

Lung Hypoplasia Without Other Congenital Anomaly- A Rarely Encountered Entity

Ghosh N¹, Das N², Nayak K³

¹Dr. Nilanjan Ghosh, M.B.B.S, M.D, RMO cum Clinical Tutor, ²Dr. Nabanita Das, M.B.B.S, Post Graduate Trainee, ³Dr. Kaustav Nayak, MBBS, DCH, MD, Professor. All from the RG Kar Medical College, Kolkata, India.

Introduction

Lung agenesis is a rare congenital anomaly arising as a result of embryological defect or secondary to some other congenital anomaly. It represents a broad range of malformations characterized by incomplete development of lung tissue. The gross morphology of the lung is essentially unremarkable but the number and size of airways, vessels and alveoli are decreased. Underdevelopment of alveolar tissue results in fibrotic and non functioning lung. Bronchiectatic changes have also been reported¹. It is very commonly associated with other congenital anomalies of the diaphragm², urinary system³, cardiovascular system (Tetralogy of Fallot)⁴, central nervous system (anencephaly, hydroencephaly)⁵, and musculoskeletal anomalies of thoracic cage, Klippel Feil syndrome and Downs syndrome⁶.

The Case

A seven years age female was admitted to our department with history of cough, difficulty in respiration, sputum production, reduced appetite and physical activity for long duration. The child had been suffering from fever on and off with cough and respiratory difficulty since infancy.

She was febrile, had respiratory distress with grade 3 clubbing. There was shifting of the trachea to the left. Movements were diminished and percussion note was dull over the left hemithorax. On auscultation air

Abstract

A case of left sided lung hypoplasia is described in a seven year old female. She was admitted in our department with complaints of recurrent chest infections since infancy. Clinical examination revealed a febrile child with respiratory distress and with grossly diminished breath sound with patchy coarse crepitations on left hemithorax. X-ray chest revealed non homogenous opaque shadow on left hemithorax. Bronchoscopy and CT thorax confirmed the diagnosis of left sided hypoplasia of lung. No other developmental anomaly was noted.

Key words: Lung hypoplasia

entry was very poor on left side with diffuse crackles. Examinations of other systems were within normal limits.

Complete blood count showed leucocytosis and sputum examination for Acid Fast Bacilli on direct smear examination was negative. Chest skiagram showed a radio opaque left hemithorax with nonhomogeneous opacity at mid zone and gross shifting of mediastinum to the left. CT scan of thorax revealed multiple cystic areas in left lung lower zone with patchy areas of consolidation. There was herniation of right lung along the mediastinum with shifting of the mediastinum to the left side. The lower lobe bronchus of left side was identified but the upper lobe bronchus could not be visualized. No collapsed segment was seen adjacent to the left lower lobe. Subsequently ultrasonography of the abdomen and echocardiography were performed, both of which were normal.

She was put on to IV antibiotics. Once her symptoms improved she was referred to department of cardio thoracic surgery of a higher center for consultation. She was advised conservative treatment.

Address for correspondence

Dr. Nilanjan Ghosh
E-mail: niltughosh@gmail.com

This work is licensed under a Creative Commons Attribution 3.0 License.



How to cite this article ?

Ghosh N, Das N, Nayak K. Lung Hypoplasia Without Other Congenital Anomaly- A Rarely Encountered Entity. J Nepal Paediatr Soc 2013;33(2):138-140.

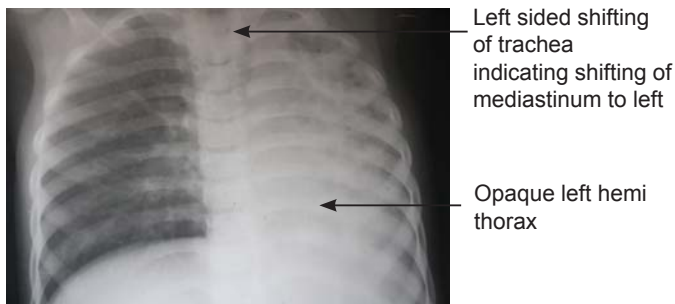


Fig 1: X-Ray showing radio opaque left hemithorax with inhomogenous opacity at midzone and gross shifting of mediastinum to left.

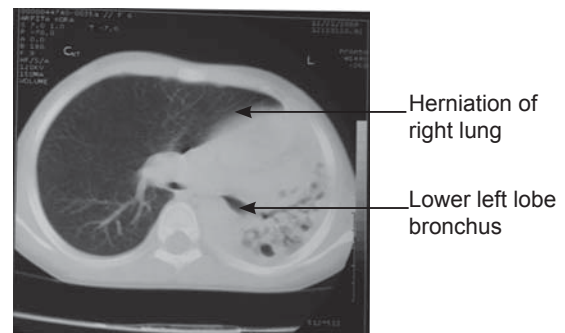


Fig 2: CT scan revealing lower left lobe bronchus but upper left lobe bronchus not seen with herniation of right lung.

Discussion

Schneider classified agenesis into three groups, which has been subsequently modified in 1955 by Boyden⁷. Depending upon the stage of development of the primitive lung bud, pulmonary agenesis is classified into three categories:

Type 1 (Agenesis) – Complete absence of lung and bronchus and no vascular supply to the affected side.

Type 2 (Aplasia) – Rudimentary bronchus with complete absence of pulmonary parenchyma.

Type 3 (Hypoplasia) – Presence of variable amounts of bronchial tree, pulmonary parenchyma and supporting vasculature.

Our patient would classify as Type 3.

Hypoplasia of the lung may be regarded as primary (idiopathic) or secondary. Hypoplasia may be either general, involving the whole lung, or partial. Primary pulmonary hypoplasia may be caused by deficient TTF-1, GATA factors, hepatocyte nuclear factor HNF310, epidermal growth factor and its receptor, EGFR; mitogen-activated protein [MAP] kinase. More commonly it is secondary arising as a result of small fetal thoracic volume, prolonged oligohydramnios, early rupture of membranes at 15-28 weeks gestation⁸, longer latent period before delivery, decreased fetal breathing, decreased pulmonary perfusion, congenital heart diseases and trisomies 18, 13, 21⁹. Incidence of pulmonary hypoplasia ranges from 9–11 per 10,000 live births and 14 per 10,000 births¹⁰ without sex predilection.

In patients with pulmonary agenesis, the clinical profile and the time of presentation vary. The basis of this variation may be related to extent and cause of hypoplasia as well as to the timing of the etiologic events that led to this anomaly¹¹. Some patients may present in infancy with severe respiratory distress

within few hours after birth whereas some may be completely asymptomatic. Others will present months or years later and even during adulthood with repeated pulmonary infections and wheezing¹². Children with pulmonary hypoplasia are prone to recurrent respiratory infections as a result of abnormal angulations in the airways, defective mucociliary clearance and pooling and spilling of secretions from blind passages¹³. Pulmonary hypoplasia may be diagnosed incidentally during childhood when complicated by pulmonary infection^{14,15,16}. Other than chest x-ray, other modalities of diagnosis are CT thorax^{17,18}, fibre optic bronchoscopy and if possible pulmonary angiography and bronchography. Thomas et al. suggested that CT is more useful than bronchography in assessing the hypoplastic lung, especially for the purposes of its surgical removal¹².

Treatment of hypoplasia is in form of medical as well as surgical care. Before delivery patient is treated medically with repeated amniotomies with or without the use of tocolytics, antibiotics and steroids. After delivery respiratory support is given ranging from oxygen to mechanical ventilation including ECMO (Extracorporeal membrane oxygenation). Surfactant administration at 4 ml/kg improves survival rate. Surgical care consists of intrauterine vesicoamniotic shunts and endoscopic ablation of valves and PLUG (Plug the Lung Until it Grows) by fetoscopic tracheal occlusion with a clip. Post-delivery surgery can be done to correct diaphragmatic hernia, cystic adenomatoid malformations and decompresses pleural effusions. Treatment in older children consists of control of recurrent infections, symptomatic treatment in form of expectorants and bronchodilators and management of other complications. Prophylaxis for respiratory syncytial virus, pneumococcus and influenza infections are recommended.

Surgery is seldom required for agenesis or aplasia, as it can be managed on conservative lines. The prognosis in these cases depends upon the functional integrity of the remaining lung as well as the

presence of associated anomalies. Left sided agenesis is more common and these subjects have longer life expectancy than those with right sided agenesis¹⁹. It is important to investigate the coexistence of this anomaly. Asymptomatic cases do not require any treatment if there are no additional anomalies, but these cases carry high-risk in any surgery because of low respiratory reserve. Pulmonary hypertension as a complication can occur, because of a reduction in the pulmonary vascular bed that, if associated with congenital heart disease (left-to-right shunt), can progress to irreversible vascular disease^{20,21}. Misdiagnosis may subject the patient to the unnecessary risks of bronchoscopy and to potential perforation of the rudimentary bronchus.

Conclusion

Here we are reporting a case of unilateral pulmonary hypoplasia without any other congenital anomaly in a seven year old child, that very late age of presentation and diagnosis is a very rare occurrence. And before starting treatment with antitubercular drugs as given in our patient, much consideration should be given to such a rare entity.

References

1. Calenoff L. Unilateral pulmonary hypoplasia in an adult. *Am J Roentgenol* 1964;91:265-272.
2. Areechon W, Reid L. Hypoplasia of lung with congenital diaphragmatic hernia. *Br Med J* 1963;1:230-233.
3. Hislap A, Hey E, Reid L. The lungs in congenital bilateral renal agenesis and dysplasia. *Arch Dis Chil.* 1979;54:32-38.
4. Johnson RJ, Howorth SG. Pulmonary vascular and alveolar development in tetralogy of fallot: A recommendation for early correction. *Thorax* 1982;37:893-901.
5. Cooney TP, Thurlbeck, wm. Lung growth and development in anencephaly and hydranencephaly. *Am Rev Respir Dis* 1985; 132: 596-601
6. Cooney TP, Thurlbeck WM, Pulmonary hypoplasia in Down syndrome. *N Engl J Med* 1982;307:1170-1173.
7. Boyden EA. Observations on the history of bronchopulmonary segments. *Minn Med* 1955;38:597-8.
8. Yoshimura S, Masuzaki H, Miura K, et al: Diagnosis of fetal pulmonary hypoplasia by measurement of blood flow velocity waveforms of pulmonary arteries with Doppler ultrasonography. *Am J Obstet Gynecol* 1999;180:441-6.
9. Warburton D, Olver BE: Coordination of genetic, epigenetic, and environmental factors in lung development, injury, and repair. *Chest* 1997;111:119S-122S.
10. Kumar P, Burton BK. Congenital Malformations. Evidenced Based Management. Chicago: The McGraw Hill companies; 2008. Chapter 22, Pulmonary Hypoplasia; p.143
11. Rutledge JC, Jensen P: Acinar dysplasia: A new form of pulmonary maldevelopment. *Hum Pathol* 1986;17:1290.
12. Thomas RJ, Lathif HC, Sen S, Zachariah N, Chako J. Varied presentations of unilateral lung hypoplasia and agenesis: a report of four cases. *Pediatr Surg Int* 1998;14:94-95.
13. Gie RP, Beyers N, Schaaf HS, Nel ED, Smuts NA, van Zyl S, Donald PR (1995) TB or not TB? An evaluation of children with an incorrect initial diagnosis of pulmonary tuberculosis. *S Afr Med J* 85: 658-662.
14. Page DV, Stocker JT: Anomalies associated with pulmonary hypoplasia. *Am Rev Resp Dis* 1982;125:216.
15. Landings BH. Cong malformations and genetic disorders of respiratory tract (larynx, trachea, bronchi and lungs) state of art: *Am Rev Resp Dis* 1979;120:151.
16. Borja AR, Ransdell HT, Villa S. Congenital developmental arrest of lung. *Am Thoracic Surg* 1970;10:317.
17. Mata JM, Caceres J, Lucaya J. et al.: CT of congenital malformations of lung. *Radiographics* 1990;10:651.
18. Bhagat R, Panchal N, Shah A. Pulmonary aplasia: A CT appearance. *Indian Paediatr* 1992;29:1410-1412.
19. Oymada A, Gasul B.M, Holinger P.M. Agenesis of the Lung. *Am J Dis Child.* 1953;85:182- 187.
20. Chou AK, Huang SC, Chen SJ, Huang PM, Wang JK, Wu MH, et al. Unilateral lung agenesis--detrimental roles of surrounding vessels. *Pediatr Pulmonol* 2007;42(3):242-8. PMID:17238192. <http://dx.doi.org/10.1002/ppul.20561>
21. Clements BS. Congenital Malformations of the Lungs and Airways. In: Taussig LM, Landau LI, editors. *Pediatric Respiratory Medicine*. St. Louis: Mosby; 1999. p. 1106-36.