



# Coronary artery fistula and lung adenocarcinoma: a case report and literature review

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**Abstract:** Coronary artery fistula (CAF) is a rare condition, whilst lung cancer is one of the most common malignant tumors worldwide. We came across an interesting case with both diseases. To the best of our knowledge, this is the first case report pertaining to a patient with a coexisting CAF and lung adenocarcinoma. The patient was a 67-year-old woman who was admitted to our hospital for evaluation of persistent cough. Through the examination she was diagnosed coronary artery fistula and lung adenocarcinoma. Both diseases were successfully treated in a single operation (artery ligation and pulmonary lobectomy). The post-operative period was uneventful. At 3-month follow-up, there were no signs of blood shunting or cancer recurrence. There is no standard guidelines to treat both diseases. We want to seek out a solution to the problem. In this patient, we successfully performed artery ligation and pulmonary lobectomy in a single operation without any complications. We believe the treatment of patients with CAFs should be individualized. But, there is still a lot of shortcomings in our research. First of all, we have not enough cases to support our approach. What's more, the long-term effects of the operation are not certain. Last but not least, we have no proof in genetics with both diseases.

**Keywords:** Coronary artery fistula (CAF); lung adenocarcinoma; combined surgery; case report

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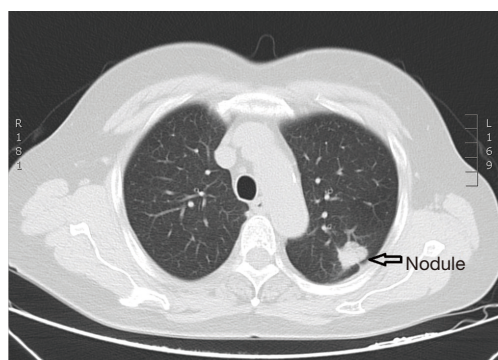
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## Introduction

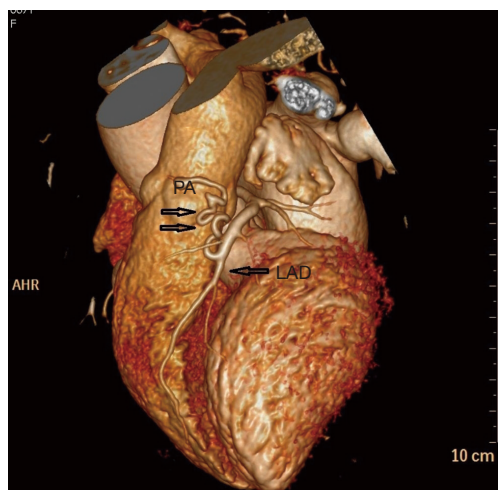
Coronary artery fistula (CAF) is a rare condition that refers to an anomalous connection between one or more coronary arteries and a cardiac chamber or great vessel (1). The majority of CAFs arise from the right coronary artery (RCA), while those arising from the left anterior descending artery (LAD) and the left circumflex artery (LCx) are less common (2). Surgery is a traditional form of treatment. Transcatheter approaches may be more beneficial than surgical approaches for eligible coronary artery fistula cases. CAF can be categorized as either congenital or acquired anomalies. Congenital coronary artery fistulas are more common but only account for 0.4% of congenital cardiac abnormalities. Acquired coronary artery fistulas develop

from injurious events, iatrogenic or traumatic in nature. Over the past two decades, lung adenocarcinoma has been consistently ranked as the most common type of lung cancer. So far, surgery is still the main treatment for lung adenocarcinoma. Other traditional treatments, such as chemotherapy and radiotherapy, are still effective. In recent years, new therapeutic methods such as targeted therapy and immunotherapy have emerged, and they have gained more and more effects and recognition. Cases with concomitant CAF and lung cancer have not been reported to date. Herein we report a patient with coexisting LAD-main pulmonary artery (PA) fistula and lung adenocarcinoma. Both conditions were successfully treated in a single surgical procedure.

We present the following article in accordance with



**Figure 1** Computed tomography (CT) radiograph showing a tumor measuring 2 cm × 1.8 cm in the upper lobe of left lung.

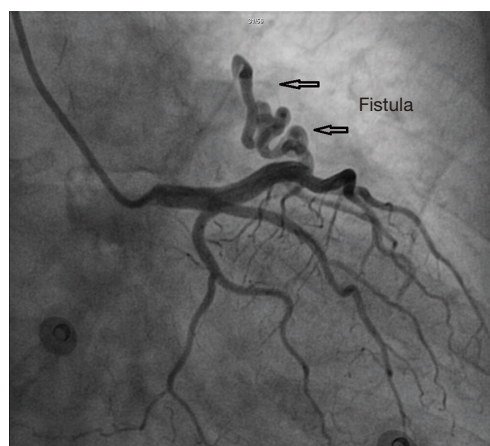


**Figure 2** Three-dimensional, volume-rendered multidetector-row computed tomography (MDCT) images showing abnormal vessels located anterior to the main pulmonary artery. PA, pulmonary artery; LAD, left anterior descending artery.

the CARE reporting checklist (available at <http://dx.doi.org/10.21037/apm-20-2015>).

### Case presentation

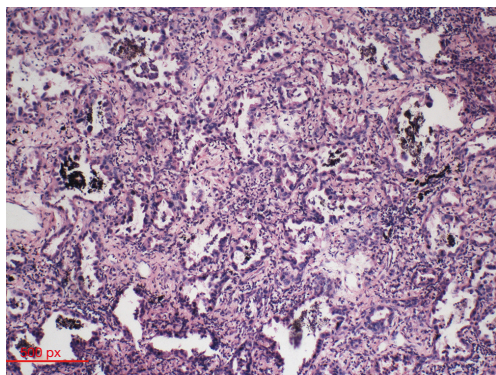
A 67-year-old woman was admitted to our hospital for evaluation of persistent cough. The patient was yellow and had no family history. Findings from the general physical examination were unremarkable. Laboratory investigations, including liver function tests and serum levels of carcinoembryonic antigen, carbohydrate antigen CA-199, and CA-125 were normal. Chest contrast-enhanced multi-detector computed tomography (MDCT) showed a nodule



**Figure 3** Coronary angiogram showing the fistula and the shunt.

(largest diameter: 2 cm) in the upper lobe of the left lung (Figure 1). The tumor had a lobulated shape with spiculated margins and had caused pleural traction. The mass was suspected to be a lung carcinoma. During the pre-operative evaluation, a fistula with active flow between the LAD and the PA was detected by trans-thoracic echocardiography. The fistula was confirmed by coronary artery multi-slice spiral CT (Figure 2) and coronary angiography (Figure 3). Therefore, a diagnosis of lung neoplasm and CAF was confirmed. Lung adenocarcinoma in early stage should be treated surgically. Surgical ligation or percutaneous transcatheter occlusion are possible treatment options to coronary artery fistula. In this case, we took into account the patient's economic factors and to avoid the damage caused by the second operation. After obtaining the consent of the patient, we adopted a combined operation. We planned for surgical treatment of both conditions in a single procedure via sternotomy.

Intraoperatively, circuitous vessels originating from the mid-portion of the LAD to the PA were observed. A thrill was auscultated over the fistula vessel. We first ligated the abnormal blood vessels on the aortic and pulmonary sides. Subsequently, we opened the mediastinal pleura to visualize the left upper lobe of the lungs. The tumor was located in the periphery. A wedge resection was performed to obtain a biopsy specimen. The tumor was whitish gray in appearance. Examination of frozen sections revealed a lung adenocarcinoma. Based on the pathology report, pulmonary lobectomy and systematic mediastinal lymphadenectomy were performed. Post-operative pathologic evaluation indicated a moderately differentiated adenocarcinoma (50% alveolar type and 40% papillary type; Figure 4). The pathologic TNM stage was T1bN0M0. The post-operative



**Figure 4** Microscopic examination of the surgical specimen showing moderately differentiated adenocarcinoma with 50% alveolar type and 40% papillary type (hematoxylin and eosin staining; magnification 40×).

period was uneventful and the patient was discharged on the 15th post-operative day. No blood shunting between the coronary and pulmonary arteries or lung cancer recurrence was observed at 3-month follow-up.

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient.

## Discussion and conclusions

CAFs were first reported by the German anatomist Krause in 1865 (3). An estimated 0.002% of the general population are affected by CAFs. Most CAFs are congenital anomalies and are typically diagnosed incidentally during coronary angiography (incidence: 0.18%) (2). CAFs originate from the right and left coronary arteries in approximately 52–60% and 30% of cases, respectively. CAFs originating from the LCx account for 18% of cases (4–6). The flow endpoint of the fistula is in the right ventricle in 40% of cases, right atrium in 25% of cases, PA in 15–20% cases, coronary sinus in 7–10% of cases, and the superior vena cava in 1–2% cases (7).

Most CAFs are congenital, however, CAFs may also occur as a complication of chest trauma, infection, coronary angioplasty, or cardiac operations (8,9). Clinical symptoms arising from CAFs are unusual. Most patients are asymptomatic for long periods of time (10). Some congenital CAFs can be identified in childhood based on murmurs and/or clinical symptoms (11). Symptoms are more likely to manifest in the elderly, and are largely

attributable to the blood flow through the shunt and the structures involved (12–15). CAFs may sometimes cause acute myocardial infarction, which may necessitate coronary artery bypass surgery (7,16). Our patient had no apparent symptoms because there was a minimal shunt from the distal coronary blood flow.

Coronary angiography and MDCT are reliable techniques used to identify and delineate the course of anomalous coronary arteries (17). Other examinations, such as magnetic resonance imaging (MRI), transthoracic echocardiography, and Doppler echocardiography are also helpful (18–21). In the present case, CAF was detected during trans-thoracic echocardiography as part of the pre-operative evaluation for lung neoplasm, and was subsequently confirmed by coronary angiography.

There are no standard guidelines for the treatment of CAFs (22). The indications for fistula closure are mainly based on clinical symptoms, such as heart failure, myocardial ischemia, aneurysmal dilatation, or high-flow shunting (23,24). In severe cases, spontaneous rupture of coronary artery-to-pulmonary artery fistulas may cause hemopericardium and cardiac tamponade (25). Current treatment options include observation, transcatheter embolization, and surgical ligation. Transcatheter approaches are preferred over the open surgical approach owing to the lower cost, lower morbidity, shorter recovery time, and better cosmetic results. Transcatheter techniques also have some shortcomings, such as distal embolization, which may cause ischemic electrocardiographic changes or myocardial infarction (26). Surgical closure is preferred for CAFs with the following characteristics: (I) locations inaccessible for percutaneous intervention; (II) multiple fistulae; (III) extreme native vessel tortuosity; (IV) aneurysmal dilation of the fistula or proximal coronary artery; and (V) presence of normal coronary branches near the fistula drainage sites (27). Biorck and Crafoord (28) were the first to report open surgery in a patient with a coronary artery-to-pulmonary artery fistula in 1947. Our case was unusual because the patient had two different medical problems (coronary artery-to-pulmonary artery fistula and lung adenocarcinoma). The clinical pre-operative TNM staging was T1bN0M0, for which surgical resection is the standard treatment. We decided to treat both conditions in a single operation. No complications happened after the surgery. The pathologic TNM stage was T1bN0M0, so we didn't do chemotherapy. The patient recovered well and was discharged 15 days after surgery, thus avoiding a second surgical procedure.



In summary, we have reported the first case of concomitant CAF and lung cancer. Our successful experience suggests that the treatment of patients with CAFs should be individualized.

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## Footnote

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**Conflicts of Interest:** All authors have completed the ICMJE uniform disclosure form (available at <http://dx.doi.org/10.21037/apm-20-2015>). The authors have no conflicts of interest to declare.

**Ethical Statement:** The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient.

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