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Rare case of congenital coronary artery fistula coexistent and coalesced with aortopulmonary fistula

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SUMMARY

Coronary artery fistula (CAF) is an uncommon congenital heart disease. Furthermore, aortopulmonary fistula is a rare congenital heart disease of adult onset. We report the case of a 79-year-old man who presented with chest pain. ECG-gated cardiac CT and coronary artery angiography revealed an anomalous vessel arising from the right coronary cusp and a CAF from the left coronary descending artery. These fistulas coalesced and drained into the same portion of the pulmonary artery. Haemodynamic studies revealed that the estimated systemic-to-pulmonary flow ratio was 1.18. The mean pulmonary pressure was 14 mm Hg. We decided against surgical intervention due to his advanced age and lack of heart failure symptoms. The patient did not have any worsening heart failure and chest pain on follow-up. This was a rare case of CAF coexistent and coalesced with an aortopulmonary fistula.

BACKGROUND

Coronary artery fistula (CAF) is a very rare disease.¹ It is a communication between the coronary arteries and the cardiac chambers or other vessels. Aortopulmonary fistula is also a rare disease. We report a case where CAF and an aortopulmonary fistula coalesced and drained into the pulmonary artery (PA). We obtained a detailed anatomical information using multidetector CT angiography and coronary artery angiography.

CASE PRESENTATION

A 79-year-old man with a 3-month history of chest discomfort unrelated to exercise was admitted to our hospital. The patient reported a medical history of asthma and chronic obstructive pulmonary disease, which were well-controlled by medication. On initial examination, his blood pressure was 115/82 mm Hg, heart rate was 78 beats/min, respiratory rate was 12 breaths/min, pulse oximetric oxygen saturation was 98% in room air and his temperature was 36.2°C. The patient's height and weight were 170 cm and 66 kg, respectively. Clinical examination of the cardiovascular system revealed no murmurs, and the lungs were clear. His jugular venous pressure was not elevated, and he did not exhibit heart failure symptoms, such as shortness of breath and leg oedema.

INVESTIGATIONS

A 12-lead ECG revealed sinus rhythm and T-wave inversion in leads V1, V2 and V3. Transthoracic ECG illustrated one flow just above the pulmonary valve, preserved left ventricular ejection fraction,

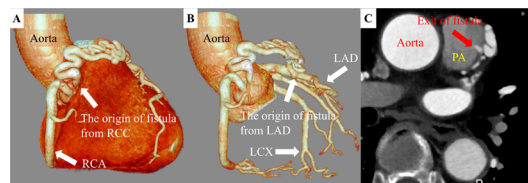


Figure 1 The ECG-gated cardiac CT revealing abnormal vessels arising from the LAD and RCC. (A) Right-lateral view illustrates the fistula from RCC. (B) Right-lateral view illustrates the fistula from the LAD. (C) Superior view illustrating the exit of fistulas. LAD, left coronary descending artery; LCX, left coronary circumflex artery; PA, pulmonary artery; RCA, right coronary artery; RCC, right coronary cusp.

no enlargement of the left and right heart, trivial mitral regurgitation and trivial tricuspid regurgitation. ECG-gated cardiac CT revealed an anomalous vessel arising from the right coronary cusp (RCC) and a CAF from the left coronary descending artery (LAD; [figure 1A,B](#)). The abnormal blood vessels emerging from the RCC and the CAF emerging from the LAD coalesced, and drained into the PA ([figure 1C](#)). Coronary artery angiography revealed no significant stenosis of the coronary artery; however, it revealed fistulas originating in both the RCC and LAD ([figure 2A,B](#); [videos 1 and 2](#)). These fistulas drained into the same portion of the main PA. Haemodynamic studies revealed that the estimated systemic-to-pulmonary flow ratio was 1.18 ([table 1](#)). Step-up of oxygen saturation was observed between the main PA and right ventricle outflow ([table 2](#)). The mean PA pressure was within normal range. Single-photon emission CT revealed no myocardial ischaemia.

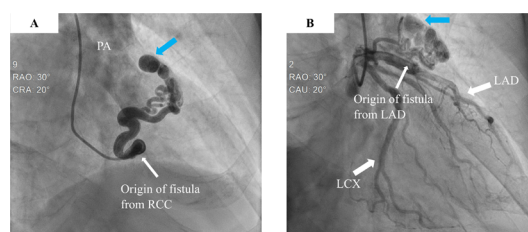
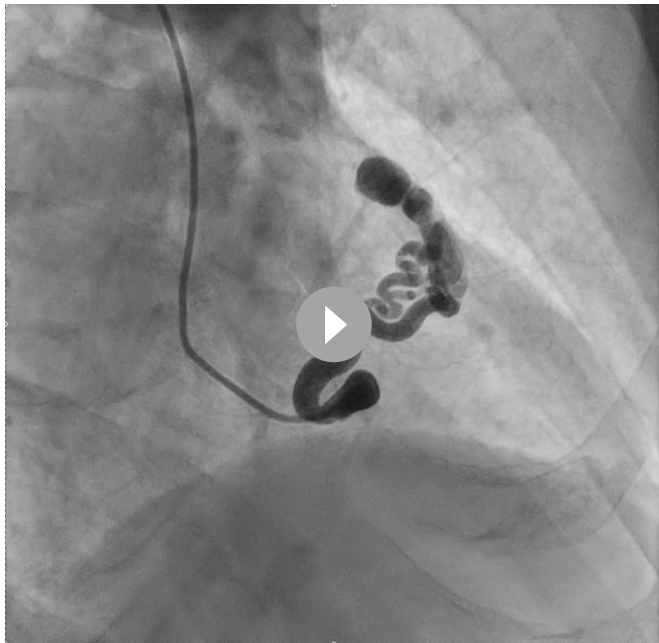


Figure 2 Angiography of coronary artery and fistulas. (A) Right-cranial view illustrating the fistula originating from the right coronary cusp. (B) Right-caudal view illustrating the fistula from the left coronary descending artery. The two fistulas draining into the same portion of the main PA. CAU, caudal; CRA, cranial; LAD, left coronary descending artery; LCX, left coronary circumflex artery; PA, pulmonary artery; RAO, right anterior oblique; RCC, right coronary cusp.



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Video 1 Angiography of coronary artery and fistulas. Right cranial view illustrating the fistula originating from the right coronary cusp.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of this case included angina pectoris, myocarditis, pericarditis, musculoskeletal disease and oesophageal disease.

TREATMENT

The estimated systemic-to-pulmonary flow ratio was low, and single-photon emission CT revealed no ischaemic myocardium. We decided against surgical intervention due to his advanced age and lack of heart failure symptoms.



Video 2 Angiography of coronary artery and fistulas. Right caudal view illustrating the fistula from the left coronary descending artery.

Table 1 Haemodynamic studies of the case

Aorta s/d/m	162/106/130	mm Hg
Pulmonary capillary wedge a-wave/v-wave/mean	6/1/3	mm Hg
PA s/d/m	21/10/14	mm Hg
Right ventricle systolic/diastolic/EDP	26/6/10	mm Hg
Right atrium a a-wave/v-wave/mean	9/8/7	mm Hg
Heart rate	71	beats/min
Cardiac output (Fick)	7.15	L/min
Cardiac index (Fick)	3.97	L/min/m ²
Qp/Qs*	1.18	

The estimated systemic-to-pulmonary flow ratio is 1.18. The pulmonary artery pressure is within the normal range.

*Pulmonary blood flow/systemic blood flow ratio.

EDP, end-diastolic pressure; PA, pulmonary artery; s/d/m, systolic/diastolic/mean.

OUTCOME AND FOLLOW-UP

After discharge, the patient was carefully followed up.

DISCUSSION

We describe a rare case of a CAF. To our knowledge, this is the first documented case of congenital aortopulmonary fistula combined with CAF. These two fistulas were connected to each other.

CAF is a rare congenital anatomical abnormality of the coronary arteries. CAF prevalence is reportedly 0.9% of all congenital heart diseases.¹ CAF may arise from the LAD, left coronary circumflex artery or the right coronary artery. Congenital coronary-PA fistula is a CAF and its characteristics are poorly understood. The majority of CAFs arise from the left coronary artery.²⁻⁴ The most common drainage site for CAF is PA (85%).⁴ In this case, the fistula connected the left descending coronary artery to the PA.

Aortocardiac fistula is a rare condition. Aortopulmonary fistula is an aortocardiac fistula. There were no reports of the prevalence of the aortopulmonary fistula. The aortocardiac fistula is common in the aortic-right atrium (37%) and aortic-PA (25%).⁵ The origin of the aortocardiac fistula in this case was RCC, namely the ascending aorta.

With the development of CT, it has become possible to determine the origins, drainage sites and the number of fistulas in detail.⁶ As in this case, the results of CT can be used to determine examination methods and treatment strategies. Angiography of coronary artery and fistulas is an invasive study and has some

Table 2 Oxygen saturation

	O ₂ saturation (%)
Right PA	76.2
Left PA	75.3
Main PA	75.9
Right ventricle outflow	69.6
Right ventricle apex	68.1
Right ventricle inflow	69.4
Superior vena cava	72
Right atrium	70.5
Inferior vena cava	76.2
Aorta	91.2

Illustration of the step-up of oxygen saturation between the main pulmonary artery and right ventricle outflow.

PA, pulmonary artery.

risks of complications. Also, the accurate diagnosis rate is low with invasive angiography.

The most commonly reported symptoms of CAF are chest pain (39%) and respiratory dyspnoea (25%), with murmur being the most common finding on physical examination (37%).² In this case, however, chest pain was the only symptom. The coronary 'steal phenomenon' is considered to be the primary cause of CAF without coronary artery stenosis. This mechanism is related to the flow from the high-pressure coronary artery to a low-resistance PA, owing to the pressure gradient. This symptom is associated with an increased myocardial oxygen demand during exercise or activity.

Researchers agree that the symptomatic patients should be treated. According to the American College of Cardiology/American Heart Association guidelines, 'percutaneous or surgical closure is a Class I recommendation for large fistulas regardless of symptoms, and for small or moderate fistulas with evidence of myocardial ischemia, arrhythmia, ventricular dysfunction, ventricular enlargement, or endarteritis'.⁷ In this case, however, we decided to treat the patient conservatively for multiple reasons. First, his symptoms were atypical of angina pectoris and did not worsen on exertion. The frequency of his symptoms was approximately once a month. Second, he exhibited no symptoms

of heart failure or pulmonary hypertension. Third, the mean PA pressure and estimated systemic-to-pulmonary flow ratio were low. Fourth, there were some risks associated with surgery due to his advanced age.

In conclusion, this was a rare case where CAF and an aortopulmonary fistula coalesced and drained into the PA.

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Learning points

- ▶ To create awareness among physicians regarding the possibility of coexistence of exceedingly rare anatomical pathologies, coronary artery fistula and aortopulmonary fistula, which also coalesced before draining into the pulmonary artery. This case study will also help physicians understand its presentation.
- ▶ To understand that multidetector CT angiography is helpful in investigating for congenital coronary artery fistulas and can be used to plan the approach towards coronary artery angiography.
- ▶ To assess the risk of the diseases, examinations and anatomical evaluation about adult congenital heart diseases should be done.

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