

Case Report

## Double Congenital Fistulae with Aneurysm Diagnosed by Combining Imaging Modalities

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Congenital coronary pulmonary artery fistula (CAF) is rare, and systemic-to-pulmonary artery fistula (SPAF) is even more so. Furthermore, congenital coronary pulmonary fistula associated with congenital SPAF is extremely rare. As far as we know, CAF and SPAF connected with an aneurysm have not been described very often. We described an 83-year-old woman with an aneurysm originating from a CAF connected to an aortopulmonary artery fistula. Chest radiography revealed a shadow at the left edge of the heart line. Multi-detector-row computed tomography (MDCT) with contrast enhancement and coronary cine angiography revealed that the shadow was an aneurysm connected to a tortuous fistula at the left anterior descending coronary artery. The aneurysm was formed by congenital coronary pulmonary and aortopulmonary artery fistulae. Echocardiography revealed predominantly systolic blood flow in the fistula from the left anterior descending coronary artery (LAD). Although neither MDCT, echocardiography nor coronary angiography alone could provide a comprehensive image of the anomaly, including the hemodynamics in the fistulae and their relationship with surrounding organs and tissues, their combination could provided important facts the led to a deeper understanding of this very uncommon occurrence.

**Key words:** coronary pulmonary artery fistula, aortopulmonary artery fistula, aneurysm

**C**oronary pulmonary artery fistula (CAF) is a rare congenital anomaly. Congenital systemic-to-pulmonary artery fistula (SPAF) is also very rare. Therefore, only a few reports have described congenital CAF associated with congenital SPAF. Since the fistulae often meander, it is sometimes difficult to image an entire view. We describe a patient with CAF and SPAF that was visualized and diagnosed using coronary angiography, Multi-detector-row computed tomography (MDCT) and transthoracic echocardiography (TTE).

### Case Report

A physician had prescribed telmisartan, cilnidipine and trichlormethiazide (40, 10 and 1mg/day, respectively) as follow up for hypertension in an 83-year-old-woman. He referred her to our hospital for further assessment of palpitations in July 2011.

She was aware and alert upon presentation; and soon thereafter, the palpitations disappeared without specific treatment and she became asymptomatic. A physical examination revealed normal heart and lungs, arterial blood pressure of 150/78mmHg and a heart rate of 91 beats/min.

Laboratory findings were essentially normal and 12-lead electrocardiographic findings were also nor-

mal. However, chest radiography revealed a shadow at the left edge of the heart line (Fig. 1). Transthoracic echocardiography (TTE) visualized a cystic mass touching the main pulmonary artery, and the left anterior descending coronary artery (LAD) seemed connected to the mass. Unlike blood flow in the normal coronary artery, that in the LAD of this patient was evident throughout the cardiac cycle, predominantly during ventricular systole. Left ventricular wall motion as well as right and left ventricular dimensions were normal. The diameter of the main pulmonary artery was 26 mm just above the pulmonary valves. The right ventricular systolic pressure calculated from the tricuspid regurgitation flow velocity was 26 mmHg, and the ratio of pulmonary to systemic flow volume was slightly increased ( $Q_p/Q_s = 1.2$ ). Color Doppler imaging showed shunt flow from the mass to the main pulmonary artery (Fig. 2).

Early enhancement of the mass was evident on contrast-enhanced MDCT images. We tentatively diagnosed the mass as an aneurysm, although MDCT did not provide sufficient information with which to evaluate stenotic lesions of native coronary arteries. The patient was admitted to our hospital for further examination.

We initially evaluated the relationship between the mass and the coronary artery by coronary angiography, which revealed 2 abnormal fistulae. One branched from the mid-region (#7) of the LAD, and the other originated from the right sinus of Valsalva at the ascending aorta. Each fistula emptied into a large ( $47 \times 33$  mm) aneurysm (Fig. 3), but angiography could not identify the point of outflow to the pulmonary artery due to the size of the aneurysm. No significant stenoses were evident in the native coronary arteries.  $^{201}\text{Tl}$  myocardial stress scintigraphy demonstrated the absence of myocardial ischemia due to coronary blood flow diversion by the fistula.

We performed electrocardiography-synchronized coronary CT to further clarify the run of the fistulae and their association with the aneurysm (Fig. 4A and B). The tortuous fistulae were entangled and emptied into the aneurysm. Echocardiography identified the point of outflow from the aneurysm in the main pulmonary artery as being just above the pulmonary valve (Fig. 5).

Based on these findings, we finally diagnosed CAF and SPAF with a single aneurysm. Although the

patient became asymptomatic while on hypotensive drugs, we were concerned about the possibility of rupture because the aneurysm was quite large. We

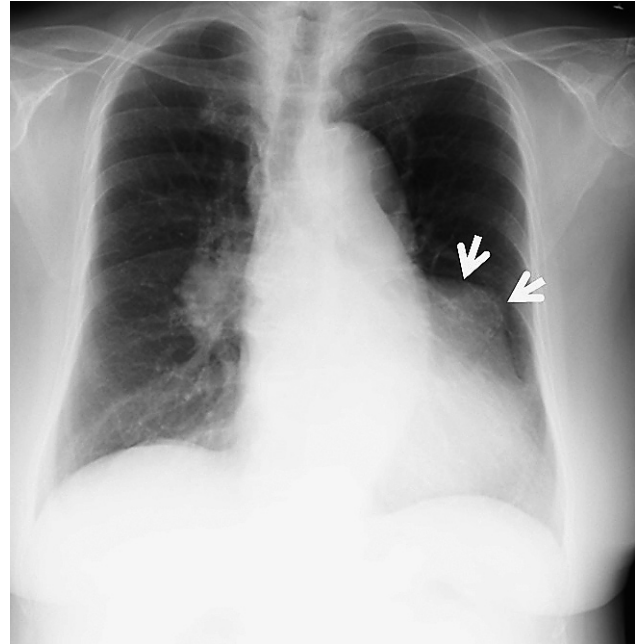


Fig. 1 Chest radiographic findings upon presentation. Shadow is in contact with left edge of heart line (arrow). Right pulmonary artery is slightly enlarged.

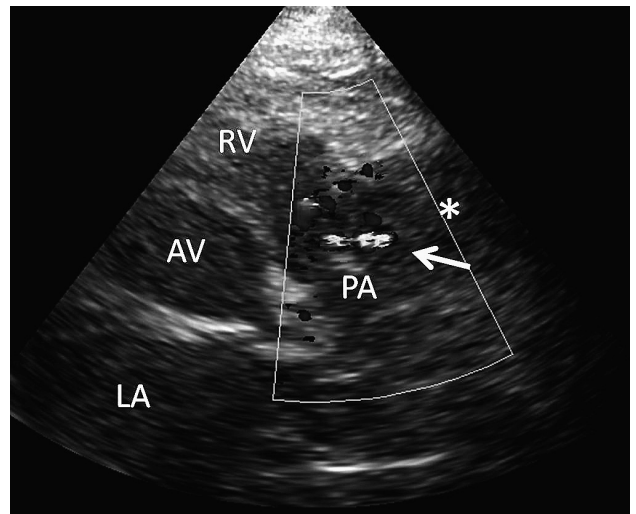
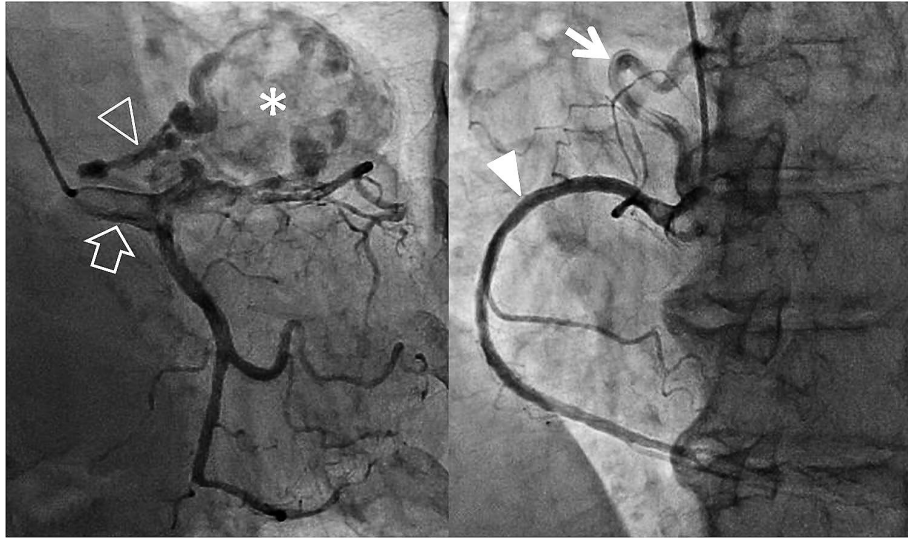
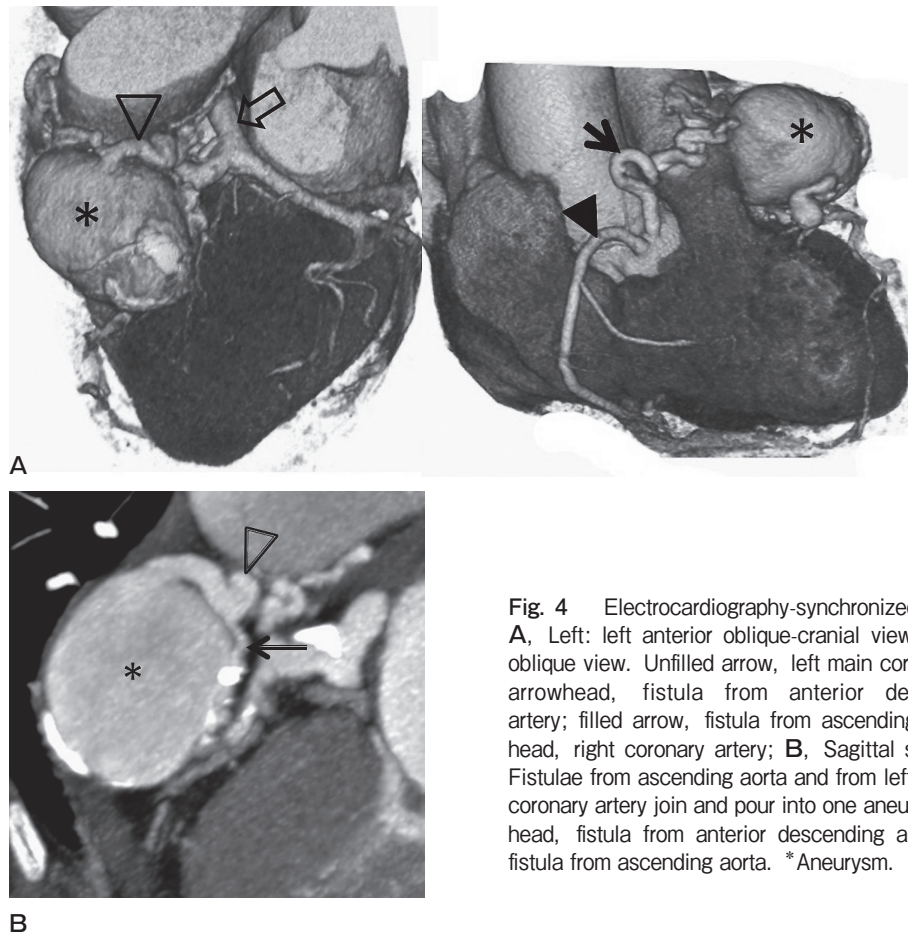


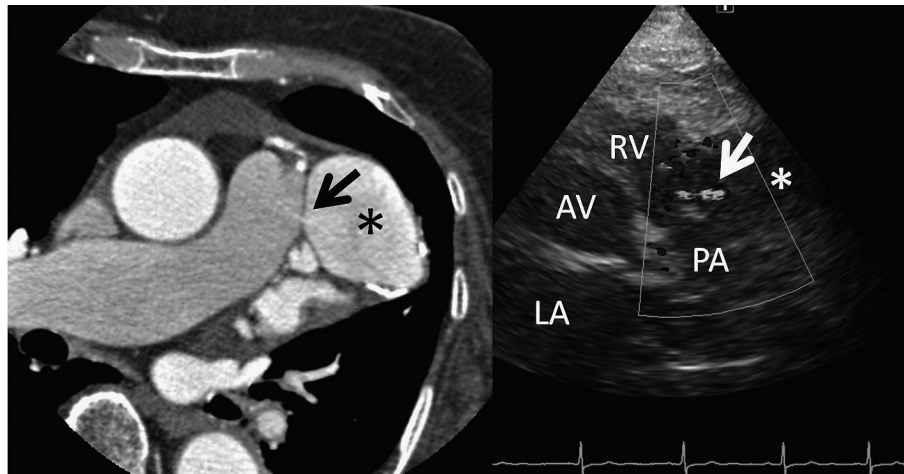
Fig. 2 Two-dimensional echocardiographic findings. Arrow, shunt flow from aneurysm to main pulmonary artery. AV, aortic valve; LA, left atrium; PA, pulmonary artery; RV, right ventricle. \*Aneurysm.



**Fig. 3** Conventional angiographic findings. One fistula branched off from the left anterior descending coronary artery and the other originated from the ascending aorta. Unfilled arrow, left main coronary artery; unfilled arrowhead, fistula from anterior descending coronary artery. Filled arrow, fistula from ascending aorta; filled arrowhead, right coronary artery. \*Aneurysm.



**Fig. 4** Electrocardiography-synchronized coronary CT (3D). **A**, Left: left anterior oblique-cranial view; right, left anterior oblique view. Unfilled arrow, left main coronary artery; unfilled arrowhead, fistula from anterior descending coronary artery; filled arrow, fistula from ascending aorta; filled arrowhead, right coronary artery; **B**, Sagittal section of aneurysm. Fistulae from ascending aorta and from left anterior descending coronary artery join and pour into one aneurysm. Unfilled arrowhead, fistula from anterior descending artery; straight arrow, fistula from ascending aorta. \*Aneurysm.



**Fig. 5** Contrast-enhanced multidetector CT with (left) and 2-D echocardiographic (right) images. Arrow (left) indicates location of out-flow from aneurysm. AV, aortic valve; LA, left atrium; PA, pulmonary artery; RV, right ventricle. \*Aneurysm.

advised the patient to undergo fistula ligation, but she declined this course of action. She remains under treatment for hypertension and undergoes regular CT assessment.

### Discussion

The etiology of CAF and SPAF is classified as acquired and congenital. Acquired fistulae could be caused by infection, trauma, neoplasms or thoracic surgery. Our patient had no history of diseases that could have led to such fistulae, indicating that they were probably congenital. Congenital CAF account for 0.2%–0.4% of all congenital cardiac anomalies [1], and since congenital SPAF is also extremely rare [2], very few reports have described congenital CAF connected with SPAF [3].

Conventional angiography has traditionally been useful for diagnosis. Although understanding the entire path of the tortuous fistulae in our patient was difficult, angiography confirmed the absence of severe stenotic changes in the coronary arteries and identified the origin of the fistulae in our patient. Coronary angiography can reliably show the proximal part of fistulae and lead to evaluations of size or number. Zenooz *et al.* [4] and Shmitt *et al.* [5] considered that conventional angiography might not be able to clearly visualize coronary fistulae draining to low-pressure chambers of the heart because the contrast medium becomes significantly diluted. Multi-detector-

row CT helped to clarify the anatomical complexity, the entire course of the fistulae and the nature of the aneurysm in our patient. However, evaluating a native coronary artery by MDCT at the time of a first medical examination is sometimes insufficient. Said [6] considered that because such anomalies are mostly discovered in elderly patients, arterial calcifications can obscure native coronary stenotic changes. Nakamura *et al.* [7] found that MDCT was a useful alternative to echocardiography and coronary angiography during follow-up. Fistulae usually have small diameters and are prone to extensive meandering, which causes vessels to easily become resistant. This explains the small amount of shunt flow and why patients remain asymptomatic until they become quite elderly. Although MDCT can easily identify a drainage vessel that lies between an aneurysm and the pulmonary artery, the aneurysm in our patient opened directly to the main pulmonary artery and the small shunt hole resulted in visualization of only the fine shunt flow line on MDCT. Only TTE could confirm the location of the drainage site of the aneurysm in our patient and confirm that it was not an artifact.

The primary therapeutic approach to CAF and SPAF is surgery or interventional catheterization. Surgery is recommended for symptomatic patients, such as those with congestive heart failure from volume overload, myocardial ischemia or bacterial endocarditis. Our patient underwent myocardial scintigraphy stress testing to evaluate asymptomatic

myocardial ischemia due to fistular coronary diversion. We did not uncover any evidence of myocardial ischemia in the LAD area. The therapeutic management of asymptomatic patients is controversial. Nakatani *et al.* [8] described spontaneous closure of a fistula secondary to spontaneous thrombosis. Therefore, immediate closure is not considered necessary in asymptomatic patients. Said *et al.* [9] recommend careful periodic evaluation and several years of follow-up. Medical therapy is seldom indicated except for congestive heart failure. Since myocardial infarction occurs in 2% of patients with or without atherosclerotic lesions, antiplatelet therapy has been recommended. On the other hand, Ueyama *et al.* [10] indicated that aneurysms with a diameter of >30 mm are likely to rupture regardless of whether or not they are symptomatic. Because our patient refused to undergo surgery, we applied a conservative strategy comprising periodic monitoring, good oral hygiene to prevent endocarditis and antihypertensive therapy to prevent aneurysmal rupture without antiplatelet therapy.

In conclusion, we combined imaging modalities to comprehend the anatomical complexity of an exceedingly rare aneurysm originating from a coronary-to-pulmonary artery fistula connected to an aortopulmonary artery fistula.

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