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Case Report

Absence of left pulmonary artery with the ipsilateral lung cancer and right aortic arch: a case report

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Abstract: Unilateral pulmonary agenesis is a medical condition characterized by the unilateral absence of pulmonary artery (UAPA). It is a rare congenital condition caused by malformation of pulmonary vascular system. Such patients often develop other congenital heart diseases, such as Tetralogy of Fallot, patent ductus arteriosus. In this report, we have described a 56-year-old male patient, who experienced an intermittent pain in the left side of the chest for two months. He also experienced persistent back pain. This patient was diagnosed with UAPA and ipsilateral lung cancer. This was an extremely rare case worldwide. After performing a contrast-enhanced lung CT, vascular imaging technology, bronchoscopy, and pathological examination are used to make a clear diagnosis of lung cancer, which was complicated by left pulmonary artery agenesis and a right aortic arch. The patient was treated with left lung resection. The postoperative recovery of the patient was good as he did not develop any complications. Therefore, the patient was discharged as his prognosis was positive.

Keywords: Unilateral pulmonary agenesis/UAPA, lung cancer, aortic arch, pneumonectomy

Introduction

Unilateral pulmonary agenesis is a condition in which there is unilateral absence of pulmonary artery (UAPA). In such patients, although the pulmonary valve exists, there is no branch from the pulmonary artery trunk. Therefore, the pulmonary valve directly connects with one side of the pulmonary artery and the corresponding lung. The pulmonary artery is absent on the other side as it originates from the aorta. Unilateral pulmonary agenesis is a rare congenital malformation of the pulmonary vascular system, and it is often associated with other congenital heart diseases, such as Tetralogy of Fallot, patent ductus arteriosus [1]. Pool *et.al* [2] first reported about UAPA in 1868. Its incidence is approximately 1 in 20 million people. Most simple cases of UAPA have been reported from a radiological point of view. In our hospital, we came across a case of UAPA, with complications of a right aortic arch and squamous cell carcinoma in the ipsilateral lung. Since this UAPA case had complications, it was an extremely rare case in the world. In this case report, we have discussed the impact of UAPA on the development and progression of cancer in ipsilateral lung. We have also investigated the relationship between left pulmonary agenesis and right aortic arch. Thereafter, we have suggested solutions for treating this anomaly.

Case report

A 56-year-old male patient was hospitalized since he experienced intermittent pain on the left side of the chest for about two months. He would cough and eject out yellow sputum and bloody sputum occasionally.

The patient had his first massive hemoptysis (about 180 ml) about 50 years ago. However, he did not undergo any medical treatment due to his impoverished family conditions. He was prescribed a hemostatic medication to ease out hemoptysis. In the past 30 years, the patient had recurrent cough with yellow sputum and hemoptysis (about 80 ml/each time). Twenty years ago, he started to use Chinese herbs that gradually alleviated the symptoms of hemoptysis. The patient also had a history of

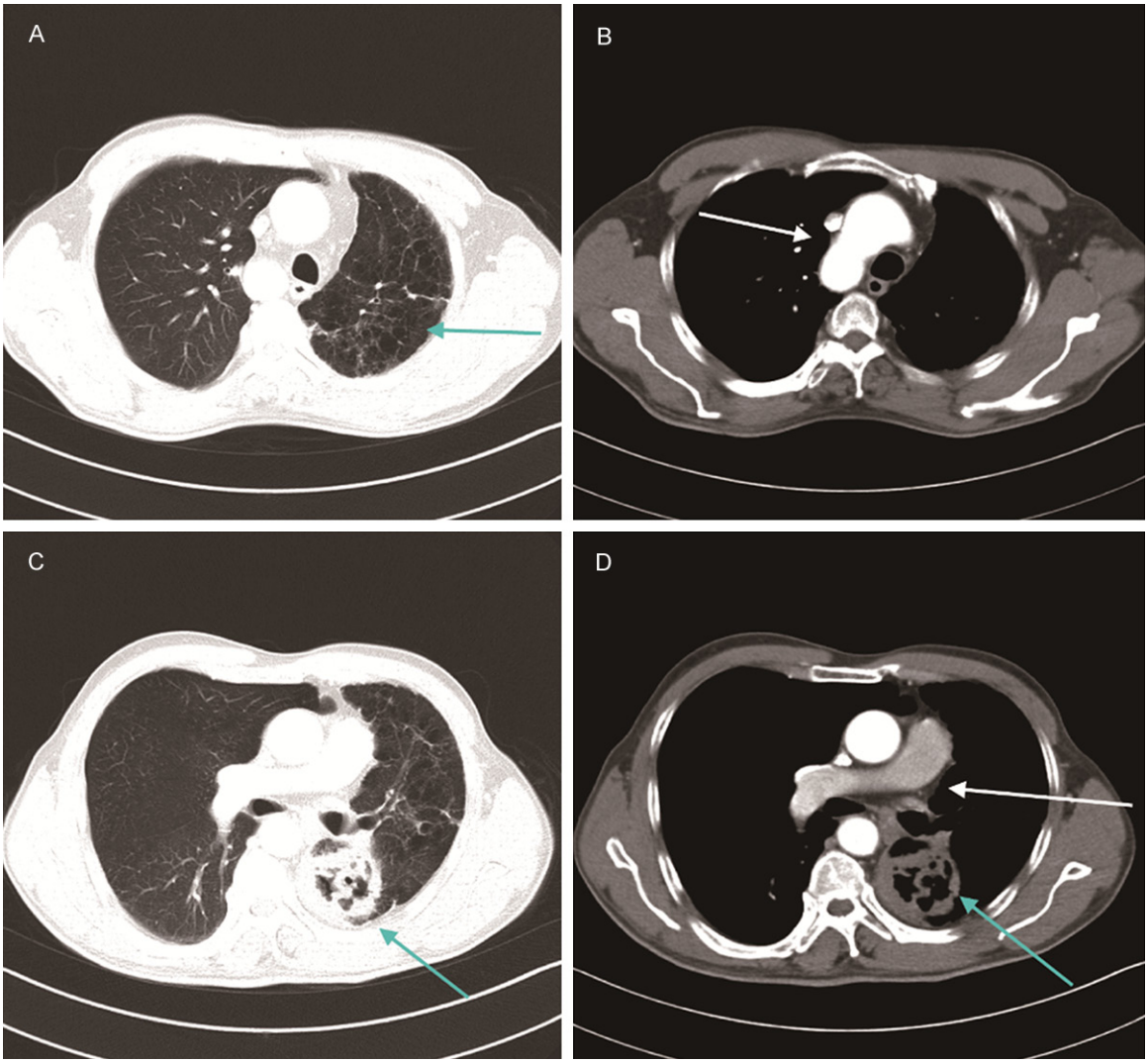

Figure 1. Lung CT scan. A. The CT imaging showed an enhanced texture of both lungs, multiple cysts, and emphysema-like changes (arrow) in the lobes of the left lung. B. The aortic arch is located in the right chest (arrow). C. The dorsal section of the lower left lung showed an occluded bronchus and a high density agglomeration around the bronchus (arrow). D. The lobe of the lower left lung showed a highly dense agglomerate, containing irregular cavities and liquid shadow (blue arrow). There was no appearance of left pulmonary artery (white arrow).

smoking for 35 years. He would smoke about 25-35 cigarettes each day. The auscultation sounds indicated that breathing was slightly weaker in the left lung. Moreover, we could also hear moist rales. The electrocardiogram (ECG) showed multiple premature atrial contractions, ST segment elevation (V1-V3), and left ventricular hypertrophy. The lung function tests a forced expiratory volume in the first second of 2.23 L (accounting for 69% of the predicted value) and pulmonary reserved volume is 84%,and residual volume/total volume ratio is increased. The arterial blood gas analysis as follows: pH: 7.33, PO2: 70.8 mmHg, PCO2: 36.8 mmHg, HCO3act: 21.9 mmol/l. In cytokeratin 19 fragment, the level of tumor markers was 9.35 ng/ ml (< 5.00). Lung enhanced CT showed trachea and mediastinum located left slightly, an enhancement of the texture of lungs, and emphysema-like changes in the left lung, the occlusion of the superior segment of the left lower lobe in the bronchus, a high density agglomeration in the irregular cavity, and liquid shadow. The left pulmonary artery did not appear, and the aortic arch was on the right chest (**Figure 1**). The 3D vascular imaging technology detected left pulmonary artery agenesis and a right aortic arch (**Figure 2**). The bronchoscopy showed that

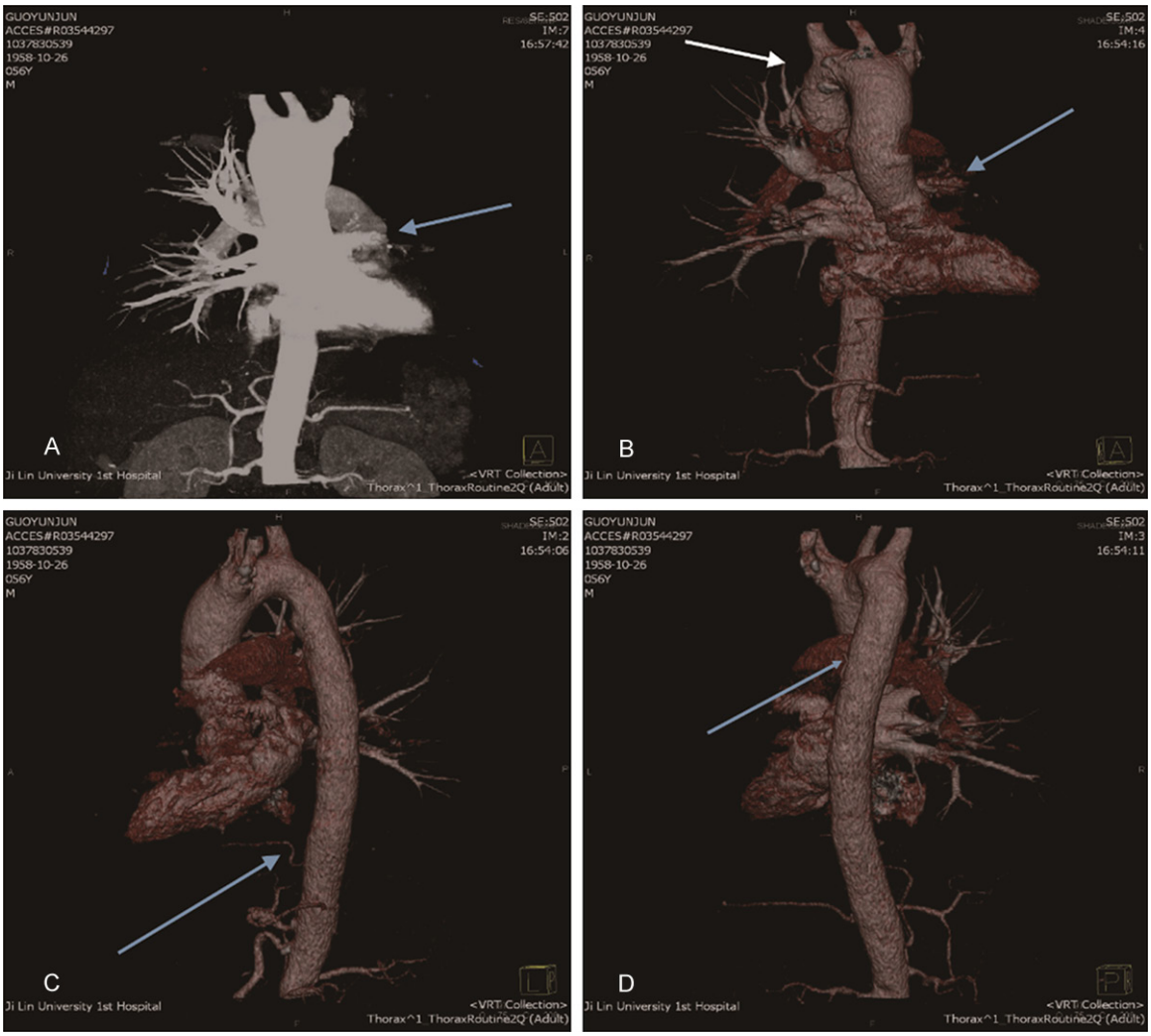

Figure 2. 3D model of an absent left pulmonary artery. A. Reconstruction of a coronal diagram depicting the absence of left pulmonary artery (arrow). B. Vascular 3D reconstruction technique showed left pulmonary artery agenesis (blue arrow) and a right aortic arch (white arrow). C and D. Vascular 3D reconstruction techniques showed blood vessels in the left lung (blue arrow).

mucosal congestion, swelling, and roughness in the superior segment of the left lower lobe's bronchial orifice had almost blocked the lumen. The biopsies from the posterior segment of the left lower lobe, and the pathological results indicated the presence of multiple cell nests, which were atypical in shape. All these observations indicated the possibility of cancer. This was confirmed by examining the cytology of exfoliated squamous cells.

Based on the CT scan and the patients' symptoms, we excluded the following two possibilities: tumor compression in the left pulmonary artery and the development of thromboembolism in the left pulmonary artery. The patient was diagnosed with squamous cancer in the left lower lobe, left pulmonary artery agenesis, and a right aortic arch.

When we performed thoracic surgery, the aortic arch couldn't be found in the left chest; however, there was a gray colored thickening, which showed poor compliance with the left lung pleural. Most parts of the lung had emphysema-like changes in the vesicles; the tumor was located in the superior segment of the left lower lobe. The tumor was hard, and invaded the posterior chest wall. It was about 6 cm × 6 cm × 4 cm in size. After sequentially ligating and cutting blood vessels in the inferior pulmonary ligament and pulmonary vein, hilum was exposed

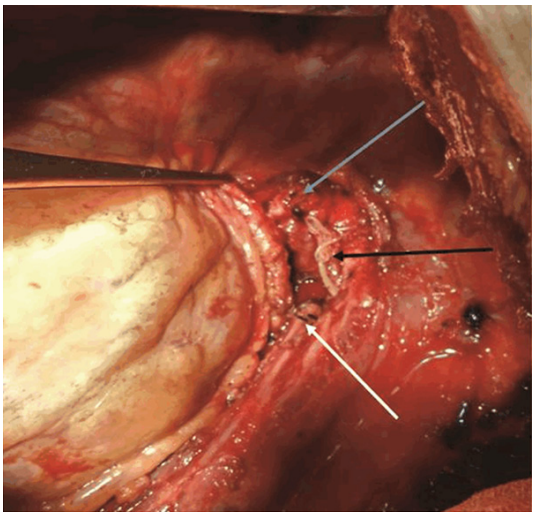

Figure 3. Mediastinal imaging after pneumonectomy showed a supra pulmonary vein stump (blue arrow), an inferior pulmonary vein stump (white arrow), bronchial stump (black arrow), and absence of a left pulmonary artery.

and we opened the mediastinal pleura. While performing the preliminary separation of hilum, we saw supra pulmonary veins; they had a small diameter of about 0.5 cm. The left pulmonary artery could not be seen, and there were no sheath and ligaments (**Figure 3**). Even with further finger palpation, the left pulmonary artery could not be detected. After ligation, the superior left pulmonary vein was cut. At this stage, we found two tortuous thickenings in the bronchial arteries; they were about 0.4 cm in diameter. With ligation and cutting, the left main bronchus were fully exposed. After cutting off the left main bronchus with tracheal occluder, left pneumonectomy was performed. Based on the pre-operative findings of three-dimensional imaging techniques (**Figure 2**), the bronchial artery from the aorta were considered as blood vessels of the left lung. During the surgical procedure, the patient did not develop any complications, and he was discharged after surgery. The pathology report showed squamous cell carcinoma in the left lung.

Discussion

This patient had a rare congenital cardiovascular malformation as he did not have a pulmonary artery. Such a congenital defect occurs either unilaterally or bilaterally. The infants with bilateral deficiency always die as soon as they are born, so UAPA cases are more common. UAPA are mainly located on the side opposite to the main artery arch, so this congenital malformation is more common on the right side. This patient is special in that he has a right aortic arch and a contralateral UAPA. Simple UAPA is rare, and most cases are children with malformations (about 78%). These patients often die young. Simple UAPA patients survive relatively long, and the reported maximum age is 68 years [3]. The prognosis depends on the severity of other cardiovascular malformations and pulmonary hypertension [4]. This patient was 56 years old. Since the UAPA and right aortic arch anomalies did not have a greater impact on the physiology and blood flow, the patient could survive from recurrent hemoptysis in previous years. Therefore, this is a very rare condition.

UAPA patients often have clinical manifestations of hemoptysis, recurrent pulmonary infections, chest pain, cyanosis, and dyspnea. In this case, the patient not only experienced typical hemoptysis and recurrent pulmonary infection but also UAPA and ipsilateral lung cancer. The etiology of UAPA and ipsilateral lung cancer could be related. Alveolar hypocapnia can cause bronchial spasms, reduced mucociliary clearance, and accumulation of inflammatory cells, thereby increasing the chances of lung infection [5]. UAPA can cause repeated lung infections, inflammation, delayed healing, and a high level of long-term inflammatory mediators, resulting in DNA damage of bronchial epithelial cells, cell mutation, vascular proliferation, squamous cell metaplasia, and ultimately cancer. However, this patient had been smoking for a long period time, a habit that aggravated the development of lung cancer.

We do not yet know whether the etiology of left pulmonary artery agenesis is related to the right aortic arch. During the fifth and sixth week of gestation period, the pulmonary arteries derive from the proximal portion of the sixth aortic arch. A right aortic arch is considered to be relevant to the aberrant formation of a ductus arteriosus which passes from the right pulmonary artery (instead of the left) to the right aortic arch. This congenital formation occurs within 5-6 weeks of pregnancy and concomitantly with persistence of the right aorta and the degradation of the fourth aortic arch [6]. Since the development of right aortic arch is

often associated with agenesis of left pulmonary artery [7], some factors are likely to trigger a pulmonary artery distortion within 5-6 weeks of pregnancy, which ultimately leads to left pulmonary artery agenesis and right aortic arch.

It is important to diagnose UAPA at an early stage. In this case, we have used a contrast-enhanced lung CT and 3D revascularization technology to make a clear diagnosis. Furthermore, we tackled the situation by treating lung cancer through thoracotomy and intraoperative pulmonary exploration.

During the surgical treatment, UAPA was corrected by revascularization, selective embolization, lobectomy, and pneumonectomy. Lobectomy or pneumonectomy is performed only on the bleeding site of the ipsilateral bronchial artery; however, there is no golden standard for patients with ipsilateral lung cancer and UAPA. Thus, it was a challenging case that necessitated strict adherence to pneumonectomy indications. Due to vascular malformations, there was absence of left pulmonary artery and a right aortic arch in this patient. Therefore, we decided to perform left pneumonectomy. In this procedure, we cut the left superior and inferior pulmonary vein along with the bronchial artery to ensure safe blood supply to the lungs. Left pneumonectomy was performed smoothly. The patient recovered post-operatively. Since there were no postoperative complications, the patient was ultimately discharged from the hospital.

In conclusion, only four cases of UAPA with lung cancer have been reported worldwide, including three cases of lung cancer in the ipsilateral side of UAPA and one case of lung cancer in the contralateral side of UAPA. All the cases are treated by performing pulmonary resection surgery. Three cases were not associated with other cardiovascular malformations [8-10], but there was one UAPA case with a right aortic arch and left lung adenocarcinoma in the upper lobe [11]. Therefore, we believe this is the first case of UAPA with pulmonary squamous cell carcinoma and right aortic arch. The preoperative diagnosis of UAPA with lung cancer is particularly important, because only clinicians with extensive experience could handle the surgical treatment of this patient. There was no standard protocol for treating this patient as this was a very rare case. Therefore, we had to implement an individualized treatment plan, which was most suitable for this specific case. We hope the diagnosis and treatment of this case would help in solving similar cases in the near future.

Disclosure of conflict of interest

None.

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