Clinical Case Report

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Isolated unilateral pulmonary vein atresia with hemoptysis in a child

A case report and literature review

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Abstract

Rationale: Hemoptysis is an uncommon chief complaint but a distressing symptom in pediatric patients. Due to the recurrence and mortality in minor patients, an accurate diagnosis of the underlying cause is quite essential for treatment. The etiologies causing hemoptysis in children are similar to that in adults. Isolated unilateral pulmonary vein atresia (PVA), as an unusual cardiovascular anomaly, has rarely been reported to be an etiology of hemoptysis in children.

Patient concerns: A 2-year and 11-month-old boy was admitted into our hospital with a complaint of recurrent hemoptysis for 2 months and the symptom became more aggravated in recent 4 days before admission. Physical examination was only remarkable for slightly diminished breath sounds over the left lung field, pale face, and colorless lip. Series of targeted laboratory evaluation were negative expect for anemia. Due to the identification of asymmetrical transparency of bilateral lung, slight emphysema of right lung, less volume of left lung with ground-glass opacity and reticular opacity, and ipsilateral mediastinal shift on chest CT, and varices of submucosal vessels in the left bronchial tree on the fiber-optic bronchoscope.

Diagnoses: It more likely indicated a congenital cardiovascular disease. The diagnosis of left isolated unilateral PVA was ultimately confirmed through chest CT angiography (CTA) with three-dimensional (3D) reconstruction.

Interventions: Since the boy did not complain with hemoptysis after admission, respiratory tract infections seldom occurred and no pulmonary hypertension was detected, a conservative approach was chosen with periodic clinical follow-up after discussing with the cardiac surgeons and in accordance to his parents' own wishes.

Outcomes: Fortunately, he was doing well after 3 months of clinical observation.

Lessons: We firstly reported a rare case of hemoptysis in children secondary to isolated unilateral PVA with no associated congenital heart disease in Chinese population. It is significant to improve the recognition and prompt diagnosis of this rare condition for pediatric clinicians, and widen the etiology spectrum of hemoptysis in children. The diagnosis of unilateral PVA should be considered for a patient with recurrent hemoptysis and imaging findings that indicate hypoplastic lung, ipsilateral mediastinal shift, and smooth margins of left atrium without evidence of rudimentary pulmonary veins.

Abbreviations: 3D = three-dimensional, CRP = C reaction protein, CT = computed tomography, CTA = computed tomography angiography, ESR = erythrocyte sedimentation rate, GBM = glomerular basement membrane, HGB = hemoglobin, PCT = procalcitonin, PVA = pulmonary vein atresia.

Keywords: children, hemoptysis, isolated unilateral pulmonary vein atresia

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1. Introduction

Hemoptysis, defined as expectoration of blood or blood tinged-sputum due to bleeding from the respiratory tract, is an uncommon chief complaint but a distressing symptom in children.[1,2] Due to the recurrence and mortality in a minority of patients, an accurate diagnosis of the underlying cause is extremely important and essential for treatment.[3] The etiologies resulting in hemoptysis in children are similar to that in adults, including infections, cardiovascular diseases, foreign body aspiration, cystic fibrosis, respiratory neoplasm, bronchiectasis, the tracheostomy-related and vasculitis syndrome as well as other idiopathic conditions such as pulmonary hemosiderosis.[2–4] Cardiovascular diseases, like left heart dysfunction, aorta pulmonary collateral arteries, pulmonary arteriovenous fistula, although relatively uncommon, with a constituent ratio of 5% to 10% in different case series, are generally recognized as the important causes for pediatric hemoptysis.[3,4] However, pulmonary vein atresia (PVA), as an unusual cardiovascular anomaly, has rarely been reported to be an etiology of hemoptysis in children.

PVA is a rare congenital malformation, which occurs due to the failure of incorporation of the common pulmonary vein into the left atrium. On the basis of the extent of involvement and the stage in which the normal development of pulmonary venous drainage is affected, it is divided into 3 categories: common, unilateral, and individual PVA.[5] Unilateral PVA is a more rare condition, with less than half of patients not accompanying with congenital cardiac anomalies.[6] Recurrent respiratory tract infections and dyspnea on exertion are the common clinical manifestations. Hemoptysis may be the presenting complaint occasionally. Rupture of the dilated bronchial veins is believed to be the mechanism of hemoptysis in these cases.[7] Due to the morbidity and mortality of this disorder, early and accurate diagnosis is essential and crucial to rule out or avoid potential life-threatening complications of pulmonary hypertension and hemoptysis.[6] In the present study, we firstly reported a rare case of hemoptysis in children secondary to isolated unilateral PVA with no associated congenital heart disease in Chinese population and have given a literature review, aiming to improve the awareness and prompt diagnosis of this rare condition for pediatric clinicians and to widen the etiology spectrum of hemoptysis in children.

2. Ethics statements

Informed consent was obtained from the patient's parents for publication of this case report and accompanying images. The study was approved by the University Ethics Committee on Human Subjects at Sichuan University.

3. Case Report

A 2-year and 11-month-old boy was admitted to our emergency department with a complaint of recurrent hemoptysis for 2 months, described as blood tinged sputum. The symptom became more aggravated (coughing up about 3–5 mL of bright red blood several times per day) in recent 4 days before admission. No other symptoms, such as fever, cough, wheeze, night sweat, weight loss, rash, arthralgia, were described during the disease course. Any history of foreign body aspiration, recurrent respiratory tract infections, direct contact with tuberculosis patients and hereditary hemorrhagic diseases prior to illness onset was denied. On arrival, he was conscious, afebrile and had no tachypnea, no hypotension and no hypoxia. Sinus tachycardia with a heart rate of 135 times per min was noted. Physical examination was only remarkable for slightly diminished breath sounds over the left lung field, pale face, and colorless lip. Severe anemia was detected in blood routine test (hemoglobin (HGB) 53 g/L), while normal platelet and coagulation panel ruled out the possibility of systemic hemorrhage diseases. After blood transfusion, the child was admitted into our cardiovascular department for further evaluation and treatment.

Series of targeted laboratory examinations and imaging were performed to help find out the original disease causing hemoptysis. The complete blood count was almost normal except for the decreased HGB (96 g/L), which was improved by transfusion without any adverse effect. C reaction protein (CRP), erythrocyte sedimentation rate (ESR), procalcitonin (PCT), liver function, renal function, blood electrolyte, autoantibody, anti-neutrophil cytoplasmic antibodies, anticardiolipin antibody, and anti-GBM were unremarkable. Additionally, sputum culture, mycoplasma IgM, T-spot and PPD skin test were also negative.

Notably, the images on the chest CT scan were suggestive of the asymmetrical transparency of bilateral lung, the slight emphysema of right lung, and the less volume of left lung with ground-glass opacity and reticular opacity, and ipsilateral mediastinal shift, but without other abnormal features such as pulmonary tubercle, cavity with shaggy walls and air-fluid level, pleural effusion, and lymphadenopathy (shown in Fig. 1). In addition, to further ascertain the unusual performances observed on the chest CT scan and detect local sites of bleeding, the fiber-optic bronchoscope was conducted. Encouragingly, mucosal hyperemia, increased secretions, derangement and varices of submucosal vessels in the left bronchial tree were revealed, whereas only mucosal hyperemia and edema were seen on the right side. The varices in the left side had a particular streaked appearance and bled on contact with the bronchoscope (shown in Fig. 2). No foreign body was found and the bronchoalveolar lavage was normal.

In light of the findings of chest CT scan and bronchoscope, the diagnosis of congenital cardiovascular disease was highly suspected. To confirm our hypothesis, the echocardiography was applied, but no intracardiac anatomic abnormality was displayed. Additionally, the chest CT angiography (CTA) with

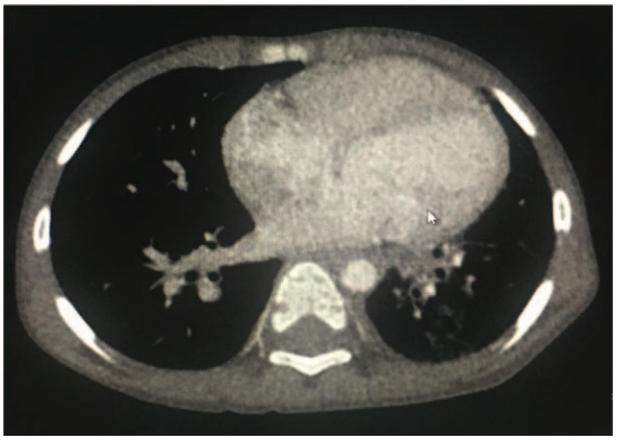

Figure 1. The images on the chest CT scan were suggestive of the asymmetrical transparency of bilateral lung, the slight emphysema of right lung, and the less volume of left lung with ground-glass opacity and reticular opacity, and ipsilateral mediastinal shift, but without other abnormal manifestations such as pulmonary tubercle, cavity with shaggy walls and air-fluid level, pleural effusion, and lymphadenopathy. CT = computed tomography.

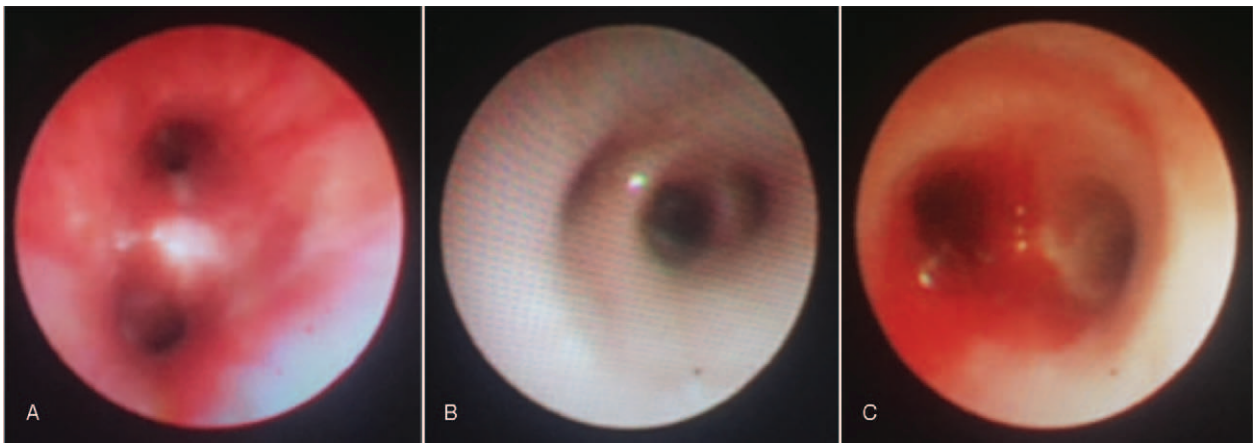

Figure 2. Fiber-optic bronchoscopy revealed mucosal hyperemia, increased secretions, derangement, and varices of submucosal vessels in the left bronchial tree (A), while only mucosal hyperemia and edema were seen on the right side (B). The varices in the left side had a particular streaked appearance and bled on contact with the bronchoscope (C).

three-dimensional (3D) reconstruction was conducted. Encouragingly, the left lung parenchyma was found to be slightly hypoplastic, and it manifested with diffused lung mesenchyme changes; the left main pulmonary artery and ipsilateral vessels were smaller in caliber in comparison with the right, and bilateral bronchial artery was circuitous and thickening, particularly the left bronchial artery more apparent. Besides, there were no left pulmonary veins draining into the left atrium visualized on the images. The margin of the left atrium in the expected location of the left pulmonary vein was entirely smooth, without evidence of vascular structures connected to it (shown in Fig. 3). Based on these findings, the diagnosis of left isolated unilateral PVA with no associated congenital heart disease was ultimately confirmed. Since the boy did not complain with hemoptysis after admission, respiratory tract infections now seldom occurred and no pulmonary hypertension was detected, a conservative approach was chosen with periodic clinical follow-up after discussing with the cardiac surgeons and in accordance to his parents' own wishes. Fortunately, he was doing well after 3 months of clinical observation.

4. Discussion

Hemoptysis in children is not a common chief complaint, but if occurs, it poses a frightening diagnosis for pediatricians and the parents as it may initially remain unnoticed because children tend to swallow their sputum and are difficult to provide a clear history. Once the presence of hemoptysis has been identified, one needs to identify the source of bleeding and primary underlying cause. The main causative disease in children is different according to the age, race and region distribution, but infection is still the most underlying causes for a majority of pediatric hemoptysis.[3] However, in the present study, we reported one extremely infrequent etiology, unilateral PVA. To our knowledge, including our case, <50 patients have been reported in the medical literature (shown in the supplementary material, http://links.lww.com/MD/C406). We could note that most patients were associated with congenital cardiac defects and manifested with recurrent episodes of respiratory infections and/or dyspnea on exertion or exercise intolerance. Isolated unilateral PVA with hemoptysis only occurred in 6 cases (as shown in Table 1). As we know, this childhood patient was the first case presented with recurrent hemoptysis secondary to isolated unilateral PVA with no associated cardiac anomalies in Chinese population.

Hemoptysis secondary to unilateral PVA was considered to result from the high pressure and consequent rupture of dilated bronchial veins. The association of bronchial varicosities with obstruction to the pulmonary venous return in the setting of unilateral PVA has rarely been described.[9] These can be

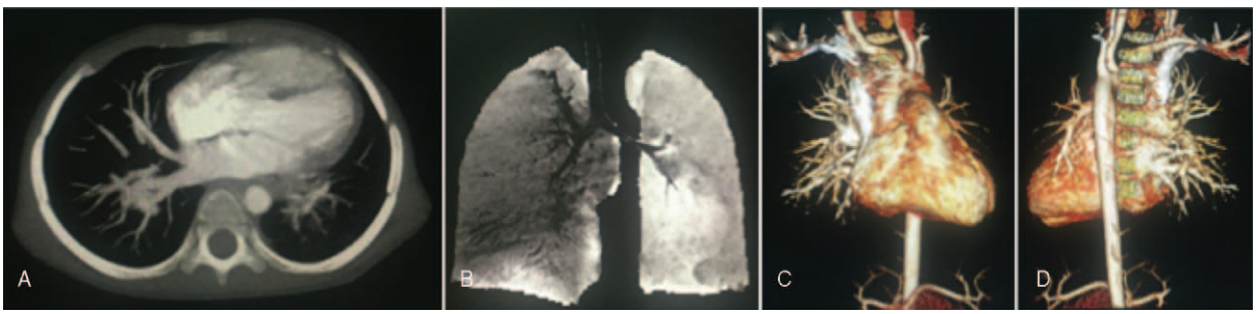

Figure 3. Chest CT scan showed a smaller caliber of the left main pulmonary artery and ipsilateral vessels compared with the right (A); chest CT with 3D reconstruction revealed the left lung parenchyma with slight hypoplasia like mesenchyme (B); chest CTA with 3D reconstruction showed that the left main pulmonary artery and ipsilateral vessels were smaller in caliber in comparison with the right, and bilateral bronchial artery was circuitous and thickening, particularly the left bronchial artery more apparent. Besides, there were no left pulmonary veins draining into the left atrium visualized on the images. The margin of the left atrium in the expected location of the left pulmonary vein was entirely smooth, without evidence of vascular structures connected to it (C, D). CT = computed tomography.

Table 1

Cases of isolated unilateral pulmonary vein atresia complaining with hemoptysis in children.

References	Age years	Sex	Side	Main Complaints	Associated cardiac lesions	Diagnostic ways	Therapy	Follow-up
Swischuck[8]								
Case 1	2.5	M	L	2	—	PA and CC	S	I
Case 2	2	F	R	1/2	—	PA and CC	S	—
Case 3	2.5	M	L	1/2	—	PA	—	—
Dixit[9]								
Case 1	7	F	R	1/2	—	Chest CT and CTA	—	—
Case 2	3	F	R	2	—	Bronchoscopy, chest CT and CTA	—	—
Cindy S[10]	12	F	R	1/2	—	Bronchoscopy, ECHO and CC	S	I
Current study	2.9	M	L	2	—	Bronchoscopy, CTA with 3D reconstruction	C	I

Main complaints: recurrent pneumonia or frequent respiratory tract infections; recurrent hemoptysis

Therapy: S: surgery; C: conservative management; Follow-up: I: Symptoms improved or completely well.

3D = three-dimensional, CC: cardiac catheterization, CT = computed tomography, CTA = computed tomography angiography, ECHO = echocardiogram F = female, L = left, M = male, PA = pulmonary arteriography, R = right.

explained by increased pulmonary venous pressure leading to communications between the pulmonary and bronchial veins. As a matter of fact, isolated bronchial varices should also be considered in the differential diagnosis of this anomaly.

Overall, the prognosis of unilateral PVA was still guarded, with an approximately 50% mortality rates in untreated patients,[11] so early and accurate diagnosis is quite pivotal and essential before pulmonary hypertension or massive pulmonary hemorrhage occurred. Being aware of the possibility of unilateral PVA and avoidance of misdiagnosis of idiopathic pulmonary hemosiderosis when confronted with childhood patients presenting with hemoptysis, is of great importance. Significantly, some specific features on the imaging may help the pediatricians to identify affected individuals. Firstly, the morphologic characteristics by chest CT and CTA should have the increased attenuation of affected lung parenchyma with hypoplasia, small hemithorax and ipsilateral mediastinal shift, thickening interlobular septa, smooth margins of left atrium without evidence of rudimentary pulmonary veins, and small ipsilateral pulmonary artery.[7,12] Secondly, bronchial varix may serve as a hallmark clinical feature of unilateral PVA for the imaging of bronchoscopy.[7,13] Additionally, it may also be a very important clue in the diagnosis of unilateral PVA that pulmonary angiography may include preferential flow to the uninvolved lung and the distension of the contralateral pulmonary veins that have normal transit time, while the absence or rather the reversal of pulmonary blood flow in the affected lung.[14] Lastly, the cardiac catheterization may reveal increased pulmonary wedge pressures as well as higher oxygen saturations in the venous system, and differential oxygen saturation in both pulmonary artery that the affected side has an even higher readings compared with the unaffected side.[6]

Owing to the extremely lower prevalence of this disorder and limited number of patients, data with respect to long term follow-up is very limited. Based on the current evidence, basically 2 therapeutic options are available. The first one is conservative approach. As in our case, if the affected patient suffer from no frequent respiratory infections, has no significant symptoms and do not develop pulmonary hypertension, this approach might be chosen. The other considerable option is pneumonectomy suitable for patients with severe symptoms and signs to obviate the life-threatening complications such as recurrent severe respiratory tract infections, pulmonary hypertension and/or recurrent hemoptysis.[12,15] Available evidence indicates that the outcome of pulmonary resection is satisfactory. Nevertheless, pneumonectomy is burdened with increased morbidity in small growing children, like scoliosis, kyphosis, or unilateral hypoplastic chest wall, all being more pronounced when the child is younger. Therefore, this operation should be delayed as much as possible to avoid these complications.[16] Since the boy in our study did not complain with hemoptysis after admission, respiratory tract infections now seldom occurred and no pulmonary hypertension was detected, a conservative approach was chosen with periodic clinical follow-up after discussing with the cardiac surgeons and in accordance to his parents' own wishes. Fortunately, he was doing well after 3 months of clinical observation. A close follow-up will be further carried out to monitor the occurrence of complications.

In conclusion, we firstly reported a rare case of hemoptysis in children secondary to isolated unilateral PVA with no associated congenital heart disease in Chinese population, and had a literature review. It is of great significance to improve the recognition and prompt diagnosis of this rare condition for pediatric clinicians, and widen the etiology spectrum of hemoptysis in children. The diagnosis should be considered for one patient with recurrent hemoptysis and imaging findings that indicate hypoplastic lung, ipsilateral mediastinal shift, and smooth margins of left atrium without evidence of rudimentary pulmonary veins. Bronchoscopic findings of unilateral submucosal bronchial varices should be clearly evocative of unilateral obstruction to the pulmonary venous return. In spite of the controversial therapeutic approach, we should bear in mind one point that surgery could be required in severely symptomatic patient with long-term or massive hemoptysis, pulmonary hypertension, and/or recurrent severe lung infections. If mildly symptomatic or even asymptomatic, the conservative alternative with a close follow-up should be highlighted against a possibly drastic and complicated surgical treatment, especially in the young children. However, due to the limited number of patients reported in the literature and the short-term follow-up results as our case, more patients with different treatment strategy and long-term follow-up outcomes need to be further accumulated, and thereby to develop a applicable guideline regarding the management of unilateral PVA and to improve its prognosis.

Author contributions

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Supervision: Yifei Li, Xiaoliang Liu.

Writing – original draft: Changqing Tang.
Writing – review & editing: Changqing Tang, Hongyu Duan, Chuan Wang.

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