Case report

Unilateral pulmonary vein atresia: Literature overview and case report

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Abstract

The unilateral absence of the pulmonary vein, known as pulmonary vein atresia, is a rare type of unilateral pulmonary venous hypoplasia caused by the congenital atrophy of the long pulmonary vein segments in one lung. The involved lung may be normal in size or present with hypoplasia and is often characterized by increased interstitial attenuation and interlobular septal thickening due to venous stasis, edema, and fibrosis. Pulmonary angiography often reveals a reduced size for the lateral pulmonary artery, peripherally sparse pulmonary vessels, contrast stasis, and the inability to visualize pulmonary veins. Symptoms include coughing up blood and infection. We present the clinical case of a patient who was initially diagnosed with recurrent hemoptysis due to pulmonary tuberculosis, followed by unsuccessful treatment. Imaging by 64-slice computed tomography with contrast injection using multiplanar reformation and volume rendering techniques allowed this case to be definitively diagnosed. This report emphasizes the epidemiological factors and clinical and imaging features of unilateral pulmonary vein atresia to prevent confusion and facilitate proper diagnosis in similar cases.

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Introduction

The absence of a unilateral pulmonary vein, known as unilateral pulmonary vein atresia, is a rare congenital anomaly. Common clinical symptoms include hemoptysis and infection, which can result in misdiagnosis as pulmonary tuberculosis [1]. Pathological examination can reveal the absence of pulmonary veins between the segmental veins and the four pulmonary veins that drain into the left atrium. Imaging plays an essential role in the proper diagnosis of this condition. The size of the ipsilateral lung may be normal or small, and affected lungs are often characterized by increased interstitial density and interlobular septal thickening due to ve-
nous stasis, edema, and fibrosis. Computed tomography (CT) angiography reveals a lack of pulmonary veins, small ipsilateral pulmonary arteries, sparse pulmonary vascular density in the periphery, and contrast stasis. Radioisotope imaging shows decreased pulmonary perfusion [1,2]. In this article, we present a case of unilateral pulmonary vein atresia to emphasize the epidemiological factors, clinical signs, subclinical symptoms, and imaging approaches that should be considered to help doctors achieve the correct diagnosis in similar cases.

Case report

A 19-year-old male patient was admitted to the hospital due to coughing up blood, with a childhood history of respiratory infection episodes. At the age of 13, he reported coughing up a small amount of blood once and was diagnosed with and treated for pulmonary tuberculosis following a 9-month regimen of continuous drug administration. During subsequent years, the patient continued to experience several episodes during which the patient coughed up small amounts of blood, which was diagnosed as pulmonary tuberculosis sequelae and treated with antibiotics. Two weeks prior to admission, the patient experienced fever, coughed up approximately 200 mL of blood. The patient was admitted to a district hospital, where the patient was diagnosed with and treated for pulmonary tuberculosis according to the prescribed regimen, after which the patient stopped coughing up blood. Then, the patient experienced a continuously high fever that was not alleviated by antipyretic treatment and was transferred to the National Lung Hospital (negative rRT-PCR of COVID-19 and two-dose mRNA COVID-19 vaccine).

On examination at admission, the patient had a fever (>38°C). Pulse and blood pressure were within normal limits, with no dyspnea, and the left lung sounded more hypoventilated than the right lung. The results of blood tests showed normal blood biochemistry and C-reactive protein levels. The acid-fast bacillus sputum test was negative. One lymph node was identified 10 mm posterior to the left sternocleidomastoid muscle. Anterior chest X-ray results suggested left pulmonary fibrosis. For diagnosis and follow-up of the negative acid-fast bacillus pulmonary tuberculosis test, the patient was sent to the pulmonary tuberculosis department for treatment. The patient continued to be treated for tuberculosis but continued to display irregular hemoptysis. Dissecting the lymph nodes behind the sternocleidomastoid muscle resulted in inflammatory lymph node hyperplasia.

The patient received two chest X-rays, one at the time of hospital admission and another 1 week later. The X-ray results and the radiologist’s interpretations are shown in Figure 1.

The patient underwent 2 chest CT scans, one at the time of hospitalization and another 1 week later. All CT images were obtained on a 64-slice machine, and images were obtained both before and after contrast injection, at a slice thickness of 3 mm, reconstructing all 0.75 mm windows. Thin slices were used to perform multiplanar reformation (MPR) and volume rendering technique (VRT) series. The CT results and the radiologist’s interpretations are shown in Figures 2 and 3.

Fig. 1 – Chest X-ray on admission (A) and 1 week later (B). Decreased left lung volume, mediastinum displaced to the left, with the left diaphragm higher than the right diaphragm. The left lung is less bright than the right lung, with a reticular opacity and ground-glass opacity in the middle-third area.

Discussion

The absence of the pulmonary vein is a rare congenital abnormality, with 50% of cases presenting alone and the remaining 50% of cases presenting in the setting of lung diseases associated with other congenital abnormalities. Pulmonary vein atresia can occur on either side, with no clear predominance, although this typically presents unilaterally. Atresia likely occurs due to errors when combining the pulmonary vein network during development, resulting in the development of a larger vein that empties into the left atrium. Although pulmonary vein atresia is typically diagnosed in neonates, often in the setting of an abnormal hypoplastic lung, it can also present in adults. The most common clinical manifestations are recurrent infections in the lungs, low-level, irregular but repeated episodes of hemoptysis, which may result in vomiting blood, followed by pulmonary hypertension indicators. Mortality can be as high as 50% among untreated patients [1,3,4,5].

Two explanations have been proposed to explain the presence of a thickened interstitial septum in cases of pulmonary artery atresia. (1) The pulmonary vein is typically only absent from the segmental vein, with the retention of the pulmonary vein system in the interlobular septum, allowing the pulmonary circulation to continue supplying blood to this venous network, resulting in venous stasis and the development of a thickened interlobular septum. (2) Due to incomplete venous circulation, blood that reaches the venous system of the interlobular septum undergoes the phenomenon of returning to the arterial circulation, resulting in the appearance of pulmonary hypoperfusion on radioisotope imaging [2]. The thick-
enling of the septum may also be related to pulmonary lymphedema. Pulmonary fibrosis can occur due to a combination of pulmonary venous infarction and chronic pulmonary edema [6]. CT scans revealed the lack of pulmonary veins and the sparse density of the ipsilateral pulmonary artery. In addition, angiographic studies have demonstrated pulmonary blood supplied by the bronchial artery and other systemic artery branches [4,7]. Although the development of arteries supporting the bronchial artery blood supply has been acknowledged in some cases of pulmonary venous insufficiency, to our knowledge, this finding is not detectable on CT angiography, which was also highlighted in previous reports. This condition was not detected in the current case.

The lateral pulmonary artery is believed to be narrowed in these cases due to preferential pulmonary perfusion to the adjacent lung, leading to impaired growth of the affected pulmonary artery [1,5], which may also cause bronchial and systemic arterial proliferation. Pulmonary vein obstruction can also lead to the formation of bronchial varicose veins [3,5], and the rupture of bronchiectasis is a common cause of hemoptysis in these patients. Although most patients present in childhood, adult manifestations have also been reported, and adult patients with this condition are often difficult to diagnose [8]. Parenchymal consolidation at the left atrium border of the mediastinum may suggest an inflammatory parenchymal lesion causing fibrosis that obstructs the ipsilateral pulmonary artery and pulmonary vein [7]. However, the presence of a small mediastinal lymph node without evidence of bronchial obstruction should suggest a congenital malformation as the potential underlying cause of chronic inflammation [6,9]. The consolidation of the parenchyma may be attributed to the production of replacement tissue by the pulmonary veins. The noninvasive diagnosis of unilateral pulmonary vein atresia can be performed by CT angiography, cardiac magnetic resonance imaging (MRI), or bronchoscopy [10,11]. Similar to cardiac MRI, chest CT can provide precise morphological information regarding the structures of the heart and extracardiac vasculature [9,12,13]. In our case, conventional cardiac MRI and angiography were not performed because both venous and arterial abnormalities were clearly visible on 64-slice CT [4,14]. We emphasize the ability of multislice CT to establish an accurate diagnosis of pulmonary venous insufficiency, eliminating the need for conventional angiography.

Dixit et al. reported 2 cases of unilateral pulmonary vein atresia in children [14]. In the first case, a 7-year-old girl was admitted to the hospital for the evaluation of recurrent chest infections starting at 4 months of age and recurrent hemoptysis starting 4 years prior, with approximately 1-2 episodes presenting each year. Despite no evidence of tuberculosis, the child received two courses of tuberculosis treatment. On admission, the child was unconscious, her hemoglobin level was 10 g%, and the Mantoux test was negative. Chest X-ray showed that the right lung was smaller than the left with a grid opacity, the mediastinum deviated to the right, and the left lung was bright, with a deep rib–diaphragm angle, revealing a state of compensatory ventilation. Chest CT showed thickening of the interlobular septum in most of the right lung, with clear peripheral areas, indicating that the interlobular septum contained pulmonary veins. CT angiography following contrast injection revealed that the right pulmonary artery was half the diameter of the left pulmonary artery. The left atrium clearly presented with two pulmonary veins, but no pulmonary veins were visible on the right atrium. No evidence of bronchial obstruction was noted. A soft tissue mass (consolidation) was
observed adjacent to the left atrium. The prominent right bronchial artery supplies blood to the right lung, along with another artery arising from the thoracic aorta, which were both well-defined on VRT image.

The second case was a 3-year-old girl presenting with a history of recurrent hemoptysis since the age of 1 year [14]. Bronchoscopy revealed mucosal hyperemia and inflammation, but no other endobronchial lesions were detected. The child presented with a hemoglobin level of 12 g% and was in good health. Lung auscultation revealed a slight decrease in the amount of air entering the right side, and chest X-ray revealed a small right hemithorax, with evidence of consolidation in the right middle and lower zones. Multislice CT showed areas of consolidation on the right side and a normal central airway. MPR CT showed a small right pulmonary artery and the complete absence of right pulmonary veins, with the clear visualization of the left superior and inferior pulmonary veins. A smooth left atrial contour and minimal soft tissue were noted. VRT revealed a prominent right bronchial artery and the absence of the right pulmonary vein.

A review of the available literature and clinical cases indicates that unilateral pulmonary vein atresia is a rare congenital abnormality that is often misdiagnosed as pulmonary tuberculosis or as a secondary malignancy in the lung when it does present [1,15]. Clinical signs of infection and repeated hemoptysis are common indicators [16]. Standard chest X-ray reveals a small lung, raised ipsilateral diaphragm, and reticular opacity. Contrast-enhanced multislice CT should be indicated to confirm the absence of pulmonary veins, especially on MPR and VRT series [12]. A small unilateral lung, thickened interlobular septum due to venous stasis, small ipsilateral pulmonary artery, and signs of ipsilateral pulmonary vein absence are imaging features that can help confirm the diagnosis. Although isotope-based imaging to assess pulmonary perfusion plays an important role in lung resection and lung transplantation, this modality does not contribute to the definitive diagnosis of pulmonary vein atresia [13]. Symptomatic treatment, unilateral pneumonectomy, or implementation of a lung transplant program are often suggested to treat these patients [1].

Conclusion

In summary, we report a rare case of congenital pulmonary vascular abnormality. This disease is often detected in adolescence, and the symptoms and clinical course are easily confused with pulmonary tuberculosis, especially in primary health care settings. The radiographic diagnosis can reveal very specific indicators, and multislice chest CT with intravenous contrast injection, combined with imaging reconstruction techniques, can help doctors diagnose this disease.

Availability of data and materials

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

Author contributions

Cung-Van C and Nguyen MD contributed equally to this article as co-first authors. All authors read and approved final version of this manuscript.

Ethics approval

Not applicable.

Patient consent

Written informed consent was obtained from the patient for the publication of patient information in this article.

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