

Unilateral pulmonary venous atresia: A rare cause of recurrent hemoptysis

Shagun Jyotin Shah, Chandrahas T Deshmukh, Jane JE David, Moushmi M Hosalkar

Department of Pediatrics, Seth G. S. Medical College and KEM Hospital, Mumbai, Maharashtra, India

ABSTRACT

Hemoptysis is a rare but distressing complaint in children. Pulmonary venous atresia (PVA) is a rare cause of recurrent hemoptysis in children. We report a 3-year-old boy with recurrent hemoptysis for 2 years. The child had tachycardia, tachypnea, tender hepatomegaly, and left-sided decreased air entry with crepitations. Coagulation and Koch's workup was negative. X-ray of the chest showed a small left hemithorax. Computed tomography with angiography showed left unilateral PVA with pulmonary artery hypoplasia and dysplastic left lung. The child has been posted for left pneumonectomy. Although uncommon, PVA should be diagnosed early to prevent life-threatening complications such as hemoptysis and pulmonary hypertension. The patient can be managed conservatively or surgically depending on the severity. Early diagnosis and intervention helps in reducing morbidity and mortality.

KEY WORDS: Dysplastic lung, hemoptysis, pneumonectomy, pulmonary venous atresia

Address for correspondence: Dr. Shagun Jyotin Shah, 46, Kailash Nagar, 658 Tardeo Road, Mumbai - 400 007, Maharashtra, India.
E-mail: shagunshah27@gmail.com

Submitted: 12-Aug-2019 **Accepted:** 26-Oct-2019 **Published:** 27-Feb-2020

INTRODUCTION

Hemoptysis is a rare but distressing complaint in children.^[1] Common causes of hemoptysis in children are bronchiectasis, tuberculosis, foreign body, and coagulopathies.^[1] Pulmonary venous atresia (PVA) is a rare cause of recurrent hemoptysis in children. Due to morbidity and mortality of this disorder, early and accurate diagnosis is essential to avoid complications such as pulmonary hypertension and hemoptysis. We report a case of a unilateral left PVA with dysplastic lung. There are very few cases reported till date.

CASE REPORT

A 3-year-old boy, born of a non-consanguineous marriage, resident of Jaunpur presented with recurrent episodes of

cough and cold with hemoptysis for 2 years. Each episode was associated with breathlessness lasting for 1 week, some requiring hospitalizations. There was no history of fever, Kochs or Koch's contact, edema, or palpitations. The child was given nebulization in each admission but showed no response. Antenatal scans were normal. He was a full-term normal delivery with a birth weight of 2.4 kg. The child did not cry immediately after birth with no other neonatal problems and had normal development. On admission, the child had tachycardia, tachypnea, pallor, and Grade 1 clubbing. Respiratory system examination revealed decreased air entry with crepitations on the left side. He had cardiomegaly with a grade 3/6 ejection systolic murmur in the left second intercostal space with a normal second heart and a tender hepatomegaly of 4 cm. Hemoglobin was 7 g/dl with normal leukocyte,

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How to cite this article: Shah SJ, Deshmukh CT, David JJ, Hosalkar MM. Unilateral pulmonary venous atresia: A rare cause of recurrent hemoptysis. Lung India 2020;37:164-6.

Access this article online	
Quick Response Code: 	Website: www.lungindia.com
	DOI: 10.4103/lungindia.lungindia_369_19

platelet count, and coagulation. Tuberculosis workup was negative.

Chest radiograph revealed small left hemithorax with cardiothoracic ratio of 0.60. Transthoracic Doppler echocardiographic examination revealed an underfilled left atrium with dilated right atrium. The left pulmonary vein could not be visualized. The interatrial and interventricular septum was normal. The pulmonary artery was dilated slightly without the evidence of pulmonary hypertension. A computed tomography (CT) of the chest with pulmonary angiography showed that the left and right lower pulmonary veins were seen for a distance of 4 mm and 3 mm from the left atrial orifice, respectively. The remaining part of the veins was atretic. The right-sided pulmonary veins were dilated and drained into the left atrium. There was no anomalous venous drainage. The left pulmonary artery was hypoplastic with severe narrowing of lobar branches. The right pulmonary artery was normal. There was also a loss of lung volume on the left side with dysplastic changes. The diagnosis of left PVA was confirmed [Figures 1-3]. Along with blood transfusion, diuretics and digoxin was started, and the patient was posted for left pneumonectomy.

DISCUSSION

During the first 2 months of fetal life, the common primitive pulmonary vein arises from the primitive left atrium which finally gets incorporated into the left atrium with four pulmonary veins arising from it. The right superior pulmonary vein drains the right upper and middle lobe. The left superior pulmonary vein drains the left upper and lingular lobe. The two inferior pulmonary veins drain the lower lobes. Abnormal resorption of the embryonic structures can lead to a change in the number or size of pulmonary veins or abnormal drainage of the veins.^[2]

PVA is one such rare congenital anomaly.^[2] It is due to the failure of incorporation of the common pulmonary vein into the left atrium.^[2] It is mostly congenital but can be secondary to veno-occlusive disease or mediastinitis.^[3] Depending on the degree of involvement, it can be divided into common, unilateral, and individual PVA. Our patient had a unilateral PVA. It may occur in either lung.^[3] They normally present in infancy or within the first 3 years of life, but some patients may become symptomatic in adulthood. It may be associated with other congenital heart diseases or an abnormal pulmonary venous drainage in 50% of the cases.^[2] The ipsilateral pulmonary artery is also affected in these patients as seen in our case due to preferential pulmonary artery perfusion to the contralateral side, with resultant impaired growth of the affected pulmonary artery. This leads to systemic pulmonary artery collaterals that cause dilatation of the bronchial vessels and can lead to bronchial varix formation.



Figure 1: Computed tomography Chest showing cystic lesions with the loss of volume of the lung suggestive of the dysplastic lung (original)

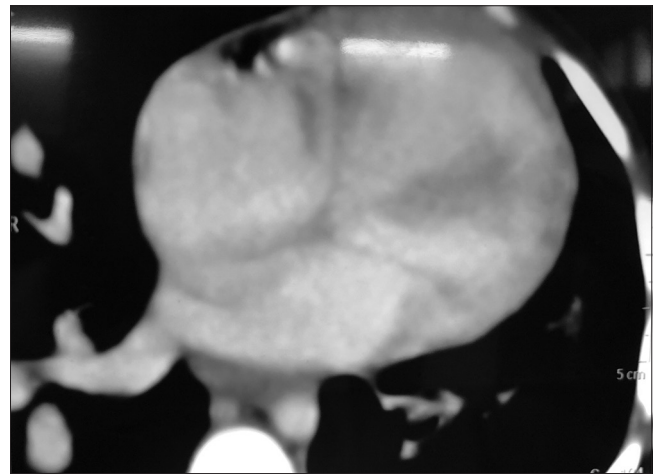


Figure 2: Computed tomography angiography showing left pulmonary vein atresia, normal right pulmonary vein, and smooth left atrium (original)

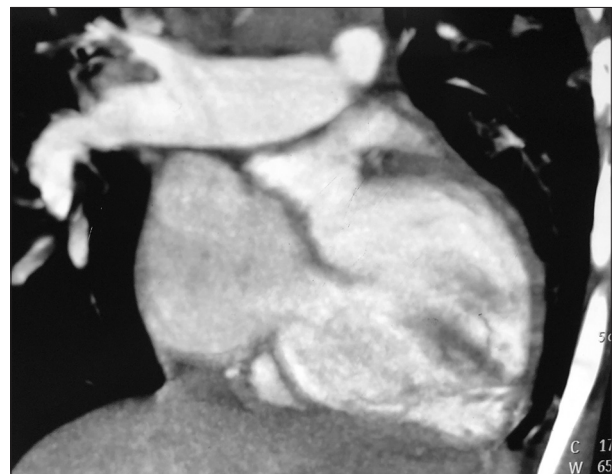


Figure 3: Left pulmonary artery hypoplasia on computed tomography angiography. The right pulmonary artery is normal (original)

Pulmonary artery hypertension is a common association. Our patient did not have any associated cardiac anomaly or pulmonary hypertension. Patients usually present

with recurrent pulmonary infections or hemoptysis. Our case presented with recurrent hemoptysis. Hemoptysis is due to the high pressure and rupture of the dilated bronchial veins.^[4]

CT scan along with angiography aids in diagnosis. It shows increased attenuation of affected lung parenchyma with hypoplasia, small hemithorax, ipsilateral mediastinal shift, thickening interlobular septa, smooth left atrium without evidence of rudimentary pulmonary veins, and small ipsilateral pulmonary artery.^[5,6] Thickening of the interlobular septa is due to the dilatation of pulmonary lymphatics and bronchial veins with bronchial varix serving as the clinical hallmark. Pulmonary angiography may be helpful in demonstrating the absence or reversal of pulmonary blood flow in the affected lung and preferential flow to the uninvolved lung.^[6] 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 arteries. The histologic hallmark in PVA is extensive intimal fibrosis of the pulmonary veins with severe medial hypertrophy. The mortality rate is about 50% in untreated patients.^[7] Therefore, early diagnosis is necessary for the early intervention to prevent life-threatening complications such as massive pulmonary hemorrhage and pulmonary hypertension.

Based on the current evidence, there are two interventions which can be done for these patients.^[8] Surgery may be required in severely symptomatic patients with long-term or massive hemoptysis, pulmonary hypertension, and/or recurrent severe lung infections. The conservative approach is used in asymptomatic or mildly symptomatic patients. Pneumonectomy has been found to remove the focus of recurrent pulmonary infections, decreases the shunting and hence hemoptysis, and decreases the dead space which improves exercise tolerance. In our case, since the child had recurrent hemoptysis, a surgical approach has been planned.

Few cases of pulmonary vein atresia in infants and young children have been reported. Nasrallah *et al.* reported a 16-month boy with unilateral pulmonary atresia treated successfully by a right pneumonectomy.^[9] DeMastes-Crabtree *et al.* reported a case of a 12-year-old girl with PVA who underwent a surgical repair which included repair of the pulmonary vein atresia and a pericardial patch angioplasty of the right pulmonary venous confluence.^[10] Our patient had dysplastic lung changes with left pulmonary vein atresia and pulmonary artery hypoplasia.

CONCLUSION

Hemoptysis is not very common in the pediatric population, with PVA being a rare cause of the same. When preliminary workup for hemoptysis is negative and X-ray is suggestive of the above-mentioned findings, PVA should be suspected. CT or magnetic resonance imaging angiography is found to be useful for a confirmatory diagnosis. Patients can be treated conservatively or surgically depending on the severity of the illness. Antenatal diagnosis of this condition may help in better survival of the patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient's parents have given their consent for the patient images and other clinical information to be reported in the journal. The patient's parents understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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