Unilateral Pulmonary Aplasia

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By Lalit Nirwan, MD [1]

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Clinical History: A 7-year-old girl presented with difficulty breathing for last three months and mild dull aching pain on left side. She was referred to the Radiology Department for evaluation.

Chest radiograph revealed a complete radiopaque left hemithorax with gross mediastinal and tracheal shift toward ipsilateral left side, with loss of lung volume, crowding of ribs and compensatory hyperinflation and herniation of right lung.

Contrast enhanced Computed Tomography of thorax was done.
Contrast enhanced CT, axial and coronal section of thorax, shows complete absence of left lung with gross mediastinal and cardiac shift toward the left side. Right lung shows compensatory hyperinflation with herniation toward left side. Images revealed single right dilated pulmonary artery, absence of left pulmonary artery. Ascending and descending aorta are visualized.
CT of thorax, lung window confirms complete absence of left lung parenchymal tissue.
Volume rendered reconstructed images shows complete absence of left lung, compensatory enlarged right lung with hyperinflation and herniation, single right pulmonary artery and rudimentary blind ended left main bronchus.
Coronal reconstructed CT images revealed associated D10 hemivertebra.

Diagnosis: Unilateral Pulmonary Aplasia
Discussion: This is a rare congenital anomaly, occurring in 1 in 1,000,000.
Agenesis — Complete absence of bronchus and lung parenchyma
Aplasia — Rudimentary bronchus with complete absence of parenchyma
Hypoplasia — Presence of variable amounts of bronchial tree, pulmonary parenchyma and vasculature
Developmental causes: Intrauterine compressions - Extrathoracic compressions oligohydraminos, fetal ascitis; Thoracic cage compressions - Thoracic bone dysplasia, muscular disease; Intrathoracic compression - Diaphragmatic defect
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Disclosures:

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