who have symptoms, have significant aneurysm enlargement, and in whom catheter closure has failed.

A symptomatic patient with congenital CAF associated with an aneurysm originating from the proximal LAD and draining into the PA is herein presented. Because a transcatheter intervention failed, the CAF was repaired by cardiopulmonary bypass (CPB) on arrested heart. This case report was written according to the SCARE guidelines [8].

2. CASE REPORT

A 55-year-old female patient complaining of fatigue, shortness of breath (NYHA-Class III-IV), and chest pain for two months was diagnosed with CAF at another center. A transcatheter embolization closure of the CAF failed, and the patient was referred to our hospital for surgery. Her medical history and examination were unremarkable. There were nonspecific ST-T wave changes on electrocardiography. Physical examination showed a rhythmic pulse of 98 beats per minute and a blood pressure of 147/72 mm Hg. The cardiothoracic ratio was normal on the posteroanterior chest radiograph. Laboratory tests were also normal (WBC: $9.7x10^3/mm^3$; Hematocrit: 39,2 %; Hemoglobin: 12.7 g/dl; PLT: $273x10^3/mm^3$; LDH: 193 U/l; BUN: 21 mg/dl; Creatinine: 0.9 mg/dl). Total cholesterol was 154 mg/l, LDLc was 87 mg/dl, and HDLc was 40 mg/dl, according to the lipid profile. Troponin-I level was 14.9 ng/l. Echocardiography revealed vascular structures draining into the main PA and causing turbulence, just above the pulmonary valve. Ejection fraction was 60%, and PA pressure was 35mm Hg. Coronary angiography and multislice computed tomography (CT) showed the CAF and its connections originating from the proximal LAD and draining into the PA. There was also an aneurysm at the fistula area, measuring 2 x 2.5 cm. The LAD was dilated (8 mm) and tortuous at the proximal CAF. It was noted that the CAF was also supplied by the conal artery (Fig. 1, Fig. 2). No atherosclerotic lesions were seen in the coronary arteries. Qp/Qs ratio was 1.7. The patient was fully informed of the surgery and informed consent was obtained.

2.1 Surgical Technique

Under general anesthesia, a median sternotomy was performed and the pericardium was opened. Next, the CAF that occupied a wide area on the PA, and the aneurysm connected to the fistula were visualized (Fig. 3). A thrill was palpated over the pulmonary artery. Systemic heparinization was made, and CPB was initiated by aorta bicaval cannulation. After placement of the aortic cross clamp, cardiac arrest was made by antegrade blood cardioplegia. Under direct vision, the CAF connections were seen. Once the LAD and fistula were exposed, the CAF was divided using a 6/0 polypropylene suture. Then the aneurysmatic fistula segment was resected. A 2-3 cm horizontal cut was made on the pulmonary artery just above the pulmonary valves. The drainage point of the fistula was found, and the fistula opening was closed primarily with 5/0 prolene sutures. The pulmonary arteriotomy was repaired. The CAF

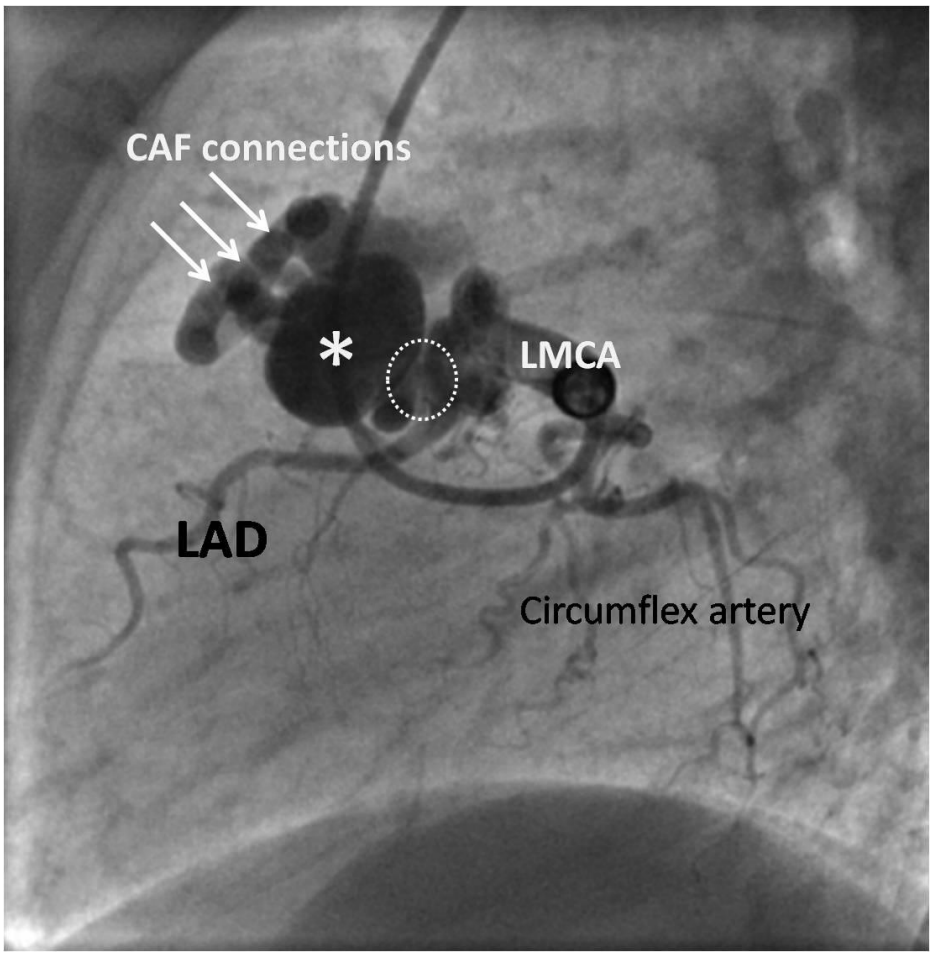

Fig. 1. Coronary angiography

Coronary angiography reveals a CAF, a fistula aneurysm (asterisk), CAF connections (arrows), and the location of the fistula (dashed circle). LMCA: Left main coronary artery; LAD: Left anterior descending artery; CAF: Coronary artery fistula

and all its connections were successfully divided under CPB. Weaning from the CPB was uneventful and under sinus rhythm. There were no intra and postoperative ischemic changes. The cross-clamp time was 27 minutes, while the CPB time was 35 minutes. The patient was extubated during the fourth postoperative hour and transported to the ward after spending one night in the critical care unit. The patient was discharged uneventfully on the 5th postoperative day with antiaggregant treatment (acetylsalicylic acid 100 mg 1x1, clopidogrel 75 mg 1x1). No residual fistula was observed in the follow-up CT angiography performed one month after the operation. The patient is still under follow-up without any problems.

3. DISCUSSION

Most coronary artery anomalies are discovered incidentally during coronary angiography or radiologic studies performed for the assessment of cardiac symptoms. Patients with CAF often do not show any symptoms in the early stages of life, especially during the first two decades. With advancing age, symptoms emerge and the frequency of associated complications increases. Dyspnea on exertion is one of the most common symptoms in patients with CAF. Patients may present with a variety of clinical conditions such as congestive heart failure, pulmonary hypertension, bacterial endocarditis, atherosclerotic changes, and thromboembolic events [1,4]. The first sign may be a rupture and the diagnosis is made intraoperatively, as in a patient without preoperative imaging who requires emergency surgery [9]. A ruptured aneurysm may manifest clinically as chest pain, pericardial effusion, cardiac tamponade, or even sudden cardiac death, and may mimic acute aortic dissection [10,11]. The risk factors for rupture of congenital aneurysmal fistulas are

female gender, saccular aneurysm, Asian ethnicity, left coronary origin of aneurysmal fistula, and hypertension [12]. Angina and myocardial infarction are rare in these patients, and ischemic symptoms are thought to be due to coronary artery steal [4,13]. Reitz et al. [14] performed a PA - circumflex coronary artery shunt model with a pulmonary to systemic flow ratio of 1.1. The proximal coronary flow increased by 211% while the distal coronary flow decreased by 26%. However, it was observed that these flow changes in the coronary arteries did not have a significant impact on the measured left ventricular function.

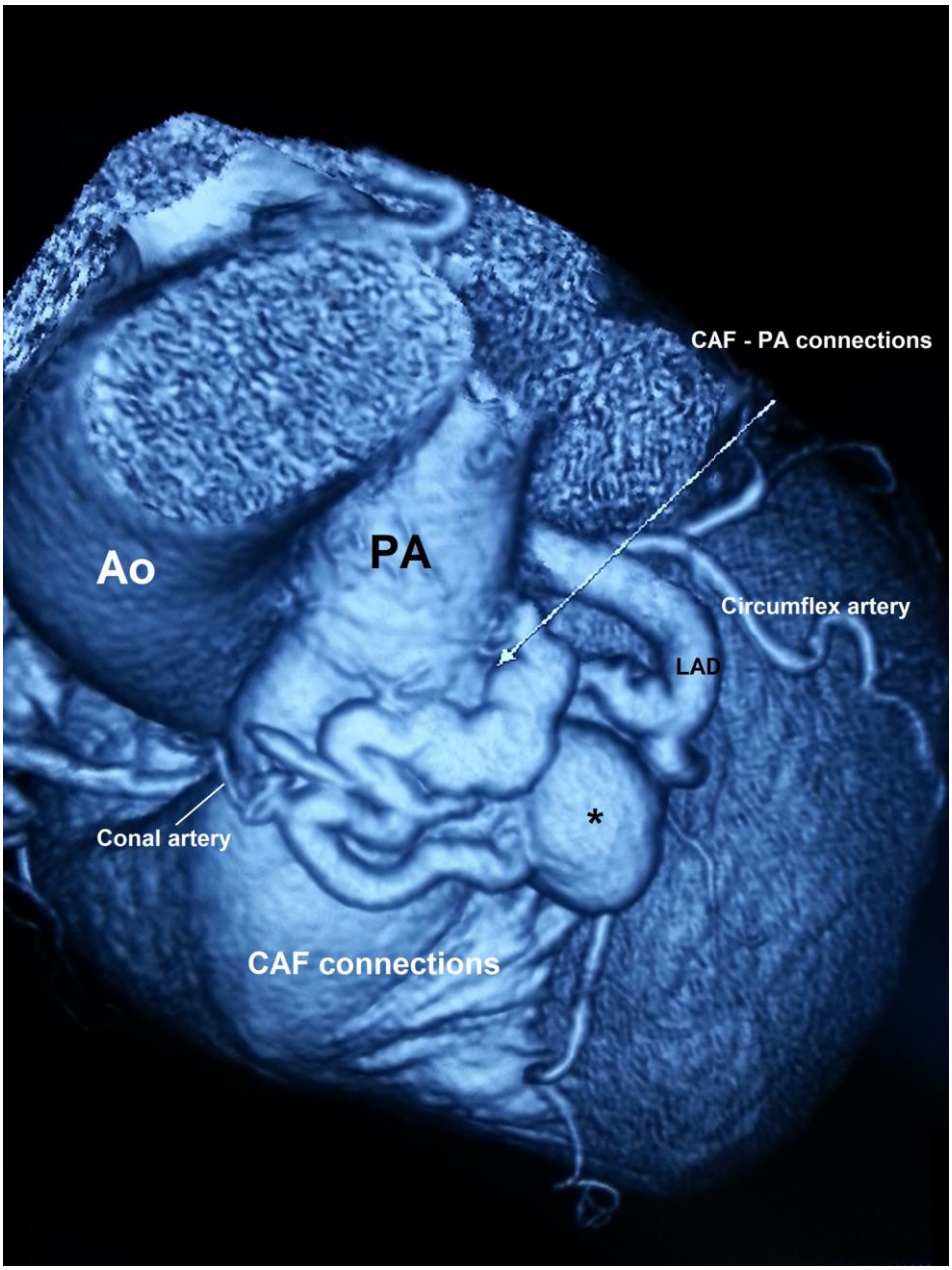

Fig. 2. Multislice-Computed Tomography

Multislice-Computed Tomography shows the formation of CAF and aneurysm (asterix) originating from the proximal left anterior descending artery (a). The opening of the fistula to the pulmonary artery (dashed arrow) (b). Ao: Aorta; PA: pulmonary artery; LAD: Left anterior descending artery; CAF: Coronary artery fistula.

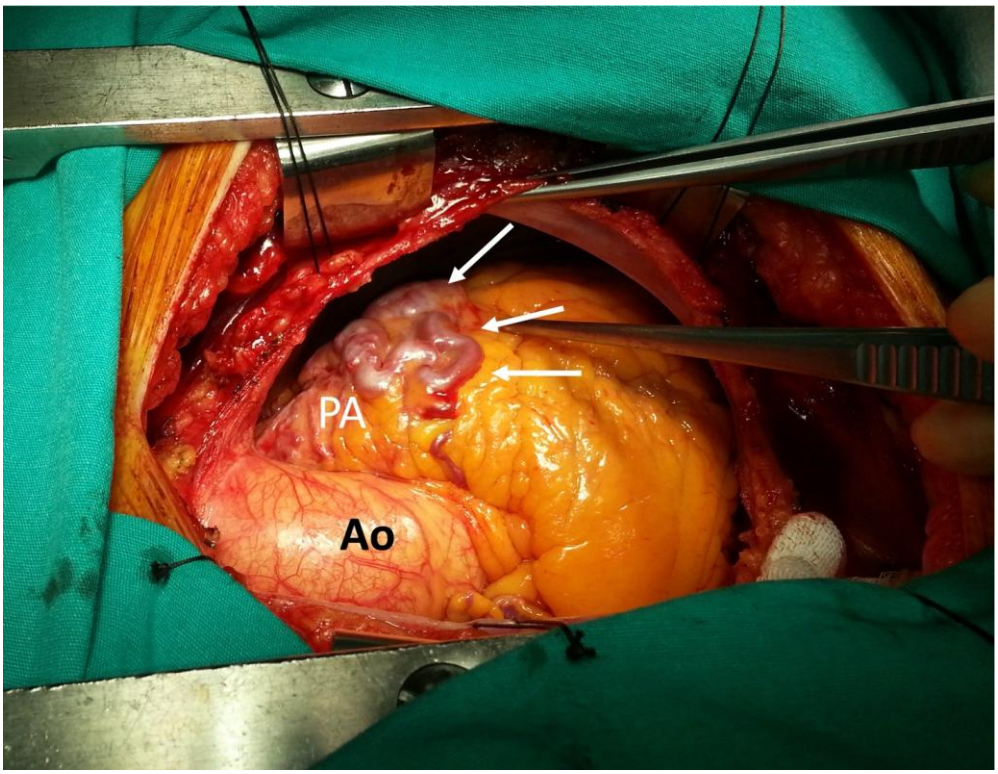

Fig. 3. Intraoperative view of the CAF

The CAF forms dense varicose vein-like clustering on the pulmonary artery (arrows). Ao: Aorta; PA: pulmonary artery

Asymptomatic patients can be treated conservatively, whereas early surgical treatment should be considered if the aneurysm shows progressive dilation during the follow-up period [15]. On the other hand, Lowe et al. suggested that surgical correction should be considered in all patients with severe CAF and that a treatment should be made electively before symptoms and pathologic changes are obvious in the heart [3]. However, certain reports indicate that the decision of an operation should be based on the volume shunt resulting from the fistula (Qp/Qs ratio ≥ 1.5:1) [4]. The location and size of the fistula should also be considered. Surgery may also be considered when myocardial ischemia and/or heart failure occurs due to coronary flow steal and / or shunt [16].

Although small fistulas can close on their own, larger or symptomatic fistulas should be treated to prevent complications. Treatment options for CAF include surgical ligation and percutaneous catheter closure. The surgical approach to CAF may be performed with off pump or with the use of CPB, and in combination with other cardiac surgery procedures if needed. Transcatheter closure is a less invasive approach preferred in cases where the anatomical features of the fistula are suitable and there are no additional cardiac pathologies that require intervention [4,5,7,12]. The selected modality should be based on the anatomical location of the CAF and the concomitant heart conditions that require treatment. In the patient here, the catheter closure method failed. Surgical treatment was preferred due to the presence of multiple fistulas with a large fistula aneurysm, drainage in the PA, and the patient's complex anatomy. The presence of symptoms and a Qp/Qs ratio greater than 1.5 were other important reasons for proceeding with surgery. CABG was not performed in the patient because there was no coronary artery disease, and the fistula and coronary artery boundaries were clearly defined. We did not find any ischemic findings either during surgery or in the postoperative period. Postoperative echocardiographic asessment showed that there were no motion defects in the left ventricular wall.

4. CONCLUSION

Small fistulas can close spontaneously, but larger symptomatic fistulas may require surgical

or transcatheter intervention closure. The location and size of the fistula, along with the resulting shunt volume must also be taken into consideration in treatment planning. The decision as to whether surgical ligation or interventional closure methods are appropriate should be made on a case-by-case basis. Surgical correction should be considered in symptomatic patients with CAF, in patients who have failed transcatheter closure methods, have significant aneurysm enlargement and progressive dilation of the aneurysmal fistula. In these cases, early surgery is a good option to avoid potential complications, and surgical closure can be performed with very low morbidity and mortality rates.

CONSENT

The author declares that 'written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office/Chief Editor/Editorial Board members of this journal.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

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COMPETING INTERESTS

Author has declared that no competing interests exist.

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