Case report: Isolated unilateral pulmonary vein atresia diagnosed on 128-slice multidetector CT

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Abstract

Unilateral pulmonary venous atresia is an uncommon entity that is generally believed to be congenital. Most patients present in infancy or childhood with recurrent chest infections or hemoptysis. Pulmonary angiography is usually used for definitive diagnosis. However, the current multislice CT scanners may obviate the need for pulmonary angiography. We report two cases diagnosed using 128-slice CT angiography. On the CT angiography images both these cases demonstrated absent pulmonary veins on the affected side, with a small pulmonary artery and prominent bronchial or other systemic arterial supply.

Key words: Atresia; computed tomography; congenital; Hemoptysis; pulmonary vein

Introduction

Unilateral pulmonary venous atresia is a rare anomaly presenting with recurrent chest infections and hemoptysis.[1,2] This condition must be considered in the differential diagnosis when children present with recurrent unilateral pneumonia, particularly when associated with hemoptysis and reduced lung volume. Multidetector CT can exquisitely demonstrate the abnormality noninvasively.

Case Reports

Case 1

A 7-year-old girl was admitted to the hospital for evaluation of recurrent chest infections since the age of 4 months and recurrent hemoptysis for the past 4 years, with about 1--2 episodes per year. Though there was no definitive evidence of tuberculosis, the child had received two courses of antituberculous treatment on an empirical basis. At admission, the child was afebrile. Her hemoglobin level was 10 g% and the Mantoux was nonreactive. Slight flattening of the right chest was noted.

Chest radiograph revealed a small right hemithorax, with mediastinal shift to the same side and increased opacity throughout the right lung field [Figure 1].

CT scan of the chest and CTA showed a small right hemithorax with interlobular septal thickening and increased attenuation throughout the right lung. No evidence of bronchial obstruction was noted [Figure 2]. CTA revealed a small right main pulmonary artery, complete absence of the pulmonary veins on the right side, and normal pulmonary veins on the left. A confluent soft tissue mass was seen adjacent to the left atrium [Figure 3]. A prominent right bronchial artery supplying the right lung and additional systemic vascular supply from a prominent left inferior phrenic artery were also identified on the angiographic images [Figure 4].

Echocardiography of the patient revealed a small right pulmonary artery. There was no evidence of any congenital cardiac disease.

Case 2

A 3-year-old girl was referred for bronchoscopy with a history of recurrent hemoptysis since the age of 1 year.
Bronchoscopy revealed patent bronchi, with mucosal hyperemia and inflammation in the right bronchi. No other intrabronchial lesion was detected. The child had a hemoglobin level of 12 g% and her general physical examination was unremarkable. Chest examination showed mildly decreased air entry on the right side.

The chest radiograph revealed a small right hemithorax, with evidence of consolidation in the right middle and lower zones [Figure 5].

CT scan of the chest revealed a small right hemithorax with ipsilateral mediastinal shift and multiple areas of consolidation in the lung parenchyma [Figure 6]. CTA depicted a hypoplastic right main pulmonary artery [Figure 7], with nonvisualization of the pulmonary veins [Figure 8]. A prominent bronchial artery supplying the lung was also identified [Figure 9].

Discussion

Unilateral pulmonary atresia, without associated structural abnormalities of the heart, is a rare congenital anomaly. It is thought to result from a failure of incorporation of the common pulmonary vein into the left atrium. It may occur in either lung, and shows no preference for either side. Other associated cardiac defects are found in 50% of the patients. Pulmonary artery hypertension is also a
frequent association. The mortality rate approaches 50% in untreated patients.\(^4\)

Patients usually present with recurrent pulmonary infection or hemoptysis. Our cases presented with recurrent hemoptysis, with one patient also having recurrent chest infections. The chest radiographs in both the cases revealed a small ipsilateral hemithorax, with mediastinal shift to the same side and parenchymal opacities.

The diagnosis was made with certainty on the basis of CT scans of the chest and 128-slice CTA findings. These findings included a small hemithorax, ipsilateral mediastinal shift, and parenchymal opacities in the form of a ground–glass haze; in addition, there was septal thickening in one case.

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**Figure 5:** The chest radiograph shows an area of consolidation (arrow) in the right mid-zone.

**Figure 6 (A, B):** Coronal CT scans (lung windows) show areas of consolidation (arrows) on the right side with a small hemithorax and normal central airways.

**Figure 7:** Axial multiplanar reconstruction CT scan shows the small right pulmonary artery (arrow) and the normal left pulmonary artery (thick arrow).

**Figure 8 (A, B):** Coronal (A) and axial (B) multiplanar reconstruction (MPR) CT scan images show the complete absence of the right pulmonary veins; the left superior and inferior pulmonary veins (arrows) are well seen. The left atrial outline is smooth, and minimal soft tissue (arrowhead) is noted adjacent to it.

**Figure 9:** Volume-rendered aortic angiogram (posterior view) shows a prominent right bronchial artery (arrow). Also note the absence of the right pulmonary veins.
and consolidation in the other case. Septal thickening is likely related to dilatation of the pulmonary lymphatics and bronchial veins, due to the obstructed venous return. Fibrosis of the lung parenchyma can also occur due to a combination of pulmonary venous infarction and chronic pulmonary edema.\[^{5-7}\] In both cases, the CT scans revealed the absence of pulmonary veins, with confluent soft tissue in the mediastinum and a small ipsilateral pulmonary artery. Angiographic studies demonstrated, in addition, bronchial vascular supply in both the cases and systemic parenchymal supply from the inferior phrenic artery in the first case. Although the development of bronchial artery collaterals is known in some cases of pulmonary venous atresia,\[^{4}\] to the best of our knowledge, the demonstration of these and systemic collaterals on CTA has not been emphasized in previous reports.

It has been suggested that the ipsilateral pulmonary artery is diminutive in these cases, likely because of preferential pulmonary artery perfusion to the contralateral side, with resultant impaired growth of the affected pulmonary artery.\[^{8}\] It is possible that this accounts for the systemic-to-pulmonary artery collaterals in these cases. Pulmonary venous obstruction also leads to bronchial varix formation. Rupture of dilated bronchial veins is one of the causes of hemoptysis in these patients.\[^{9,10}\]

Although most patients present in childhood, presentation in adults has also been reported.\[^{2}\] Adult patients with this entity may present a diagnostic dilemma. The confluent mediastinal soft tissue may suggest a mass lesion or fibrosing mediastinitis resulting in ipsilateral pulmonary artery and pulmonary vein obstruction. However, the presence of a small hemithorax without evidence of bronchial obstruction should suggest a congenital anomaly. The confluent soft tissue mass can be attributed to venous collaterals.\[^{2}\]

Noninvasive diagnosis of unilateral pulmonary venous atresia by CTA,\[^{2,9}\] cardiac MRI,\[^{11}\] or bronchoscopy\[^{12}\] has been described in recent publications. CT scan in a manner similar to MRI provides accurate morphological information concerning the heart and extracardiac vascular structures. In both of our patients, conventional angiography was not done as the depiction of both the venous and arterial abnormalities was excellent on the 128-slice CTA.

Here, we emphasize the ability of multislice (128-slice) scan to conclusively establish the diagnosis of pulmonary venous atresia and thus obviate the need for conventional angiography.

**References**