Coronary artery fistulas (CAFs) are defined as abnormal communications between a coronary artery and a cardiac chamber or major vessel, such as the vena cava, right or left ventricle, pulmonary vein, or pulmonary artery [1]. CAF was first described by Krause in 1865 and its prevalence based on coronary angiography and echocardiography studies is about 0.1 to 0.8% [2-5]. While 5 to 30% of CAFs are associated with a congenital anomaly, the true incidence is difficult to evaluate because about half of the cases may be asymptomatic and clinically undetectable.

In patients undergoing coronary angiography (CAG) for various reasons, the coronary-pulmonary artery fistula (CPAF) has been reported to be approximately 15–30% of all CAFs, and multiple fistulas were reported to occur in approximately 10.7–16% [6]. CPAF is a subgroup of CAF and more rare than CAF, detected in 0.1% to 0.2% of CAG.

Coronary angiography has been known to be the standard modality for the diagnosis of CPAF. However, coronary fistulas drain into low-pressure chambers of the heart, which may not be well visualized with conventional angiography because of significant dilution of the contrast medium [7].

Recently, coronary MDCT has become a widely used technique that has proven useful to evaluate the cardiovascular anatomy and anomalies [8]. MDCT provides very useful information for treatment planning of CPAF in either surgery or transcatheter embolization; it provides information of the presence or absence of bilateral origins, accurate size of aneurysmal dilatation, and the drainage site of the fistula.

Clinical presentation of CPAF depends on factors such as the age of the patient, amount of shunting, development of cardiac ischemia, and resistance of the recipient vessel or chamber. Nonetheless, most patients are asymptomatic. In patients presenting with symptoms, chest pain has been shown to be the most frequent symptom. In addition,
fatigue, chest tightness, angina pectoris, dyspnea, rupture or thrombosis of the fistula, arrhythmia, pulmonary hypertension, arterial aneurysm, congestive heart failure, infective endocarditis, sudden death and development of myocardial ischemia or infarction (presumably resulting from the coronary steal phenomenon). Frank congestive heart failure is a frequent complication, especially in patients above 40 years of age [9].

However, the optimal strategy to manage symptomatic patients still remains controversial due to the lack of long-term clinical follow-up. Conservative medical treatment remains the major treatment for most CPAF patients with limited symptoms/signs [1].

CASE REPORT

A 61-year-old female had a history of regularly controlled hypertension and hyperlipidemia for 2 years. Her health examination of the coronary artery showed a calcium score of 193 indicating a possibly significant narrowing with at least moderate atherosclerotic plaque (90-95%). She suffered from aggravated effort angina and was referred to our cardiovascular clinic to survey coronary artery disease. Her treadmill revealed positive and echocardiography had no obvious regional wall motion abnormality. The patient was placed on a regimen of aspirin, bisoprolol and isosorbide mononitrate, but since her situation did not improve, coronary angiography was indicated for unstable angina.

Coronary angiography revealed a few insignificant lesions and two macro-fistulas which originated from the proximal segment near the ostium of the RCA and branch of mid segment of LAD, complicated by a large saccular aneurysm (15 mm x 17 mm in diameter) and draining into the pulmonary trunk (Fig. 1). The right side fistula was larger than the RCA and left side fistula, and the patient’s typical ischemic symptoms were most likely caused by the coronary steal phenomenon secondary to the fistulas. After we discussed the therapeutic strategies with the patient and her family, they chose coil embolization of the right side fistula rather than surgical closure of the bilateral fistulas or cover stent.

Before intervention therapy, the chest CT used to survey coronary artery ostium in the normal position. An enlarged arterial fistula was found arising from the proximal RCA connected to a vascular pouch about 1.7cm in size in front of the pulmonary trunk. Another fistula was noted arising from the proximal RCA connected to another vascular pouch above the previous one, with both of these vascular structures draining into the pulmonary trunk (Fig. 3a).

Transcatheter closure was performed via the right femoral artery access with a 7 French (Fr) sheath, and the RCA ostium to the CPAF was engaged with a 7 Fr JR5 guiding catheter (Medtronic, USA), and a floppy guidewire (Runthrough NS®) was inserted into the aneurysm of the CPAF. We attempted to advance the wire to the pulmonary artery, but it failed to get into the distal small orifice of the large aneurysm. The delivery catheter, a 2.6 Fr x 150 cm Excelsior 1018 microcatheter guide (Boston Scientific, USA), was gently advanced under fluoroscopic guidance over a floppy wire directly to the aneurysm of the fistula in order to occlude the drainage orifice and minimize the risk of embolization. Because of the high resistance force due to the severe distortion of the fistula, the guiding catheter, microcatheter and wire was dislodged three times. The buddy wire technique to the RCA and fistula also failed. Fortunately, the deep seating technique successfully advanced a 5 Fr JR5 guiding catheter (Boston Scientific, USA) to the second curve with good support and the microcatheter was positioned in the aneurysm of the fistula. Two GDCs (Guglielmi detachable coils), 14mm x 30cm coil (Boston Scientific, USA) were first deployed within the aneurysm sac, to secure the distal fistula orifice, in order to prevent future distal coil migration (Fig. 2a, 2b). After delivery of the device, coronary angiography confirmed the partially successful occlusion of the fistula with less visible residual flow from the fistula. Another additional twenty microcoils (stainless steel MWCE® 18S-6/2-TORNADO; 6-2 mm) were deployed sequentially and completely obliterated the RCA fistula vessel (Fig. 2c, 2d). Following the above procedures, the coils were implanted so as to merge together forming a conglomeration. Coronary angiography was repeated to confirm the quality of the occlusion, and heparin (60 U per kg) was given during the procedure. There were no ECG changes noted that might suggest myocardial ischemia following the procedure, no procedural complication occurred and the patient was discharged on the following day. The patient subsequently reported that her earlier clinical symptoms had improved, and that her recovery continued uneventfully. After one year, the chest CT was followed up and revealed the status of the post embolization of the coronary CPFA. There was no obvious shunting flow from the right coronary artery, but there was still some shunting flow from the left coronary artery (Fig. 3b-3d).

DISCUSSION

Most CAFs have been reported from studies on Caucasian people. The first report of Oriental CAF patients described incidence (0.4%), observed that 58% of CAFs originated from the left anterior descending artery and 29% from the right coronary artery, and noted that most CAFs (63%) drained to the pulmonary artery [10]. Most CAFs manifest as a single fistula and cases of multiple fistulas are generally rare.

The morphologic features of CPAFs are different from CAFs. The origin of CPAFs was 29.4% in the left coronary artery, 11.8% in the right coronary artery and 58.8% in both coronary arteries; and 29.4% cases were associated with aneurysm. Fistulas were located primarily in the left
Coronary pulmonary artery fistula

anterolateral aspect of the pulmonary trunk (82.3%) and mostly the drainage site was the left lateral side of the pulmonary trunk (82.3%) [11].

Coronary-pulmonary artery fistulas may appear as isolated anomalies in most cases, or they are associated with diverse patterns of congenital heart diseases, such as left or right ventricular hypoplasia [12]. In acquired cases, they are associated with the complications developed after

Figure 1. Coronary angiography showed two tortuous fistulas complicated by a large saccular aneurysm (15 mm x 17 mm in diameter) and drained into the pulmonary trunk. a. Left anterior oblique (LAO) 40 degree and Cranial 20 degree; b. Right anterior oblique (RAO) 40 degree and Cranial 15 degree) which originated from the proximal segment near ostium of the RCA. c. RAO 30 degree and Caudal 30 degree; d. RAO 0 degree and Caudal 38 degree) which originated from the branch of the mid segment of the LAD.
cardiac operation, secondary to endomyocardial biopsy, chest irradiation, chest trauma, percutaneous intervention, and myocardial infarction [13]. The CPAF of this patient was likely congenital, given that she denied previous invasive procedures or history of chest trauma.

In the past, coronary angiography was used primarily for the diagnosis of CPAF. Recently, as multidetector computed tomography (MDCT) has become widely used in coronary artery diseases, the diagnosis of CPAF by MDCT is gradually on the increase. There are several case reports of CPAFs diagnosed by MDCT [13]. It has been shown that 19–29% of CPAF cases are associated with aneurysmal...
Coronary pulmonary artery fistula
dilatation, and most of them are of the fusiform type, with the saccular form being rare [14]. Aneurysmal dilatation of the CPAF is explained to be due to the turbulent flow and arteriosclerotic change caused by acquired inflammation, trauma, kinking, and stenosis [14].

No defined risk factor has been reported for predicting the rupture of an aneurysm [15]. Surgery is required for cases developing aneurysmal dilatation because of the risk of rupture and embolism in CPAF. Although some studies have reported that cases with coronary artery aneurysm larger than 3 cm in size are an absolute indication for surgery [14], there is still a debate in the treatment of aneurysm in CPAF.

Clear guidelines for the treatment protocol for CPAF have not been established yet. According to Liberthson et al. [16], if patients are treated prior to the age of 20 years, the operative mortality (1%) and the complication rate (7%) are low. If they are treated after the age of 20 years, the possibility of developing significant postoperative mortality (7%), postoperative myocardial infarction (7%), and other

Figure 3.

3a. Chest CT showed bilateral arterial fistula connected to a vascular pouch about 1.7 cm in size front to the pulmonary trunk draining into the pulmonary trunk. c-d. The follow-up chest CT revealed the status post-embolization of the coronary CFA. There is no obvious shunting flow from the right coronary artery, but there is still some shunting flow from the left coronary artery.
coronary pulmonary artery fistula

Complications (23%) increase noticeably. Therefore, the authors recommend treatment during the childhood period, even though the treatment protocols are under debate.

Treatment of asymptomatic patients with insignificant shunting is still a matter of dispute [14]. Generally, asymptomatic patients are under follow-up observation without special treatment and even spontaneous closure has been reported in some cases [17]. However, although it is rare at approximately 1% and the mechanism is not clear, it has been speculated to be due to atherosclerosis, embolism, and vasodilatation [17]. Medical treatment with either beta blockers or calcium channel blockers is suggested. Prophylaxis for bacterial endocarditis is recommended in all CAF patients and in patients after complete fistula occlusion for at least 1 year [18].

For cases with symptoms caused by fistula, developing complications, or with the large left-to-right shunt, surgical intervention or percutaneous closure have been recommended. Recently, minimally invasive percutaneous transcatheter embolic occlusion, or the transcatheter closure technique, which is minimally invasive in comparison with surgery, has been introduced as a new therapeutic option for CPAF treatment [19].

The first therapeutic embolization was performed in 1974 by Zuberbuhler et al. Since transcatheter closure of the CPAF is associated with a much shorter recovery time and does not leave a scar, it is considered the procedure of choice when fistula closure is indicated. Catheter closure can be performed with a variety of techniques, including detachable balloons, stainless steel coils, controlled-release coils, controlled-release patent ductus arteriosus coils, patent ductus arteriosus plugs, regular and covered stents, and various chemicals [18]. Surgical ligation should be reserved for patients who have a complex and distally located fistula, or who have adjacent vessels at risk. In addition, surgical ligation may be preferred when correction of other congenital defects or coronary artery bypass grafting is required. Mortality related to surgical closure or transcatheter closure of isolated CAF is low (< 1%). Incomplete closure has been seen in ~10% of patients treated by catheter techniques or surgical ligation.

Latson et al. proposed a way to classify the size of CAFs. Fistulas that, at any point, are larger than two times but less than three times the expected proximal normal coronary artery diameter, or that are associated with a similar range of dilation of the proximal associated coronary artery, are considered to be medium-size fistulas. Fistulas that are more than three times the proximal coronary artery diameter are considered large. This classification can be useful in making clinical decisions [20]. Based on these criteria, the right CPAF of this patient was a medium to large fistula and the left CPAF a small one. The clinical symptoms and signs of coronary steal phenomenon with angina pectoris are largely caused by the right CPAF rather than the left CPAF.

In our patient, the cover stent could not be used in the right CPFA and there was a high probability that it would have caused restenosis in the left CPFA. This patient was in the end treated with coil embolization in the right CPFA, and the left CPFA was treated with medical therapy.

In conclusion, the optimal strategy to manage symptomatic patients still remains controversial due to the lack of long-term clinical follow-up. The transcatheter coil embolization for CPAF is a less invasive procedure and has recently been considered as an alternative to surgical therapy. If the coil embolization had failed in my patient and angina pectoris had been recurrent, surgical closure would have been another therapy.

REFERENCES

   Am J Med 2006; 332: 79
Coronary pulmonary artery fistula