Pulmonary Agenesis

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Pulmonary agenesis is an extremely rare condition with a reported prevalence of 34 per 10 lacs live births(l). About half the cases have associated congenital malformations of the cardiovascular, skeletal, gastrointestinal or genitourinary systems(2). A history of recurrent chest infections during first year of life may be elicited. More often, the patient may be asymptomatic and the diagnosis is suspected from chest X-ray or detected during autopsy. The first case from India was reported at autopsy in 1923(3). Subsequently, few other cases have been reported(4-7). The current communication documents 5 cases of pulmonary agenesis with varying presentations picked up over a period of 4 years. These cases were suspected and then documented during life.

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Case Reports

Five cases of pulmonary agenesis diagnosed at G.T.B. Hospital, Delhi are presented. Four cases presented with recurrent history of chest infection and respiratory distress for the last 1 months to 2 years. All these patients were being treated by private practitioners for collapse consolidation before admission to our hospital. At our institution, the diagnosis of pulmonary agenesis was suspected after chest skiagram and special efforts were made to rule out the entity. One case who was asymptomatic was diagnosed accidentally by X-ray which was done to rule out traumatic injury after a fall from height. Of th 4 symptomatic cases, 3 had congenital anomalies in the form of hemivertebrae and one case also had an associated ventricular septal defect *(Table I).* Clinically, all cases had mediastimal shift with dullness and decreased air entry on the affected side. X-ray chest revealed symmetric bony cage, opaque hemithorax, mediastinal shift with herniation of the normal lung towards the affected side *(Fig. 1).* Ultrasonography done in all cases revealed absence of lung tissue and shifting of heart to the affected side. In 3 cases' the diagnosis was confirmed by CT scan of the chest *(Figs. 2-4)* and in two by bronchoscopy *(Table I).*

TABLE I - *Clinical Features and Diagnosis of Pulmonary Agenesis.*

Cases	Age at presentation	Age at onset of symptoms	Symptoms	Side congenital	Associated anomalies	Diagnostic Modalities	
						Bronchoscopy	CT Scan
1-	6 mo	3 mo	Fever, cough, respiratory	Right	Hemivertebrae D_{12}, L_1,	-	Righ lung agenesis
2.	12 yr	—	Asymptomatic	Right	None	Rudimentary right main bronchus seen	-
3.	4 yr	$2^{1/2}$ mo	Fever, cough, respiratory distress	Left	Hemivertebrae T_{10}-T_{12}; Ventricular septal defect	-	Agenesis of left lung
4.	6 yr	4 yr	-do-	Right	Hemivertebrae T_7-T_9	-	Aplasia of right lung
5.	4 yr	1 mo prior to presentation	-do-	Right	None	Opening of right upper lobe bronchus closed. Rest of bronchial tree normal	-

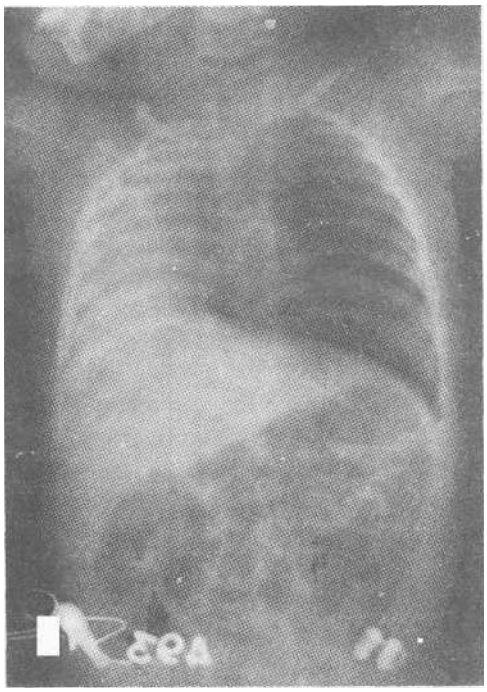

Fig. 1. Skiagram showing opaque right hemithorax with herniation of left lung.

Discussion

The developmental anomalies of the lung can be categorized into 3 groups(8): (i) Group I (Agenesis)-complete absence of bronchus, lung tissue and vessels; *(ii)* Group II (Aplasia)-Rudimentary bronchus is present but there is no lung tissue and pulmonary vessels; and *(iii)* Group III (Hy-poplasia)-Rudimentary bronchus as well as hypoplasia of lung tissue and pulmonary vessels.

The age of presentation is variable and depends on the typo of lesion present. Pulmonary agenesis is commonly associated with other congenital anomalies and may present in newborn period or in early life(9). Sometimes an isolated lesion may get detected in later years on routine X-ray done for some other ailment. Majority of patients present with recurrent chest infections since early life. Four of the five cases that we saw also presented with history of recurrent chest infection since early child-
hood. Clinically, the disease closely mimics pulmonary collapse of the affected side.
The entity should be kept in mind and

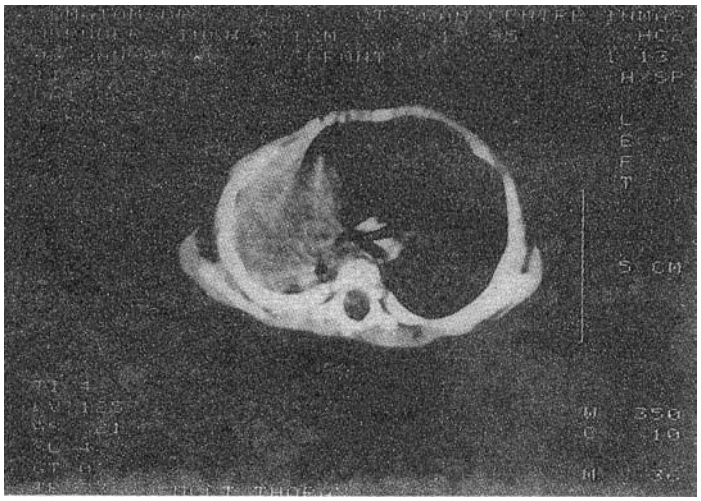

Fig. *2. CT scan of chest showing origin of left main bronchus and its branching. The right main bronchus is not seen.*

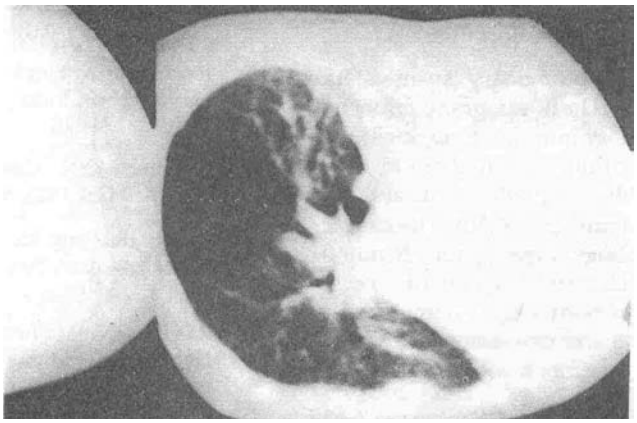

Fig. 3. CT scan of chest depicting right main bronchus with emphysematous changes in the right middle lobe. The left main bronchus is not visualized.

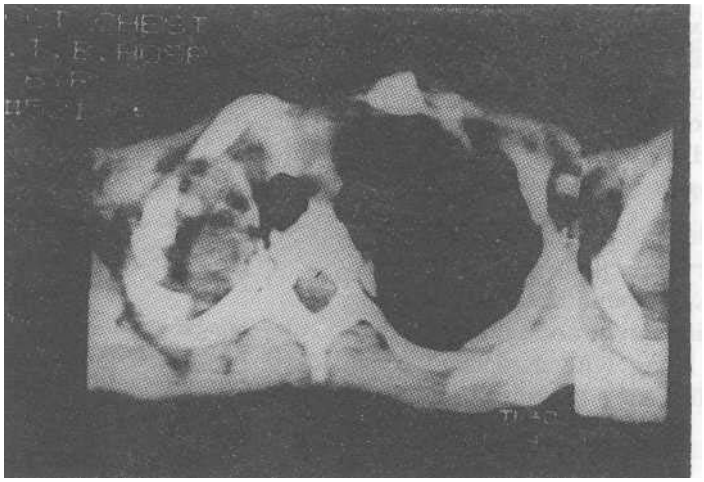

Fig. 4. Aplasia of the right lung. The proximal right bronchus is seen. There is no aerated lung parenchyma on the right side.

strongly suspected when chest X-ray reveals bony symmetry, opaque hemithorax with mediastinal shift and herniation of the contralateral lung to the affected side.

Three of the five cases described had associated hemivertebrae out of which one case also had a ventricular septal defect. A variety of congenital abnormalities may be associated with pulmonary agenesis. These may include cardiovascular (ventricular septal and atrial septal defects, tetralogy of Fallot), skeletal (hemivertebrae, absent ribs), gastrointestinal (esophageal atresia, imperforate anus), genitourinary (absent or

polycystic kidney) or other anomalies (hypoplastic trachea, ear deformities).

Diagnosis of pulmonary agenesis has been made on chest skiagram, bronchoscopy, bronchography and angiography. With the advent of CT scan, these invasive procedures which entail significant risk have become unnecessary(l0). The characteristic CT findings reveal opaque hemithorax with mediastinal shift towards the affected side and bony cage symmetry. The CT scan also clearly delineates lung parenchyma with pulmonary and bronchial tree(11).

No treatment is required in asymptomatic cases. Treatment is necessary for chest infections. Patients having stumps (hypoplast bud) may require surgical removal if postural drainage and antibiotics fail to resolve the infection. Corrective surgery of associated congenital anomalies, wherever feasible, may be undertaken(7). Overal, prognosis depends on two factors. Firstly, the severity of associated congenital anomalies and secondly, involvement of the normal lung in any disease process.

To conclude, clinical findings of recurrent respiratory infections and radiologic evidence of opaque hemithorax, bony symmetry and herination of normal lung to the affected side, along with associated congenital anomalies, are suggestive of pulmonary agenesis.

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