Clinical Case Report

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Hypoplasia of the left lung presenting as hemoptysis

A case report

Lei Zhang, Master[a], Tao Ai, Bachelor[a,*], Li Wang, Bachelor[a], Libin Zhang, Doctor[b], Ying Zhang, Master[a]

Abstract

Rationale: Congenital pulmonary dysplasia (CPD), a congenital lung anomaly, is a heterogeneous group of developmental disorders with diverse clinical and imaging manifestations, including pulmonary agenesis, pulmonary aplasia, and pulmonary hypoplasia. Patients with CPD always have recurrent respiratory tract infections, dyspnea, and/or wheezing. To the best of the author's knowledge, no case of CPD with mild to moderate hemoptysis has been reported so far. Considering the rarity of this presentation, we herein report a case of hemoptysis caused by CPD.

Patient concerns: We report on an 11-year-old girl who survived for 11 years with hypoplasia of the left lung without any associated complications except hemoptysis.

Diagnosis: Left pulmonary hypoplasia.

Interventions: The patient underwent left lower pulmonary lobe resection.

Outcomes: The patient recovered favorably after the operation, and no complications were observed. The hemoptysis was controlled and the cough disappeared after the surgery.

Lessons: CPD is a life-threatening disorder rarely encountered in the clinic, which plays an important role in various pediatric respiratory diseases. Misdiagnosis or delayed diagnosis of pulmonary malformations results in unnecessary treatments and hospitalization. The patients should undergo pulmonary contrast-enhanced CT and bronchoscopy for accurate and timely diagnosis, followed by surgical treatment.

Abbreviations: CPD = congenital pulmonary dysplasia, CT = computed tomography, IPH = idiopathic pulmonary hemosiderosis.

Keywords: hemoptysis, pediatric, pulmonary hypoplasia

1. Introduction

Congenital pulmonary dysplasia (CPD), a congenital lung anomaly, is a heterogeneous group of developmental disorders with diverse clinical and imaging manifestations, ranging from large masses requiring immediate surgical intervention to small and asymptomatic lesions. The incidence of CPD is between 30 and 42 per 100,000 individuals.[1,2] However, there are few accurate reports of the incidence of CPD in China. Patients with concomitant CPD have poor clinical courses due to recurrent respiratory tract infections, dyspnea, and/or wheezing. However, to the best of the author's knowledge, no case of CPD with mild to moderate hemoptysis has been reported so far. Considering the rarity of this presentation, we herein report a case of hemoptysis caused by congenital pulmonary dysplasia.

2. Case report

This study was approved by the ethics committee of the Chengdu Women and Children's Hospital, Sichuan, China. The patient has provided informed consent for publication of this case.

An 11-year-old girl was admitted to Chengdu Women and Children's Central Hospital due to an 8-month history of cough and 2 episodes of mild to moderate hemoptysis (100–200 mL of blood loss over 24 hours). Her cough was occasional and dry at first, but later it became more frequent and productive, without fever, chest pain, wheezing, or dyspnea. She received a 7-day course of amoxicillin and clavulanate potassium for pulmonary infection in a local hospital. Unfortunately, she responded poorly to the treatment. The second episode of hemoptysis had occurred 2 days before admission to our hospital, after which she was referred to us for treatment.

It is noteworthy that the girl had been full-term at birth, healthy, and without tuberculosis or measles.

At the time of admission, the patient was not febrile, and had a respiratory rate of 21 breaths/min, heart rate of 90 beats/min, blood pressure of 101/71 mmHg, and oxygen saturation of 98% in ambient air. The breath sounds were harsh and neither rales nor wheezing were heard. The results of other physical examinations were unremarkable.

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[a] Pediatric Pulmonology Department, [b] The First Department of Pediatric Surgery, Chengdu Women's and Children's Central Hospital, Chengdu, Sichuan, China.

[*] Correspondence: Tao Ai, Chengdu Womens and Childrens Central Hospital, Chengdu, Sichuan, China (e-mail: 534167313@qq.com).

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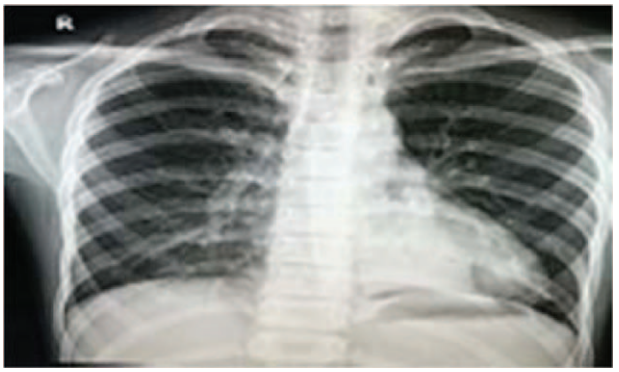

Figure 1. Chest x-ray image showing hyperintensity in the lower left lung.

The white blood cell count was 6.79×10^9/mL, with 69.7% neutrophils, and the hemoglobin level and platelet count were 116 g/L and 381×10^9/mL, respectively. The results of biochemical tests were normal. Tuberculin skin test (TST) was negative.

Serum test for anti-mycoplasma pneumoniae antibodies yielded positive results, with a titer of 1:160. Other pathogens including common respiratory viruses and tuberculosis bacteria were not found.

Chest computed tomography revealed pneumonia and a lung abscess was suspected in the lower lobe of the left lung (Fig. 1). Pulmonary contrast-enhanced CT revealed a dense, cuneate shadow in the left lower lobe with a multiple-capsule shape, and the left lower lobe bronchus was significantly narrow. These were speculated to be congenital malformations (Fig. 2A and B).

Subsequently, the patient underwent bronchoscopy. The diameter of the left bronchus was found to be reduced and a previous pulmonary hemorrhage was observed (Fig. 3). The patient then underwent surgical resection of the left lower pulmonary lobe. Gross pathologic examination of the lung tissue revealed dilation of the bronchia and blood vessels (Fig. 4A and B).

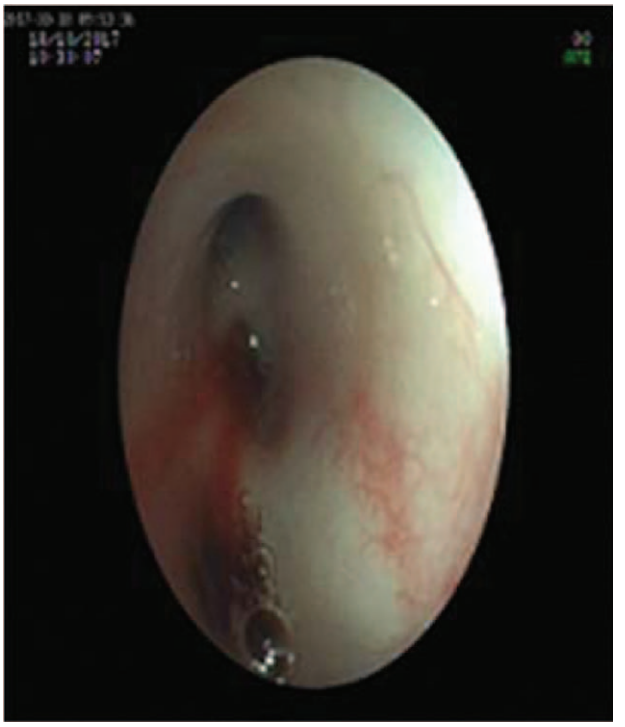

Figure 3. A bronchoscopy image showing previous pulmonary hemorrhage.

In a part of the lung tissue, the bronchia and alveoli were stunted and bronchiectasic. Chronic inflammation of the bronchia had invaded the alveoli with acute episodes, and some of the alveoli were fractured and fused. Hemorrhage and fibrinoid exudation were observed on the pathologic slice (Fig. 5A and B).

The girl recovered favorably after the operation, without any complications. The hemoptysis was controlled and the cough

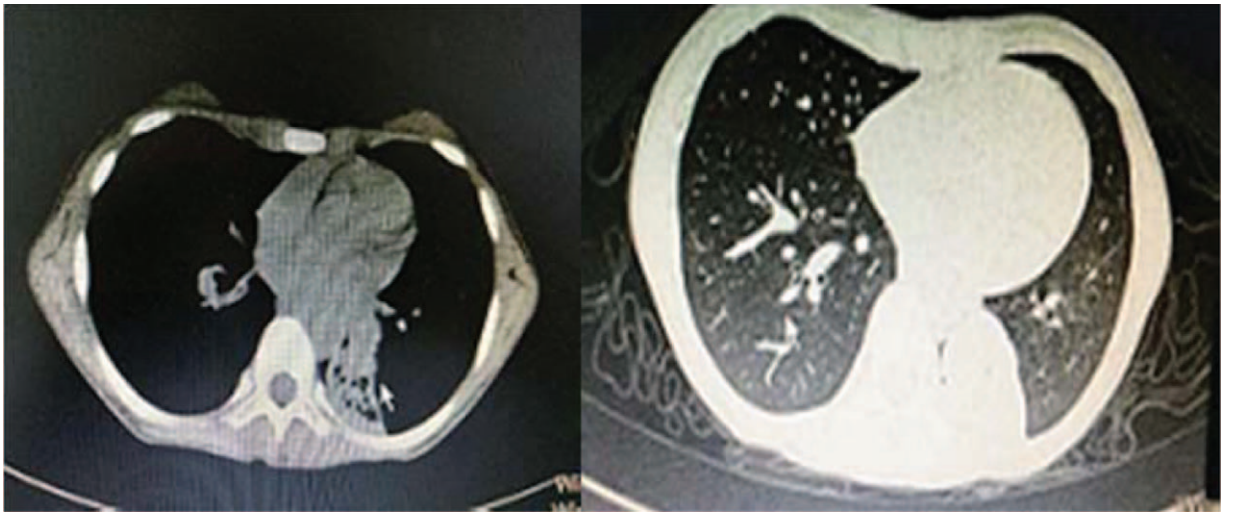

Figure 2. (A) Wedge-shaped patchy dense shadows in the lower lobe of the left lung, with air bronchogram sign inside the region; inhomogeneous enhancement in the lesion on an enhanced scan, with multiple non-enhanced small cystic shadows; patchy shadows supplied by pulmonary artery from the left lower lobe are visible in the enhanced scan, associated with a slightly thin pulmonary artery, drainage of the left lower vein back to the left atrium, and no narrowing of the pulmonary vein. (B) Strip-like high-density shadows in the double pulmonary apex and lingual segment of the upper lobe of the left lung; unobstructed trachea and main bronchus, and obvious narrowing of the opening in the lower lobe of the left lung; normal size of the double hilus of the lung; slightly left shift of the heart, without abnormalities in size and morphology; and no presence of lymph node enlargement in the mediastinum. No abnormalities are observed in the pleura, ribs, or soft tissues of the chest wall.

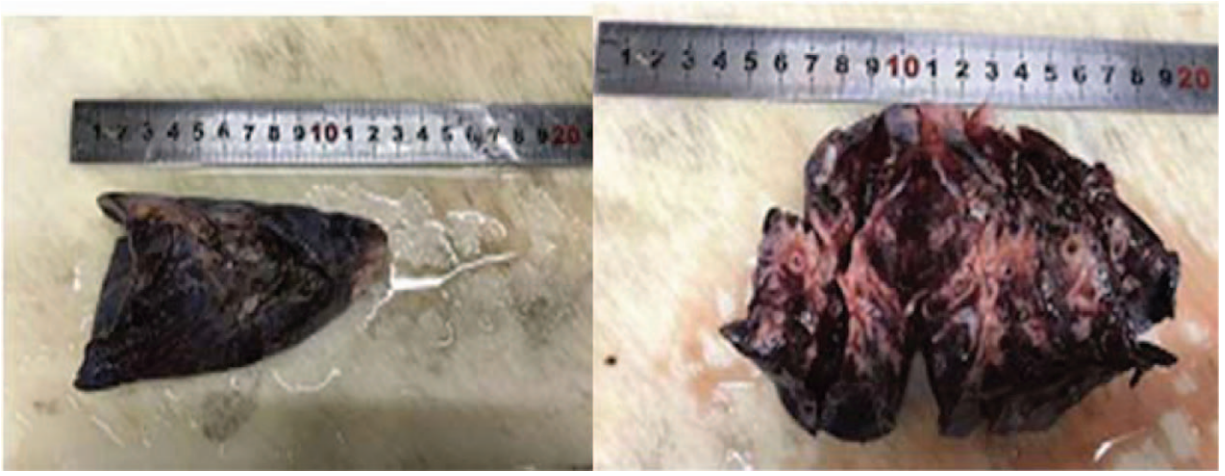

Figure 4. (A) The size of the left lung lobe is approximately 11.5 cm × 10 cm × 5 cm, and the surface is uneven, showing an abnormal grayish black color. (B) A section of the lung lobe is grayish brown, packed with some capsule cavities of different sizes, and the maximum diameter is approximately 2.0 cm.

disappeared after surgery. At a follow-up visit in January 2018, we performed a chest x-ray examination, and the results were normal (Fig. 6).

3. Discussion

Congenital pulmonary dysplasia is caused by the defective budding of the tracheobronchial tree developing from the primitive foregut, and manifests as pulmonary agenesis, pulmonary aplasia, and pulmonary hypoplasia. Pulmonary agenesis refers to the lack of unilateral or bilateral lung tissues, including bronchia, vessels, and pulmonary parenchyma. Pulmonary aplasia is described as presence of only the cecum of the bronchia without nutrient vessels and pulmonary parenchyma.[3] Pulmonary hypoplasia is defined as the condition in which the bronchia, vessels, and pulmonary parenchyma exist, but are fewer in number and smaller than normal. It is usually diagnosed during childhood, and can be accompanied by arteriovenous malformations. Pulmonary agenesis and pulmonary aplasia are serious conditions and the prognosis is poor. However, some patients with pulmonary hypoplasia have no significant clinical symptoms, while others have only recurrent airway infection and damaged exercise performance.[4] In the case described in this report, the patient who did not experience any physical discomfort for >10 years belongs to the third category and her principal manifestation was hemoptysis.

Generally speaking, the common etiologies of hemoptysis in childhood are pneumonia, bronchitis, pulmonary tuberculosis, and idiopathic pulmonary hemosiderosis (IPH), while connective tissue diseases, pulmonary vascular manifestations, pulmonary arteriovenous fistulae, and bronchiectasis are rare.[5] Hemoptysis is considered massive if blood loss is >200 mL per day in children, and 500 mL per day in adults. Hemoptysis in case of respiratory tract infection, IPH, and bronchiectasis is often mild to moderate, while pulmonary vascular manifestations and pulmonary arteriovenous fistulae can cause moderate to massive hemoptysis.[6–8] CPD is not mentioned in most of the reports discussing hemoptysis. In our patient, the local lesion of pulmonary hypoplasia showed pulmonary vascular manifestations and bronchiectasis, and this could have been the direct cause of the hemoptysis, while the recent MP infection which caused the inflammation of local lesion would be the inducement.

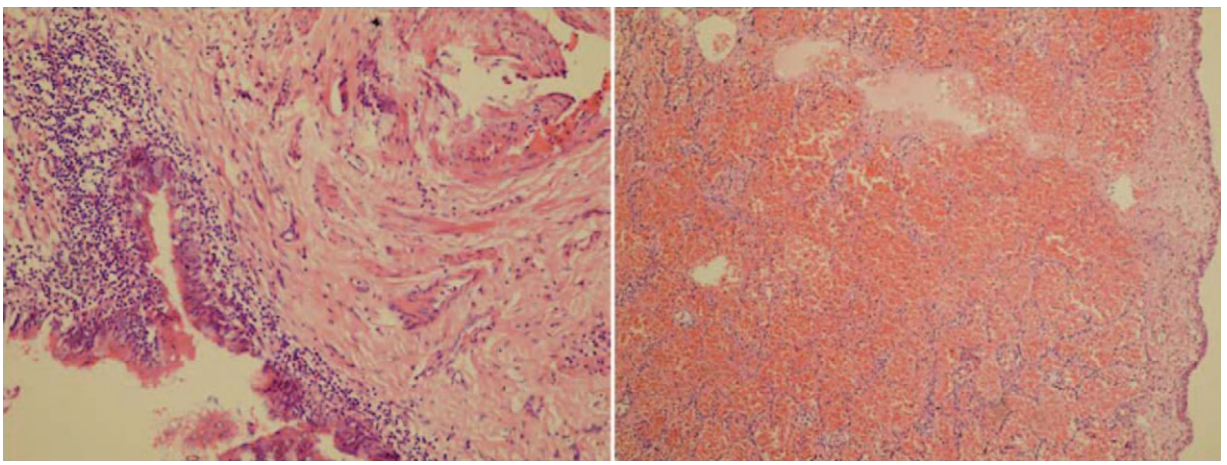

Figure 5. (A) Gross pathological examination of the lung tissue showing bronchiectasis, chronic inflammatory mucosal membrane, and fibrosis of the perivascular interstitium. (B) The interstitium of the alveoli has significant hyperemia, the septum is thickened, and pulmonary bullae are observed.

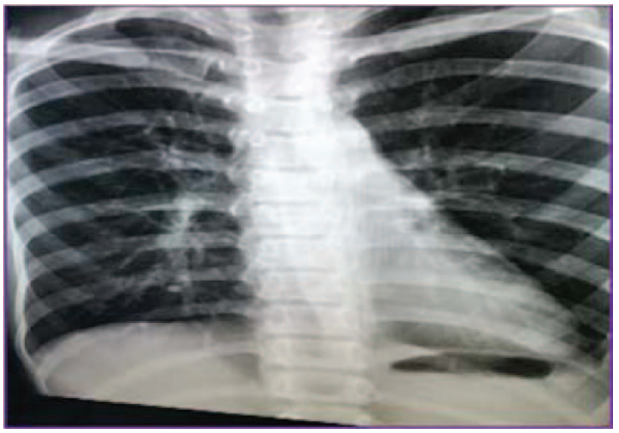

Figure 6. A chest x-ray image showing normal lung at the follow up visit.

Our case shows that when a patient with massive hemoptysis, we should not only consider infections, CPD should be considered even though little clinical evidence is available to indicate pulmonary involvement at the beginning of the disease. Contrast-enhanced CT and bronchoscopy facilitate accurate diagnosis and surgical resection is an appropriate treatment for this condition. Misdiagnosis or delayed diagnosis of pulmonary malformations could result in unnecessary treatments and hospitalization.

CPD is a rare but life-threatening disorder, which plays an important role in various pediatric respiratory diseases. It is characterized by symptoms such as recurrent airway infection, damaged exercise performance, and hemoptysis. Pulmonary contrast-enhanced CT and bronchoscopy are recommended for accurate and timely diagnosis and surgical resection for treatment.

Author contributions

Conceptualization: Lei Zhang.
Data curation: Lei Zhang, Li Bin Zhang.
Formal analysis: Tao Ai.
Funding acquisition: Tao Ai.
Resources: Ying Zhang.
Visualization: Li Wang.
Writing – original draft: Lei Zhang.

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