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CASE REPORT

A Young Woman with Hemoptysis as the main Manifestation of Isolated UAPA with Multiple Vascular Malformations

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SUMMARY

Background: **The unilateral absence of a pulmonary artery (UAPA) is a very rare congenital pulmonary vascular malformation. Since the first case was reported in 1868, the overall incidence of UAPA has been at a level of 1/300,000 - 1,200,000. Isolated UAPA without other cardiac anomalies is rare. Here we report a young woman who came to the clinic with hemoptysis as the main manifestation. Isolated UAPA with multiple vascular malformations was finally diagnosed by DSA angiography. The diagnosis and treatment experience and gains of UAPA are discussed in combination with the treatment process of this patient.**

Methods: **Appropriate laboratory tests, chest computed tomography (CT), Computed Tomography Pulmonary Angiogram (CTPA), and DSA angiography of the pulmonary arteries were used to explore the etiology.**

Results: **Chest CT showed that the bronchial tubes in the right lung segment and below were not obvious, and CTPA showed that the pulmonary arteries in the middle and lower lobes of the right lung were not visible, and the diagnosis of isolated unilateral pulmonary artery agenesis with multiple vascular malformations was clearly made by pulmonary artery DSA angiography.**

Conclusions: **In patients with unexplained hemoptysis, we should be alert to the possibility of vasculopathy, and angiographic techniques should be applied in a timely manner when plain CT fails to make a definitive diagnosis.**
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KEYWORDS

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CASE REPORT

A female patient, 33 years old, was admitted for 2 days mainly due to intermittent hemoptysis. The patient showed no obvious reason for intermittent hemoptysis 2 days before admission, the amount was small, about 3 - 4 times a day, and the total amount was about 5 mL. In most cases, there is blood in the sputum and a little blood in the mouth. When hemoptysis occurs, the patient may have an intermittent cough, no fever, chest pain and dyspnea, no abnormal bleeding of the gums, skin and mucous membranes, no restricted movement, etc. A chest CT scan revealed multiple patchy high-density shadows in the right lung. Considering inflammato-

ry lesions, a small amount of pleural effusion on the right and adhesion of the right pleura was seen. Oral azithromycin for 3 days was ineffective. She still had hemoptysis intermittently. She was then treated in our hospital. Previous patients have denied basic medical histories such as congenital heart disease, rheumatic heart disease, hypertension, diabetes, and blood system diseases. However, the patient reported that she had repeatedly had right-sided interstitial pneumonia over the past few years, and she had several visits. After symptomatic anti-inflammatory treatment, her condition was relieved, but no hemoptysis occurred. She is married with 2 sons and still breastfeeding. On admission, the vital signs were stable. Only auscultation could be heard and the right lung breathing sound was lower than that on the left. No significant positive signs were found in the rest. The abnormal indicators in the laboratory examination are only the blood routine leukocytes 10.3 x 10^9/L, neutrophil percentage 83%, and monocyte percentage 85%. No obvious abnormalities in the remaining items.

After visiting the hospital, the patient CT imaging data from a different hospital was viewed and showed that the bronchus in the right lung segment and below was not obvious, and the left part of the lung tissue had obvious low-density translucent images. A review of CT pulmonary angiography (CTPA) reconstruction showed that the right and middle lobe pulmonary arteries were not visualized, the right lung bronchial artery was enlarged, the right pulmonary veins were delayed in development, the density was not uniform, and the left pulmonary artery trunk and its branches were well filled (Figure 1). At this time, the patient still had hemoptysis intermittently. To further clarify the cause, after fully communicating with the patient, DSA angiography was performed on the right bronchial artery, inferior iliac artery, internal thoracic artery, and pulmonary artery. During the operation, the right middle lobe, lower lobe, and some upper lobe pulmonary arteries were seen to be absent. The right two bronchial arteries, the right inferior iliac artery, and the right internal thoracic artery were tortuous, thickened, and multiple arterial fistulas formed by the above, and residual pulmonary arteries were seen. So, the specific cause of hemoptysis in patients is clear (Figure 2, 3).

DISCUSSION

Unilateral absence of pulmonary artery, as a rare pulmonary vascular disease, has been pointed out that its cause may be related to the continuous connection between the proximal sixth aortic arch and the pulmonary artery and the distal sixth aortic arch [1,2]. However, most patients have congenital heart defects such as ventricular septal defect and insufficiency of arterial catheters. Isolated UAPA without any manifestation of cardiac damage is even more rare. As in this case [3], patients with isolated UAPA are more likely to have a missing right pulmonary artery [4]. Most patients with isolated UAPA are found in childhood as a result of congestive heart failure [5]. A small number of people can survive to adulthood without symptoms, but the main clinical manifestations of impaired exercise tolerance and dyspnea after exercise are found, and the majority of patients are elderly [4]. This patient was a young woman. Prior to this diagnosis, there were no obvious symptoms of cardiac impairment such as dyspnea after exercise. The recurrence of right interstitial pneumonia in previous patients may be related to the absence of pulmonary arteries on the affected side. A retrospective study [6] showed that patients with solitary UAPA are often confused with old tuberculosis, chronic pulmonary embolism, pulmonary vasculitis, and Swyer-James-Macleod syndrome due to non-specific clinical manifestations. In the progression of these diseases, recurrent occlusive bronchial inflammatory changes may occur, pulmonary ventilation is reduced, angiography of the pulmonary artery and its branches show they are small or occluded [7]. However, after this patient was admitted to hospital, mycobacterium tuberculosis infection and pulmonary vasculitis were gradually eliminated through a series of examinations, and the patient did not have a long history of cough and pulmonary infection during childhood, so the Swyer-James-Macleod syndrome was further excluded. The patient repeatedly suffered from pulmonary infection on the affected side in the past. At present, studies have identified the following two causes [5,8]: First, UAPA patients have other collateral circulation blood supply due to pulmonary artery disease. However, there may be obstacles to inflammatory cell transmission that cannot reach the site of inflammatory damage to the lung tissue. Second, insufficient blood supply to the lung will cause abnormal pulmonary blood flow and ventilation ratios, causing intrapulmonary ventilation disorders and local hypocapnia in the lung. This abnormality can lead to secondary bronchoconstriction and can cause bronchial mucosal cilia dyskinesia which prevents the mucus from being effectively removed, leading to chronic lung inflammation and bronchiectasis in the long run. This also shows that for this patient, the correct anti-inflammatory and phlegm-reducing treatment to promote airway drainage is very necessary.

In addition to recurrent lung infections, the main clinical manifestation of the patient at this visit was hemoptysis. Studies have shown that pulmonary artery occlusion can promote the generation of blood vessels in the pulmonary system on that side and increase blood flow to the corresponding lung tissue [9]. The time and extent of pulmonary outflow dysplasia may affect the distribution of the origin of collateral arteries [5]. Combined with the existing literature, it is found that the pulmonary collateral circulation in the affected side of UAPA patients mostly originates from the bronchial artery, but also originates from the sub-iliac artery, internal thoracic artery, subclavian artery, and even coronary arteries [4,5,9]. After DSA angiography in this patient,

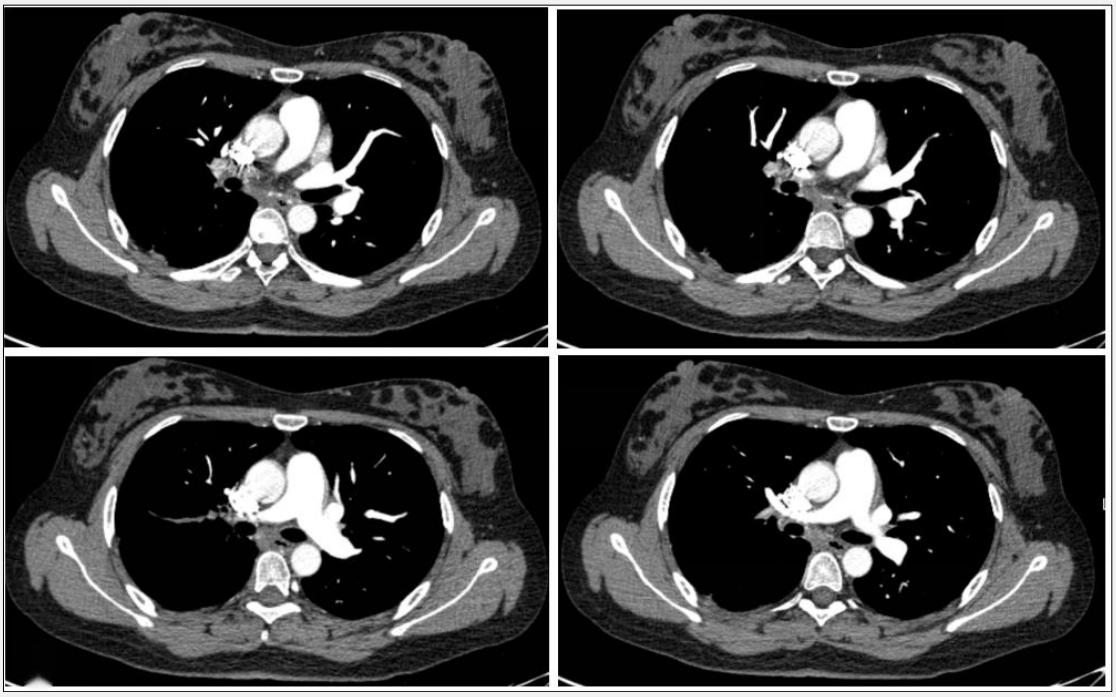

Figure 1. CTPA: The right and middle lobe pulmonary arteries were not visualized.

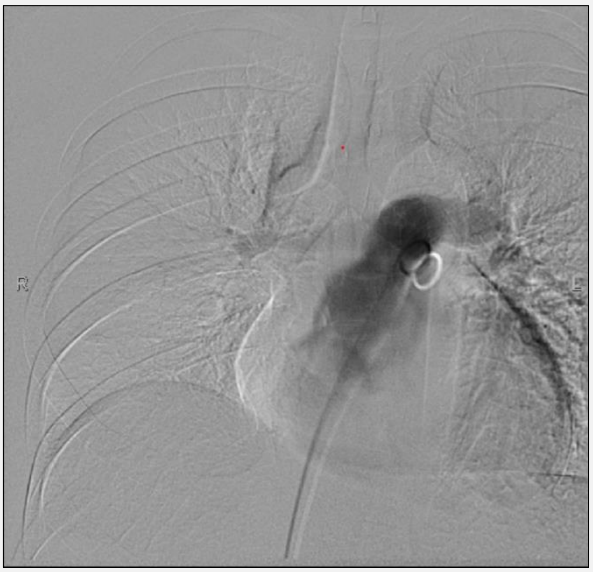

Figure 2. DSA: The right middle lobe, lower lobe, and some upper lobe pulmonary arteries were seen to be absent.

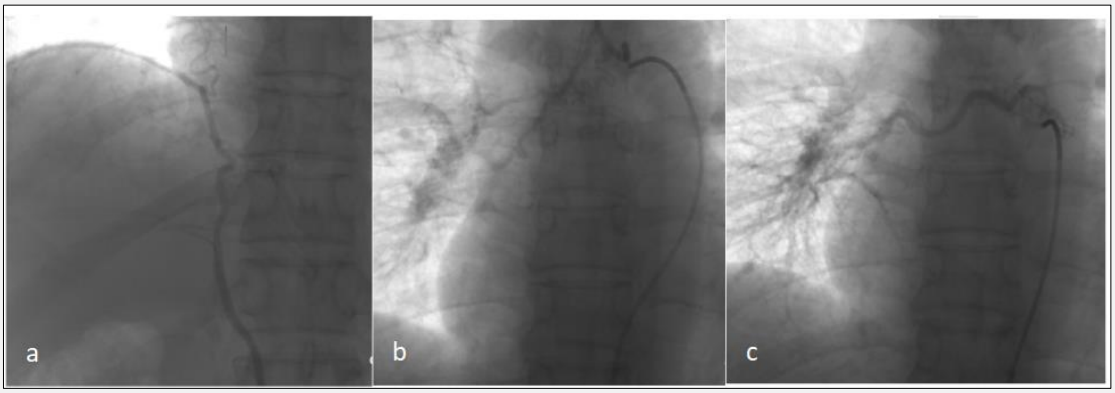

Figure 3. The right two bronchial arteries, the right inferior iliac artery, and the right internal thoracic artery were tortuous, thickened, and multiple arterial fistulas formed by the above. Residual pulmonary arteries were seen.

it was found that the collateral circulation in the affected side of the patient originated from the right bronchial artery, inferior iliac artery, and internal thoracic artery. It occurred in multiple sources and is rarely found in existing literature. However, bronchiectasis, lung lattice changes, and pulmonary cystic changes in UAPA patients due to prolonged hypoperfusion of intrapulmonary blood flow have been reported [2,5]. With age, pulmonary collateral circulation becomes more common [10]. Due to long-term damage to the structure of the lungs, the uneven intravascular pressure, the long-term chronic inflammatory stimulation leading to the damage of the vascular wall, the formation of collateral circulation vascular fistula can cause rupture of the blood vessels and bleeding. DSA angiography also proved that hemoptysis in this case was bleeding due to collateral vascular malformations and vascular fistula formation, which is consistent with most cases reported in the previous literature. However, due to the multi-source collateral blood supply compensation, the patient's lung parenchyma did not show obvious infarcts, dysplasia, or other manifestations. This may be one of the reasons for the delay in the onset of the disease since childhood.

For the treatment of hemoptysis in patients, we have adopted conservative treatment schemes such as allowing patients to take Yunnan Baiyao orally. The first is because the patient has a short hemoptysis time and a small amount of hemoptysis, which can stop the bleeding on its own. It has not affected the patient's normal coagulation mechanism and does not affect the patient's normal physiological activity; Second, according to previous literature reports, for UAPA patients, arterial interventional embolization has little trauma to hemostasis, and the bleeding site can be directly embolized to achieve hemostasis. However, due to the continuous generation of collateral circulation or excessive bleeding sites, local embolism cannot achieve the goal of radical cure. Arterial embolism has a high recurrence rate for hemostasis [11,12]. However, in this case, the patient was younger and the diseased blood vessels involved all the right lung lobes. Although traditional surgical lung lobectomy can achieve radical treatment, after lung lobectomy, it will inevitably affect the patient's normal physiological function and quality of life [13, 14], and thus affect the patient's prognosis. Other treatments, such as anastomosis of the collateral artery with the central pulmonary artery and staged repair of the affected pulmonary artery are still under study [2,8,12]. However, if acute massive hemoptysis is life-threatening for patients in the short term, lobectomy may be the best choice to save lives.

It is worth noting that pulmonary hypertension is one of the most common serious complications in patients with UAPA [4,14,15]. The absence or incompleteness of the affected pulmonary artery leads to an increase in blood flow to the contralateral pulmonary artery, which in turn promotes the release of endothelin from vascular endothelial cells to stimulate vasoconstriction, while chronic contraction of the pulmonary arterioles can rebuild pulmonary blood vessels, leading to pulmonary vessels and pulmonary arterial resistance increases, which leads to the occurrence of pulmonary hypertension. However, some studies suggest that pulmonary hypertension in UAPA patients generally occurs early, and they may die because of right heart failure. However, if pulmonary hypertension does not progress in the early stage, the possibility of subsequent occurrence will gradually decrease [2,15]. In this case, no significant pulmonary hypertension is present and the condition is stable. Long-term follow-up observation is needed for follow-up treatment and development of the patient. Early identifi-

cation and early treatment of UAPA patients may be more helpful for the patients' early benefits.

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Ethical Approval:
This study was approved by the ethics committee of North China University of Science and Technology Affiliated Hospital. All procedures performed in the study were in accordance with the ethical standards. Informed consent was obtained.

Declaration of Interest:
No conflicts of interest.

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