Unilateral Absence of a Pulmonary Artery

Dyspnea and one episode of blood-tinged sputum the night before. The clinical course of this patient had been complicated by slight impaired exercise tolerance since childhood. Physical examination revealed no abnormalities.

Angiographic findings were consistent with unilateral absence of the right pulmonary artery (UPAA) and underdevelopment of the right lung.

Discussion:

Congenital absence of a pulmonary artery is a rare anomaly. It is frequently associated with other cardiovascular anomalies, such as tetralogy of Fallot, right aortic arch, septal defects and patent ductus arteriosus. There is no predilection for the right or left side, although the condition is somewhat more common on the right. Although the side of the agenesis is not predictive for the presence of associated cardiac pathology, right-sided agenesis is less frequently associated with cardiac abnormalities. Physical examination is mostly unremarkable with exception for decreased breath sounds on the involved side. An isolated UPAA shows a characteristic pattern of chest roentgenography findings, including cardiac and mediastinal displacement, absence of the pulmonary artery shadow, smaller hemithorax, elevation of the diaphragm and paucity of the vascular markings, all on the involved side. On the opposite side, there may be hyperinflation and herniation of the lung across the midline. Ventilation-perfusion scan is an important diagnostic tool but DSA is definite in establishing the diagnosis of UPAA. The affected lung is supplied by systemic collaterals (hypertrophic bronchial arteries, intercostal and other transpleural collaterals), but pulmonary venous return is normal in most cases. The clinical course is mostly benign and when symptoms occur, they are often related to recurrent respiratory tract infections with bronchiectasis. Since symptoms are subtle and physical examination mostly normal, clinical diagnosis of UPAA of the adult is difficult. It should always be considered in the differential diagnosis of a unilateral hyperlucent lung.

Differential Diagnosis List: Congenital absence of a pulmonary artery

Final Diagnosis: Congenital absence of a pulmonary artery
References:


**Description:** Chest radiograph, posterior-anterior view shows underdevelopment of the right hemithorax and lung, with deviation of the mediastinal structures towards the right side. There is an increased vascularity of the left lung. **Origin:**
Description: Contrast-enhanced CT scan of the thorax visualizes a sharp delineation and normal tapering of the left pulmonary artery. The right pulmonary artery is not visible. The right internal thoracic artery is enlarged (arrow). Origin:
Description: Arteriography of the pulmonary arteries demonstrates opacification of an enlarged left pulmonary artery, with normal tapering towards the periphery. There is no obvious perfusion of the right lung. Origin:
Description: Selective arteriography of the right subclavian artery demonstrates the enlarged right internal thoracic artery with filling of small mediastinal and bronchial arteries supplying the right pulmonary hilus and perihilar region. Origin:
Description: Selective arteriography of the right supreme intercostal artery shows filling of the posterior intercostal arteries and collateral network supplying the right lung. Origin: