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Case Report

Thoracoscopic Sublobectomy for Lung Cancer in a Patient with Unilateral Absence of Pulmonary Artery: Case Report and Narrative Review

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Unilateral absence of a pulmonary artery (UAPA) is an uncommon congenital anomaly. Among the rarer conditions, UAPA with lung cancer has been previously reported in 13 cases; however, there remains controversy regarding the surgical approach and precautions. Herein, we present a case study of a 56-year-old female patient who was incidentally diagnosed with a nodule in the right lower lobe of the lung during a routine physical examination and subsequently found to have an absent right pulmonary artery upon preoperative evaluation. A wedge resection of the right lower lobe was performed as treatment. Postoperative pathology confirmed invasive adenocarcinoma (pT1N0M0). We provide a narrative review of existing literature on these patients and discuss optimal surgical management strategies.

Keywords: lung cancer, sublobectomy, unilateral absence of pulmonary artery

Introduction

Unilateral absence of a pulmonary artery (UAPA) is a rare congenital anomaly, first reported by Frentzel in 1868,[1] many of which are associated with other cardiac malformations, such as ventricular septal defect and tetralogy of Fallot. The incidence of isolated UAPA without associated cardiac abnormalities is approximately 1 in 200000 to 1 in 300000.[2,3] Its most common symptoms are recurrent lung infections and exertional dyspnea. Hemoptysis and pulmonary hypertension are associated complications; however, most cases are asymptomatic, and they are not recognized until adulthood.

There is a much rarer condition, which is UAPA associated with lung cancer, according to our literature review, only 13 such cases have been reported before. The special anatomical characteristics of UAPA make the operation different from the routine, and the small number of cases leads to a lack of experience in surgical treatment. Therefore, we report a case of UAPA associated with ipsilateral lung cancer and discuss the matters requiring attention and the risks of surgical treatment. The works of literature on these 13 rare cases were reviewed, with emphasis on the surgical treatment process and patient outcomes.

Case Report

The patient was a 56-year-old woman who was found to have a lung nodule during routine physical examination without related symptoms and had no lifelong history of smoking. The patient wished to undergo surgery,

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and the preoperative pulmonary function assessment showed moderate diffuse dysfunction. The patient underwent pulmonary function exercise and aerosol inhalation therapy for 2 weeks, and the reexamination of pulmonary function showed mild obstructive pulmonary dysfunction. Forced expiratory volume in 1 s of 1.97 (80.6% predicted), forced vital capacity of 2.69 (93.6% predicted), and forced expiratory volume in 1 s to forced vital capacity ratio of 0.73. After admission, further examinations were performed to exclude distant metastasis of the tumor. Cardiac ultrasound showed mitral valve prolapse, pulmonary artery systolic pressure of 25 mmHg, no other cardiac structural deformity was found, chest enhanced CT showed mixed ground-glass nodules in the dorsal segment of the right lower lobe, with a diameter of 20 × 24 mm, and multiple bullae in the right lung (**Fig. 1A**). Pulmonary artery CT showed the absence of the right pulmonary artery and veins, and abnormally large left pulmonary artery (**Figs. 1B** and **2**). After excluding contraindications, the patient was treated surgically.

The procedure was performed via video-assisted thoracoscopy. Exploration revealed severe pleural adhesions in the patient, with densely covered collateral vessels adhering from the chest wall or diaphragm to the lung surface, which is the source of intercostal arteries, internal thoracic arteries, and diaphragmatic vessels; these vessels have thin walls and are prone to bleeding (**Fig. 3**). Notably, collateral vessels were particularly abundant in the diaphragm and lower lung ligament region which were difficult to dissociate. Following surgical exploration, it was determined that lobectomy might result in excessive intraoperative bleeding. To minimize trauma, a wedge resection was performed to ensure adequate tumor margin clearance. Postoperative pathology confirmed moderately differentiated invasive adenocarcinoma (adherent type 78%, acinar type 20%, micropapillary type 2%), with no lymph node metastasis detected during dissection.

The patient's postoperative recovery was not smooth. Despite adequate atomization treatment, analgesic treatment, and expectoration care, the patient had postoperative dyspnea several times with the oxygen saturation decreased to 70%–80%, especially at night. Arterial blood gas analysis showed that the oxygen partial pressure decreased to 38–47 mmHg, and the carbon dioxide partial pressure increased to 57–60 mmHg. The patient gradually improved after being given oxygen inhalation with a high-flow mask and the application

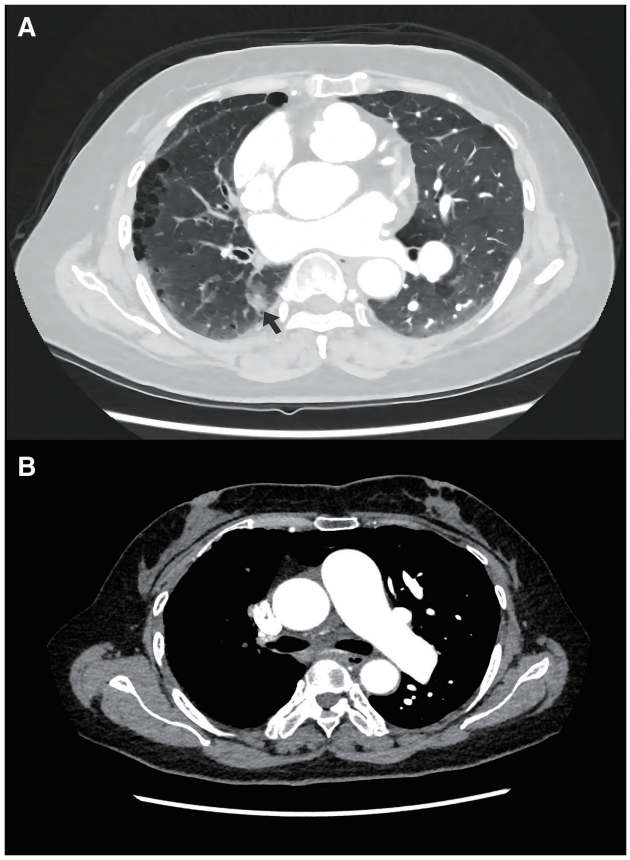

Fig. 1 Chest CT of the patient. (**A**) A mixed ground-glass nodule was seen at the black arrow in the lower lobe of the right lung and (**B**) CT angiography of the pulmonary artery showed that the right pulmonary artery was absent and the left pulmonary artery was enlarged.

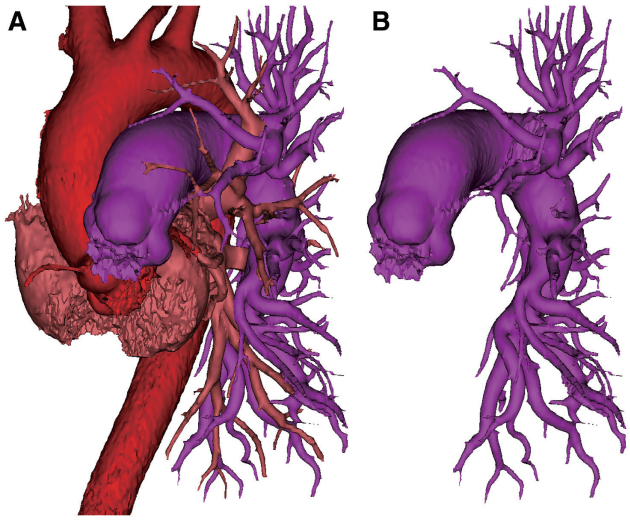

Fig. 2 Three-dimensional reconstruction of the patient's blood vessels. (**A**) Red blood vessels as the main artery, purple for the pulmonary artery and brown for the pulmonary vein. (**B**) Reconstruction of the pulmonary artery alone showed the absence of a right pulmonary artery.

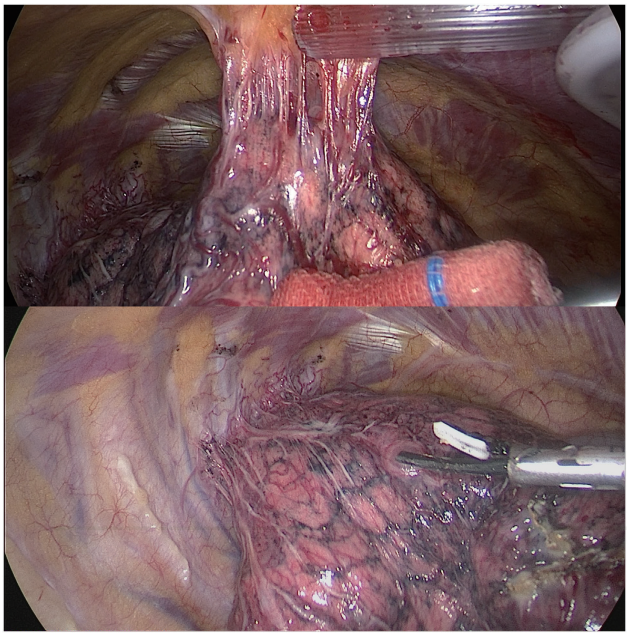

Fig. 3 Thoracoscope exploration found several collateral vessels from the chest wall

of theophylline. The chest drainage tube was removed on the ninth day after surgery, and the patient was discharged on the tenth day after surgery. After discharge, the patient had no symptoms of chest tightness and breathlessness, and his daily activities were not affected.

Literature Review

After a literature search, we identified a total of 13 UAPA patients associated with lung cancer that have been reported from 1975 to 2024. This case is the 14th patient. **Table 1** shows a summary of the information on these 14 patients.[4–16)]

The 14 patients included 5 males and 9 females, with a median age of 56.5 years at diagnosis, 7 patients had the absence of a left pulmonary artery, and 7 patients had the absence of a right pulmonary artery. Among them, the tumor and absence of arteries were located on the ipsilateral side in 10 patients and on the contralateral side in 4 patients. 10 of them were asymptomatic, while 4 patients had various symptoms such as fever, cough, hemoptysis, chest pain, and dyspnea.

Surgeries or biopsies were performed in all patients, including lobectomy in six patients, pneumonectomy in four patients, wedge resection in two patients, open biopsy in one patient, and mediastinoscope biopsy in one patient. The pathology results showed that nine patients had adenocarcinoma, two patients had squamous cell carcinoma, one patient had undifferentiated carcinoma, one non-small-cell lung cancer (NSCLC) patient (specific pathological type unknown), and one patient's pathologic type was not mentioned.

Of the 14 patients after treatment, 9 patients survived, 2 patients died after an operation or biopsy, and 3 patients were not mentioned.

Discussion

UAPA can be diagnosed by many methods, such as chest-enhanced CT, pulmonary artery computed tomography angiography (CTA), pulmonary angiography, and lung ventilation perfusion scan. On imaging examination, one side of the pulmonary artery is absent, accompanied by reduced lung volume on the affected side and hypoplastic lung tissue. Furthermore, multiple pulmonary bullae may coexist.

It remains unclear whether there is a correlation between UAPA and lung cancer. The role of chronic hypoxia as an impetus for DNA damage and the development of lung malignancy has been highlighted,[17)] suggesting that pulmonary insufficiency could potentially increase the risk of tumor growth. However, it should also be noted that reduced blood supply may decrease the likelihood of further tumor development and hematogenous metastasis. Currently, due to the low incidence of UAPA, data on concurrent lung cancer cannot be obtained. Among the 14 reported patients, one had distant metastasis, and another had mediastinal lymph node metastasis; most patients were in a resectable state. Most patients underwent anatomical lobectomy, which is indeed the standard procedure, but as sublobar resection is noninferior to lobectomy for some early peripheral lung cancers, sublobar resection may also be a treatment option under the circumstances.[18,19)]

UAPA can result in a range of anatomical variations. Insufficient perfusion within the affected pulmonary circulation leads to extensive collateral circulation compensation from the systemic circulation, primarily through thoracic aortic collaterals, intercostal arteries, internal thoracic arteries, and diaphragmatic vessels. Due to impaired ventilation and blood flow in the absent side lung tissue, there is generally poor lung development observed, characterized by reduced lung volume, widespread emphysema, multiple pulmonary bullae, and inadequate development of pulmonary fissures. This might be because the development of independent units

Table 1 Summary of cases of unilateral absence of pulmonary artery accompanied by lung cancer

Study	Year	Age/sex	Contra or ipsilateral	Absent pulmonary artery	Tumor location	Management	Outcomes
Mancebo[4]	1975	49/F	Ipsi	Right	Right upper lobe	Mediastinoscopy	Unknown
Roman[5]	1995	54/M	Ipsi	Left	Left lower lobe	Pneumonectomy	Unknown
Ito[6]	2010	57/M	Contra	Left	Right middle lobes	Lobectomy	Survived
Anstadt[7]	2011	67/F	Ipsi	Left	Left upper lobe	Lobectomy	Survived
Makdisi[8]	2015	50/F	Ipsi	Right	Right three lobes	Pneumonectomy	Survived
Watanabe[9]	2015	76/F	Ipsi	Right	Right lower lobe	Lobectomy	Unknown
Zhang[10]	2016	60/F	Contra	Right	Left lower lobe	Lobectomy	Expired
Kim[11]	2018	56/M	Contra	Left	Right lower lobe	Lobectomy	Survived
Agzarian[12]	2019	55/F	Ipsi	Left	Left lower lobe	Open biopsy	Expired
Matsumoto[13]	2020	80/M	Ipsi	Right	Right lower lobe	Lobectomy	Survived
Wang[14]	2020	56/M	Ipsi	Left	Left lower lobe	Pneumonectomy	Survived
Liu[15]	2021	60/F	Contra	Right	Left lower lobe	Sublobectomy	Survived
Kononets[16]	2022	59/F	Ipsi	Left	Left lower lobe	Pneumonectomy	Survived
Ai	2024	56/F	Ipsi	Right	Right lower lobe	Sublobectomy	Survived

of pulmonary artery-pulmonary vein-trachea is crucial for the formation of healthy lung segments, even lung lobes, and interlobular fissures.

Due to the aforementioned anatomical variation, surgical procedures present unique challenges. The location of the tumor and UAPA, whether ipsilateral or contralateral, has distinct implications for the operation. When the tumor is situated on the same side as the absent artery, surgical difficulties arise from collateral vessel bleeding, reduced lung compliance due to extensive pulmonary bullae, and pleural space adhesion resulting from chronic inflammation over an extended period. In our case, the patient exhibited numerous collateral vessels and was prone to bleeding; in addition, severe adhesion in the pleural cavity was observed necessitating a wedge resection. The postoperative response confirmed that this choice was appropriate. The patient had extremely poor pulmonary function reserve; although partial resection of the right lung would theoretically have no impact on overall pulmonary function, surgical shock still caused postoperative oxygenation to decline many times in this instance. This also suggests that strict control over the scope of surgical resection is imperative for patients with UAPA. As this patient's tumor contains ground-glass opacity, and combined with the postoperative pathological diagnosis of adenocarcinoma (adherent type 78%), we hypothesized that such patients would have a good tumor prognosis after sublobectomy with enough margin.

When the tumor is located contralaterally to the absence of a lung artery, surgical risk increases significantly due to the non-functional gas exchange capacity of the patient's other side lung. Therefore, a two-lung ventilation or intermittent two-lung ventilation approach should be adopted during the operation. It is crucial to carefully evaluate the patient's pulmonary function, as extensive resection may result in inadequate pulmonary function. To date, only four cases of contralateral surgery have been reported, all requiring completion under two-lung ventilation. The increased perfusion in the remaining lung lobes post-operation inevitably leads to a certain degree of pulmonary hypertension. Consequently, Zhang's case[10] resulted in rapid right heart failure and hemoptysis, leading to death on the second day after surgery. Among three successful patients, one underwent right middle lobectomy while two underwent wedge resection; however, even with small resections, long-term oxygenation decline may occur post-surgery for these patients. In Liu's case,[15] simple activities can cause significant dyspnea and decreased oxygenation, which improved slowly over a 10-month follow-up period after surgery. So, when encountering tumors located in UAPA on the contralateral side, careful consideration should be given before opting for surgery; alternative treatments, such as radiofrequency ablation, can be considered to minimize lung function loss. If surgery is still deemed necessary despite the potential risks mentioned above, extracorporeal membrane oxygenation (ECMO) preparation should be made for possible dips in oxygenation during the procedure, and home oxygen therapy should be arranged thereafter.

Conclusion

In summary, we reported a case of lung cancer with ipsilateral pulmonary artery absence and reviewed another 13 cases of such patients reported in the literature so far. The choice of surgical procedure for such patients should be determined based on preoperative examination and intraoperative exploration. For this patient, sublobectomy should be the right choice.

Declarations

Informed consent

Informed consent was obtained by the patient for publication of this report.

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Author contributions

Jiangshan Ai contributed to the research concept, drafted the article, and approved the submitted and final versions. Lianzheng Zhao contributed to data acquisition and analysis, and the approval of the submitted and final versions. HuiJiang Gao contributed to data acquisition and analysis, and the approval of the submitted and final versions.

Disclosure statement

All authors have no conflicts of interest.

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