Congenital Absence of the Right Pulmonary Artery: Four Cases Report

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Congenital absence of the right pulmonary artery is an uncommon congenital disorder. We describe the clinical and radiologic presentations of this disease in 4 cases (3 men, 1 woman; mean age: 22.8y/o) in our hospital. Decrease of the right lung volume is the most common radiologic finding. Other characteristic findings include trachea and mediastinum shift, right hemidiaphragm elevation and loss of normal right pulmonary vascularity. CT scan and angiography enable us to confirm the diagnosis precisely. However, plain film and CT scan offer a safer and non-invasive way to make the diagnosis.

Key words: Angiography; Computed tomography; Right pulmonary artery, anomaly

Case 1
A 29-year-old male complained of chronic cough with chest discomfort for more than two years. His symptoms did not improve after medical treatment. Physical examination at the outpatient clinic revealed decreased breathing sounds in the right lung. No wheezing or rales was noted. The plain radiograph of chest showed a small, hypoplastic right lung, mediastinum and trachea shift to right and left lung herniation in the superior mediastinum (Fig 1a). Small right hemithorax with collateral arterial circulation in the subpleural parenchyma and absence of the right main pulmonary artery were demonstrated on enhanced chest CT (Fig 1b,c).

Case 2
A 22-year-old male was admitted because of shortness of breath and repeated attacks of hemoptysis throughout his childhood. He often caught colds with difficult recoveries. Physical examination on admission revealed a flattened right chest wall less expansile than the left side. Breathing sounds were decreased over the right lung field. Chest X-ray showed a slightly contracted right lung with inconspicuous hilum. The heart and trachea were displaced to the right side (Fig. 2a). The left upper lung was hyper-expanded with herniation across the midline.
Pulmonary angiography (Fig. 2b) and chest CT demonstrated absence of the right pulmonary artery with normal left side anatomy. A selective aortogram showed dilated bronchial artery (Fig. 2c) and replacement arteries from the subphrenic artery supplying the lower lung (Fig. 2d).

**Case 3**

A 35-year-old female was first admitted to our hospital for evaluation of an abnormal chest X-ray since childhood. Her only symptom was chronic cough with whitish sputum for six months. Physical examination at the time of admission was unremarkable. Chest X-ray revealed decreased size of the entire right lung and hemithorax compared to the left and slight mediastinum shift to the right side (Fig. 3a). Enhanced chest CT showed complete absence of the right pulmonary artery (Fig. 3b) with collateral arterial circulation in the subpleural parenchyma. In lung window setting, herniation of the left apical lung to the right side was also found.

**Case 4**

A 5-year-old boy suffered from intermittent fever for 2 weeks and cough for 1 day. His mother had had a normal full-term delivery following an uneventful pregnancy. Physical examination revealed rales and decreased breathing sounds over his right lower lung field. Initial laboratory data showed a white blood cell count of $16.01 \times 10^3$/ml with 75% neutrophils. Blood chemistry and urinalysis were within normal limits. Chest X-ray showed a cavitary lesion with air fluid level in the right lower lung field (Fig. 4a). The mediastinum and trachea were shifted to the right side. Chest CT scan demonstrated a cavitary lesion in the right lower lobe of the lung with irregular marginal enhancement after intravenous injection of contrast medium. Absence of the right main pulmonary artery

Figure 1. Case 1. a. Chest plain film shows right lung volume loss with mediastinum and trachea shift to right side. Elevation of right hemidiaphragm was also found. b. Enhanced chest CT reveals absence of right main pulmonary artery. c. In lung window setting, small right hemithorax with collateral artery circulation at subpleural parenchyma and hyperaerated left upper lung herniated into right side are demonstrated.

1b

1c
with hyperexpansion of left lung was also noted (Fig. 4b). Under the impression of lung abscess with absence of right pulmonary artery, the patient received antibiotic therapy and his symptoms resolved after 2 days.

**DISCUSSION**

Although various theories have been proposed to explain the absence of pulmonary arteries, it is widely accepted that disruption of normal embryonic development of the primitive sixth aortic arch results in an anomaly in the pulmonary arteries [1,2,6]. Abnormalities in the rotation and migration of the primitive arch may result in agenesis of one pulmonary artery [6]. Absence of the left pulmonary artery tends to be associated with an intracardiac defect, particularly the Tetralogy of Fallot [7], while absence of the right pulmonary artery is usually not associated with any other anomaly of the heart or great vessels [6,8]. Anderson et al [6] reported that the aortic arch is usually on the side opposite to the absent pulmonary artery. The normal heart usually has a left-sided aortic arch, so the absence of the right pulmonary artery is much more common than the absence of the left pulmonary artery.

The diagnosis of congenital absence of right pulmonary artery is made on the basis of clinical and

![Figure 2. Case 2. a. Chest plain film shows decreased right lung volume, left lung herniated into right side and mediastinum shifted to right side. b. Pulmonary angiography shows absence of right pulmonary artery. c. Dilated bronchial artery supplying the right upper lung from aorta is seen. d. Replacement arteries from subphrenic artery supplying the right lower lung is demonstrated.](image)
radiographic findings. Clinically, patients may be asymptomatic or may suffer from recurrent pulmonary infection, dyspnea or hemoptysis [2,3,4]. Chronic cough is the most common complaint among these patients, and hemoptysis is another common symptom. High pressure of the replacement supplying arteries from the aorta result in ruptured venules in the affected lung. However, according to Bryne et al [4], most patients with congenital absence of right pulmonary artery are asymptomatic. Symptoms are only present in less than 10 percent of cases. Radiologically, absence of the right pulmonary artery is characterized by ipsilateral small lung volume with tracheal deviation, hemidiaphragm elevation, hyper-aerated left lung and right side narrowed intercostal spaces [1,5,7]. In our cases, the typical imaging findings of congenital absence of the right pulmonary artery were well demonstrated. In particular, CT enabled us to diagnose this disease by demonstration of a small right hemithorax with collateral arterial cir-

Figure 3. Case 3. a. Chest plain film reveals similar radiographic appearance as that of case 2. b. Enhanced chest CT reveals absence of right main pulmonary artery.

Figure 4. Case 4. a. Chest plain film reveals a cavitary lesion (arrows) with air-fluid level in the right lower lung field. The mediastinum and trachea are shifted to the right side. b. Chest CT shows similar radiographic appearance as that of case 3.
culation in the subpleural parenchyma and absence of the right main pulmonary artery [9]. Furthermore, angiography precisely demonstrated the replacement arteries from the ascending or descending aorta supplying the right lung.

The differential diagnosis of congenital absence of right pulmonary artery include parenchymal diseases such as pulmonary venous atresia, Swyer-James syndrome (unilateral hyperlucent lung syndrome) and vascular diseases including chronic thromboembolism of one pulmonary artery. Pulmonary venous atresia is an unusual congenital anomaly [10,11]. Embryologically, it results from improper incorporation of the common pulmonary vein into the left atrium. CT and angiography can demonstrate pulmonary hypoplasia of the affected lung with the small pulmonary artery and pulmonary venous atresia. Swyer-James syndrome results from chronic expiratory obstruction with decreased air outflow and distal air trapping. It generally develops after lower respiratory tract infection during early childhood [12]. The differential diagnosis from absence of the right pulmonary artery depends on the inspiratory and expiratory chest films. Swyer-James syndrome is characterized by the mediastinum shifted to the affected side on inspiration and away on expiration [13]. Chronic pulmonary thromboembolism is relatively uncommon. A hypolucnt affected lung with diminished number and size of vessels is demonstrated. CT is helpful to confirm the presence of mural thrombus in the main pulmonary artery [14]. The CT finding of absence of right main pulmonary artery with intact pulmonary trunk and left pulmonary artery is a highly discriminating feature that helps to confirm the diagnosis.

In summary, radiographic features of congenital absence of right pulmonary artery are characteristic and diagnostic. CT scan and angiography enable us to confirm the diagnosis precisely. However, plain film and CT scan offer a safer and non-invasive way to make this diagnosis.

REFERENCES

先天性右肺主動脈發育不全：四病例報告

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先天性右肺主動脈發育不全是一種罕見的先天性疾病。我們在此報告四例患有先天性右肺主動脈發育不全的病例，並回顧相關文獻。在影像學方面最主要的表徵為右側肺容積減少。其他的表徵則包含總氣管和縱隔腔移位，右側橫隔膜升高，右側正常的肺血管陰影減少。電腦斷層掃描和血管攝影能使我們精確地診斷此疾病。然而，胸部X光攝影和電腦斷層掃描便足以提供既安全且非侵入性的方式來做正確的診斷。

關鍵詞：血管攝影；電腦斷層掃描；先天性右肺主動脈發育不全