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CASE REPORT

Congenital Agenesis of Lung - A Case Report

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Abstract

The term 'Agenesis of Lung' is taken to mean Partial or almost complete absence of growth in the lung.[1]The rarity of this condition is evident by the infrequent reporting of such cases in literature with prevalence of 34 per million live births. The condition was first discovered accidentally at the autopsy of an adult female in 1673, by De Pozze[1]. From India, the first case was reported by Muhamed[2]in 1923, of a left sided pulmonary agenesis in a medicolegal autopsy. Munch Meyer[3]diagnosed it clinically in 1885. Subsequently a few more case reports have appeared and by 1977, over 200 cases of under development of the lung have been reported.Agenesis of lung,may present in adult life with features of recurrent chest infections and radiologically may mimic many common conditions presenting as opaque hemithorax with ipsilateral shifting of mediastinum.Here, a case of a young man presenting with frequent attacks of cough expectoration and progressive dyspnoea since childhood,proved to be a case of left pulmonary agenesis on CT scan and bronchoscopy, is to be discussed.

Keywords:Pulmonary agenesis, Recurrent childhood respiratory infection, Herniation of right lung

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Introduction:

The term 'Agenesis of Lung' is taken to mean Partial or almost complete absence of growth in the lung.1The rarity of this condition is evident by the infrequent reporting of such cases in literature with prevalence of 34 per million live births. The condition was first discovered accidentally at the autopsy of an adult female in 1673, by De Pozze[1]. From India, the first case was reported by Muhamed[2]in 1923, of a left sided pulmonary agenesis in a medicolegal autopsy. Munch Meyer[3]diagnosed it clinically in 1885. Subsequently a few more case reports have appeared and by 1977, over 200 cases of under development of the lung have been reported. Most authors describe a single or a small number of cases. The most exhaustive reviews are those of Oyamada et al[4]., Vale[5], Maltz and Nadas[6]and Sbokos and McMillan[7] Here, a case of a young man presenting with frequent attacks of cough and fever since childhood.

Needless to say, bilateral agenesis is incompatible with life. Unilateral agenesis of the lung is much less rare and may present with varying degrees of

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severity. They are often wrongly diagnosed for more common conditions of unilateral volume loss and it is even more challenging if it comes to notice in adult life.Here we report a case of young man presenting with right pulmonary agenesis. Probably this is the first case report for Lung Agenesis.

Case History:

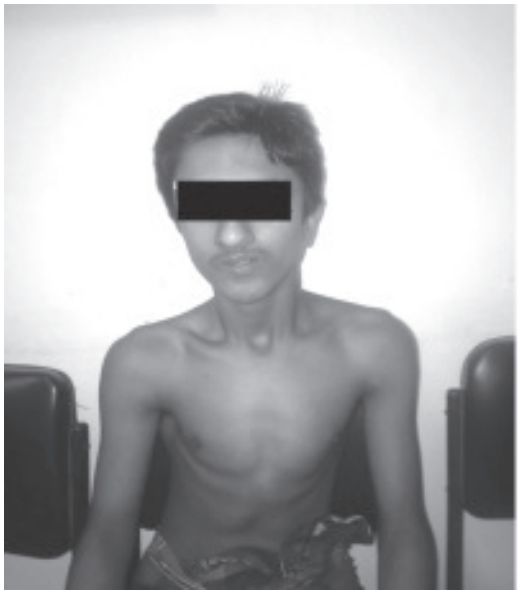
A 24 year old male presented with insidious onset, progressive shortness of breath since childhood and frequent episodes of cough with muco-purulent sputum, often one cupful per day, yellowish in colour. There were no history of orthopnea, palpitation, wheezing, chest pain, coughing out of blood,anorexia and weight loss.He had no past history suggestive of pulmonary tuberculosis. His perinatal history was insignificant and no history of similar complaints in any of his siblings. On examination, he was an average built male, malnourished, preferring right lateral decubitus. Pallor, icterus, clubbing, engorged neck veins and lymphadenopathy were absent.On inspection visible pulsation in right side of chest,drooping of shoulder seen in right side .On palpation, movement diminished in right side ,trachea deviated to right and apex beat placed at right 4^{th} intercostal space in mid clavicular line. Expansion of chest was 2cm and vocal fremitus diminished throughout the right side except 2^{nd} intercostal space to upwards .On percussion ,right side had impaired notefrom 2^{nd} intercostal space downward along all three line,resonant in rest of the areas .On auscultation, vesicular breath sound heard with reduced vocal resonant in above mentioned area, bronchial breath sound heard in right side from 2^{nd} intercostal space to upwards .Liver was not palpable , other systems were within normal limits.

Chest radiograph showed homogenous opacity in the almost all zone in right side except apical area and part of lower zone , obliterating the right cardiophrenic angle with gross shifting of the mediastinum to the right and (Fig. 1).Echodoppler study revealed heart shifted right thoracic cavity .Contrast enhanced computed tomogram showed right lung is hypoplastic and herniation of left lung to the right and (Fig. 3).congenital hepatic herniation through congenital defect of right dome of diaphragm.He was *diagnosed* as *congenital agenesis right of lung with associated defectin right dome of the diaphragm*.

Chest Radiograph showing homogenous opacity in the Right lower zone,obliterating the Right costophrenic angle with gross shifting of the mediastinum to the Right and scoliosis with convexity to the left and reticulonodular shadows in the right lower zone.

Mediastinalwindow with contrast showing absence of opening of Right main bronchus and ight pulmonary artery not seen.

Parenchymal window of contrast enhanced CT scan chest showing absence of Right lung and herniation of leftlung to the right,also there are bronchiectatic changes.



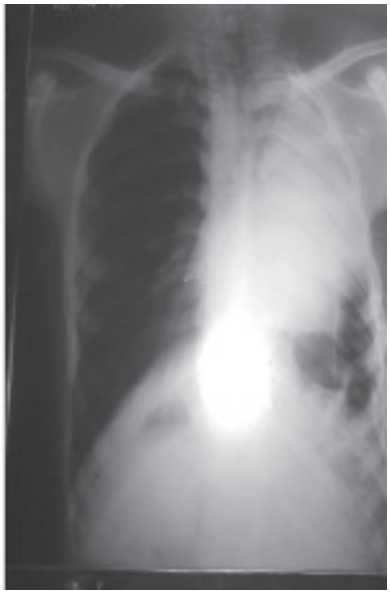


Fig. 1(a) Fig: 1(b)

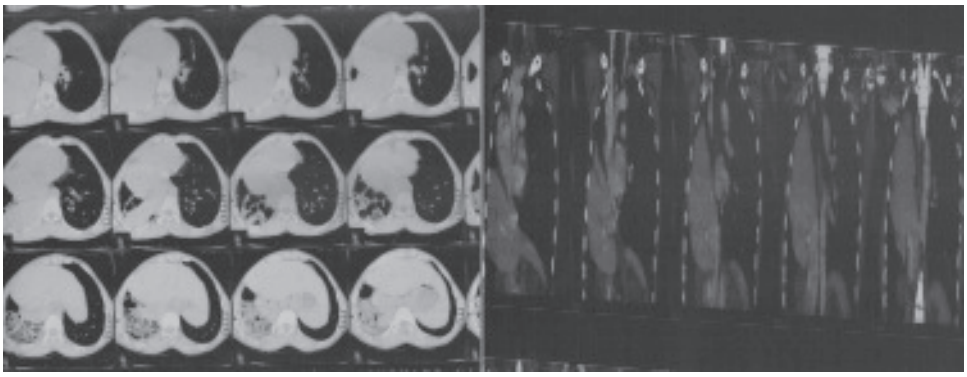


Fig.-3

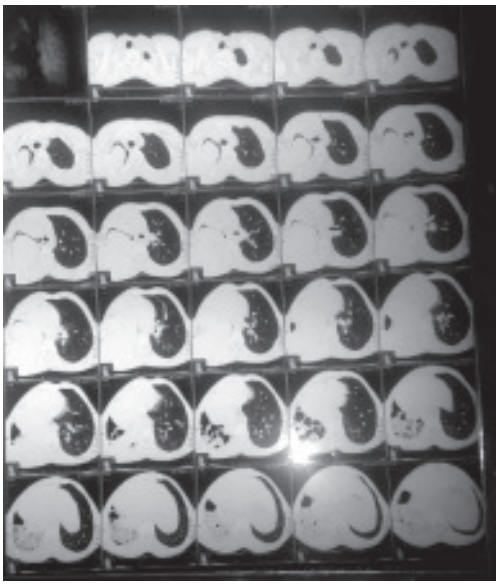


Fig.-3

Discussion:

Unilateral agenesis of the lung may be present to varying degrees of severity. The left lung is affected more frequently than the right, males predominate over females and the majority of cases exhibit other congenital abnormalities like patent ductus arteriosus, pulmonary artery atresia, cardiac malformation, tracheo-esophageal fistula, cardiac malformation and horse-shoe kidney.However, several older reports prove that other anomalies are more associated with right sided agenesis and persons with right sided agenesis mostly die within first year of their life,due to associated cardiac malformations.[2] Originally Schneider (1912)[3] classified agenesis into three groups which was later on modified by Boyden[4] as-

Type I(Agenesis): Complete absence of lung and bronchus and absence of blood vessels to the affected side.

Type II(Aplasia):Rudimentary bronchus with complete absence of lung tissue.

Type III(Hypoplasia):Presence of variable amounts of lung parenchyma,bronchial tree and supporting vasculature.

Our patient has been classified as Type III .Schneider's agenesis type I and typeII, the affected side contains no lung tissue, and only the existing lung gets the branch from the main pulmonary artery. In Schneider's agenesis Type III Presence of variable amounts of lung parenchyma, bronchial tree and supporting vasculature, an observations has been seen in our case also. Clinical presentation of agenesis lung is marked by its variety from recurrent childhood respiratory infection resulting from imperfect drainage of lung secretions or from the spillover of pooled secretions from a blind bronchial stump into initially normal lung tissue, frequent haemoptysis due to bronchiectasis of remaining lung to major organ malformation leading the patient to succumb in early life. A similar case was reported in Turkey as, a 30-year-old man presenting with dyspnoea was diagnosed to have right lung agenesis and left pulmonary bronchiectasis.[5]

Autosomal recessive chromosomal aberration associated with consanguineous marriage[6], deficiency of vitamin A,intrauterine infections, environmental factors have been held responsible for the etiology of congenital lung malformations. During normal development, the heart shifts to the left in the 4th week of foetal life and simultaneously the trachea develops as a ventral diverticulum arising from the foregut.[7] Pulmonary agenesis or aplasia occurs perhaps due to the failure

of the bronchial analogue to divide equally between the two lung buds. If this balance is not established, one side will develop normally while the other will fail completely (agenesis/aplasia) or undergo only limited development (dysplasia or hypoplasia).

In adults, unilateral agenesis of lung may mimic collapse, thickening of pleura, destroyed lung, pneumonectomy, scoliosis with pleural effusion, diaphragmatic hernia, adenomatoid cystic malformations and sequestrations.CT Chest, which provides detailed description of bronchial tree, parenchyma and vasculature is considered to be the most definitive investigation to diagnose agenesis when chest radiograph is not diagnostic.[8]Bronchography is almost obsolete now, but bronchoscopy is useful to demonstrate rudimentary bronchus. Pulmonary angiography or MRI Angiography is considered to show the absence of ipsilateral pulmonary vessel and cardiac catheterization may be needed to rule out cardiac malformations and to quantify Pulmonary artery pressure. In our case these could not be done as the patient denied to do Pulmonary angiography and Bronchoscopy .

No treatment is required in asymptomatic cases. Treatment is necessary for recurrent chest infections. Patients having bronchial stumps 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.[9]

Conflict of interest statement

We have no conflict of interest regarding the article.

References:

1. De Pozze, Brescia MA, Amermon EE, Sharma KK. 1960;77:485–490.
2. Muhamed K.S.N. Absence of left lung. Ind. Med. Gaz. 1923;58:262–64.
3. Munchmeyer cited by Ferguson FF, Meuhauser E.B.D. Congenital absence of lung and other anomalies of tracheobronchial tree. Am. J. Reontgen, 1944;52:459–471.
4. Oymada A, Gasul B.M, Holinger P.M. Agenesis of the Lung. Am J Dis Child. 1953;85:182–187.
5. Vale A.R. Agenesis of the lung. Am. J. Surg, 1955;89:90–100.
6. Maltz D.L, Nadas A.S. Agenesis of the lung–Presentation of eight new cases and review of the literature.Paediatrics. Springfield, 1968;42:175–188.
7. Sbokos C.B., McMillan IRK. Agenesis of the lung. Brit J Dis Chest. 1977;3:183–197.
8. Schneider P, Schawatbe E. E. Die Morphologie der Missbildungen Des Menschen Under Thiere. Jena: Gustav Fischar. 1912;3 Part.2:817–822.
9. Boyden E. Developmental anomalies. AM. J. Surg, 1955;89:79–88.