Unilateral pulmonary opacity with herniation of contralateral lung to same side

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A 19-year-girl was referred to our Institute from a primary healthcare for lower respiratory symptoms. She gave history of recurrent respiratory symptoms since early childhood. Her chest radiograph showed left hemithorax having large opacity, mediastinal structures shifted to left, and right lung crossing the midline to left (Figure 1). Her sputum samples were submitted for microbiological examination and she was started with empirical antibiotics. Her cough and sputum subsided after antibiotics.

Question: what is the diagnosis?

She underwent detailed contrast enhanced computed tomography study of thorax (Figure 2). There was no left bronchial tree or left pulmonary vasculature.

(In all scans, symmetry of thoracic cage is maintained despite of major anatomical changes in thoracic organs suggesting a congenital etiology).

Bronchoscopic examination revealed absence of origin of left main bronchus, trachea being followed by dilated right main bronchus.

Figure 1: Chest radiograph, posterio-anterior view, showing left hemithorax having large almost uniform opacity, trachea and mediastinal structures shifted to left and right lung crossing the midline to left (arrow heads).
Figure 2: Chest CT scan: (Figure 2A & B) axial scans showing mediastinum grossly displaced to left, right lung crossing the midline to left. There is no associated thoracic cage volume loss over left side (that is seen in acquired pathology) suggestive of congenital disorder. (Figure 2C & D) axial scans, trachea being continued to right main bronchus only, not even rudimentary left main bronchus seen. (Figure 2C to 2E) right pulmonary vasculature enlarged due to the absence of left pulmonary vasculature, entire pulmonary blood being channeled through right. (Figure 2E & F) heart is shifted near left posterior thorax. (Figure 2G & H) axial scans showing details of cardiac chambers fully displaced to posterior thoracic space, descending aorta displaced to left. Bifurcation of trachea to right upper lobe and right intermediate bronchus is seen. (Figure 2H) axial scan with right pulmonary artery elongated due to displaced heart to further left.
bronchus and all airways on right side were grossly altered in size and position (Figure 3, images obtained during bronchoscopy; and Appendix Video 1, Video clip obtained during bronchoscopy). Airways were full of secretions; bronchial aspirate was submitted for microbiological culture that confirmed *streptococcus pneumoniae*; that matched with sputum culture submitted earlier.

**Diagnosis**

Based on the features described here, a diagnosis of *left pulmonary agenesis with superadded infection of remaining right lung* was made.

**Discussion**

Unilateral whole lung opacity is frequently encountered in clinical practice that requires careful consideration of available clues, including position of trachea, thoracic cage symmetry and telltale signs of thoracic trauma or surgery. Differential diagnosis of unilateral lung opacity extends from congenital conditions to inflammatory, infective and malignant conditions (Table 1). Complete atelectasis secondary to main bronchial obstruction is most often due to a central mass, but occasionally may result from other causes like foreign bodies, mucus plugs, endobronchial tuberculosis or external compression due to lymphnodes, tumors or aortic aneurism. Traumatic bronchial rupture is always having history of blunt trauma; rib fractures are usually associated with bronchial rupture in patients over age 30. Pneumonectomy can be easily be detected due to a history of prior thoracic surgery, presence of post-operative skin scar and a radiological appearance of asymmetrical thoracic cage. In pulmonary agenesis, thoracic cage usually appears symmetric, although the trachea and mediastinum are deviated to the affected side.

Pulmonary agenesis is a rare congenital disorder characterised by complete absence of bronchus, lung parenchyma and

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**Figure 3:** Images captured during fibre-optic bronchoscopy, seen from head end; (3A to 3C) trachea is continuing to right main bronchus, no evidence of any left main bronchus—not even rudimentary one—seen. (3D) Bronchoscope in right intermediate bronchus, bronchial openings of right lower lobe, right middle lobe and superior segment of right lower lobe seen.
Table 1: Lung diseases and disorders with a radiological appearance of opaque hemithorax.

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<th>Position of trachea</th>
<th>Possible underlying etiology</th>
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| Pushed (shifted to contralateral side) | Large pleural effusion (most frequent)  
Large intrathoracic thoracic mass |
| Pulled (shifted to ipsilateral side) | Pulmonary agenesis  
Complete atelectasis  
[Foreign body (in children), endobronchial tumor (in adults),  
malpositioned endotrachial intubation, external compression,  
traumatic bronchial rupture]  
Pneumonecтомy |
| No change (central) | Adults—bronchial carcinoma, accompanied by pleural effusion and atelectasis  
Children—extensive pneumonia  
Pleural mass |

Pulmonary vasculature; the prevalence being around 34 per million live births.\(^1\) Pulmonary agenesis co-exists with other systemic anomalies including cardiovascular, gastrointestinal, musculoskeletal or urogenital system in over half of the patients.\(^2\) It is often diagnosed during childhood; half of those with unilateral lung agenesis die before the age of five years. Congenital heart diseases are seen in nearly one-third of patients. Left lung agenesis is comparatively having a longer life expectancy.\(^2\) First expression in adult life is still rarer. The natural course of pulmonary agenesis is highly variable being dependent on associated congenital malformations and development of complications. Patients may remain asymptomatic till early adulthood or present with respiratory insufficiency since birth. Isolated unilateral lung agenesis may, however, be compatible with a normal life.\(^2\) Contralateral normal lung involvement by infection or atelectasis often leads to respiratory distress.

Pulmonary agenesis may be overlooked as a collapse or pleural effusion by primary care physicians. Pulmonary agenesis and aplasia are to be identified as the later is characterised by the presence of a rudimentary bronchus. Further, both of them differ from pulmonary hypoplasia having no immature pulmonary tissues seen in later disorder. Diagnosis of pulmonary agenesis can be secured by demonstrating absence of even rudimentary bronchus, lung parenchyma and any pulmonary vasculature using radiological imaging like contrast-enhanced CT, pulmonary angiography or magnetic resonance imaging and carrying out bronchoscopy.\(^3\)

Asymptomatic pulmonary agenesis cases require no treatment apart from treatment of chest infections.\(^3\) Long-term prognosis is highly variable being dependent on the presence of co-existing congenital anomalies and involvement of the remaining single lung in disease process.
Appendix

Appendix Video 1: Video clip obtained during bronchoscopy. No evidence of any left main bronchus seen. Bronchoscope is moving from trachea to right main bronchus, opening of right upper lobe bronchus seen at 1 o’clock position and then continuing to right intermediate bronchus, bronchial openings of right lower lobe, right middle lobe and superior segment of right lower lobe seen.

Competing interests:
Nil.

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