Journal Of Case Reports: Clinical & Medical

Case Report

A Rare Clinical Presentation of Unilateral Pulmonary Agenesis in a Neonate–Pneumothorax

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ARTICLE INFO

Received Date: December 14, 2019
Accepted Date: January 16, 2020
Published Date: January 17, 2020

KEYWORDS

Congenital malformation
Lung agenesis
Pneumothorax

Citation for this article: Pradyumna Pan. A Rare Clinical Presentation of Unilateral Pulmonary Agenesis in a Neonate–Pneumothorax. Journal Of Case Reports: Clinical & Medical. 2020; 3(1):143

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ABSTRACT

Congenital pulmonary agenesis is extremely rare anomaly. We present a case report of a one-day old boy with pneumothorax and respiratory distress with unilateral right lung agenesis.

INTRODUCTION

Pulmonary agenesis is the complete absence of the lung parenchyma, its vasculature, and its bronchus [1]. The presentation is usually with respiratory symptoms. The onset & mode of presentation is highly variable. Around 50% of cases are associated with congenital VACTERL anomalies.

CASE REPORT

A full term, 2.6 kg, male baby delivered by normal vaginal delivery was brought with complaints of respiratory distress since birth. He was born out of a non-consanguineous marriage to a 34-year-old second gravida mother with no perinatal complications. The baby cried immediately. On admission: HR-176/min, RR- 74/min with subcostal & intercostal retractions and peripheral cyanosis. His chest was normal shaped, with slightly decreased movements on the right side. The trachea was slightly deviated towards the right side. The apex beat was palpable on the right side at fifth intercostal space in anterior axillary line. There was dull percussion note on the whole of the right side of chest, and there was no cardiac dullness on the left side. Breath sounds were reduced on both side. The abdominal examination was within normal limits. Chest X- ray showed complete opacification of the right hemi thorax with the mediastinal shift towards the right and a left side showed pneumothorax (Figure 1). The gastrografin study excluded congenital diaphragmatic hernia (Figure 1). CT scan chest (Figure 2) showed complete opacification of right hemithorax with an ipsilateral absence of pulmonary artery and vein. There was a shift of mediastinum towards the right side and abrupt cut-off of right main-stem bronchus. Echo showed small PDA, dilated right atrium, right ventricle and pulmonary artery with moderate pulmonary artery hypertension.

Diagnosis of right pulmonary agenesis with left pneumothorax was made. The patient was intubated and put on the ventilator. A left intercostal tube was placed to drain the pneumothorax (Figure 3). After four days he was weaned from the ventilator and put on nasal oxygen. Left lung expanded and the intercostal tube was removed thereafter. Baby clinically improved and discharged after 17 days in stable condition. The infant showed normal development at 12 months, and the unilateral lung agenesis did not hamper the quality of life of the infant. In the last twelve months, he had only

one episode of cough and fever that was successfully managed without hospitalization.

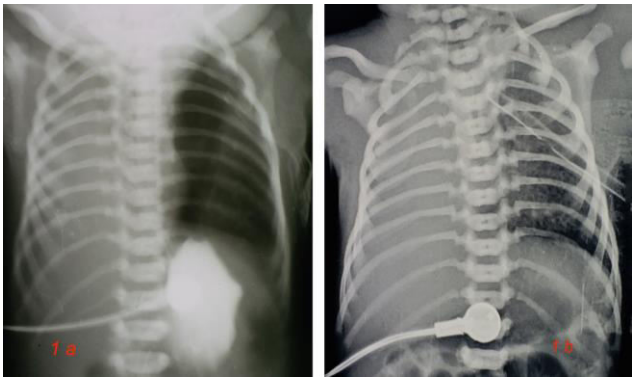

Figure 1a: X Ray chest showing right sided pulmonary aplasia with pneumothorax of left lung. Gastrografin dye is seen in the stomach.
Figure 1b: X Ray chest showing intercostal tube in situ and expansion of left lung.

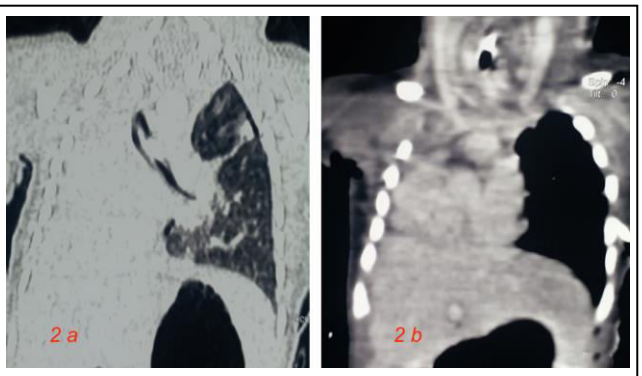

Figure 2a: CT scan chest (coronal section) showing absence of lung, pulmonary artery and veins on right side.
Figure 2b: CT scan chest showing shifting of mediastinum towards right side.

DISCUSSION

Pulmonary agenesis is a rare congenital defect first described by De Pozze in 1673 [2] with an incidence between 34-97 per million births [3]. Embryologically it is due to a failure in the development of the respiratory system from the foregut during the fourth week of gestation. Arrest at this stage of the primitive lung bud results in bilateral pulmonary agenesis while it develops only unilaterally at the later stage and leads to lung agenesis. On day twenty-two, the fetal respiratory system begins to form as a ventral outpouching of the foregut endoderm below the pharynx. This laryngo-tracheal diverticulum gradually becomes separated from the digestive tract, and the trachea bifurcates in two bronchial buds that penetrate the neighboring mesenchyma. Bronchial buds are formed at the fifth week of intrauterine life. Subsequent bronchial ramifications from the pulmonary structures are covered by blood vessels derived from the foregut mesoderm [4].

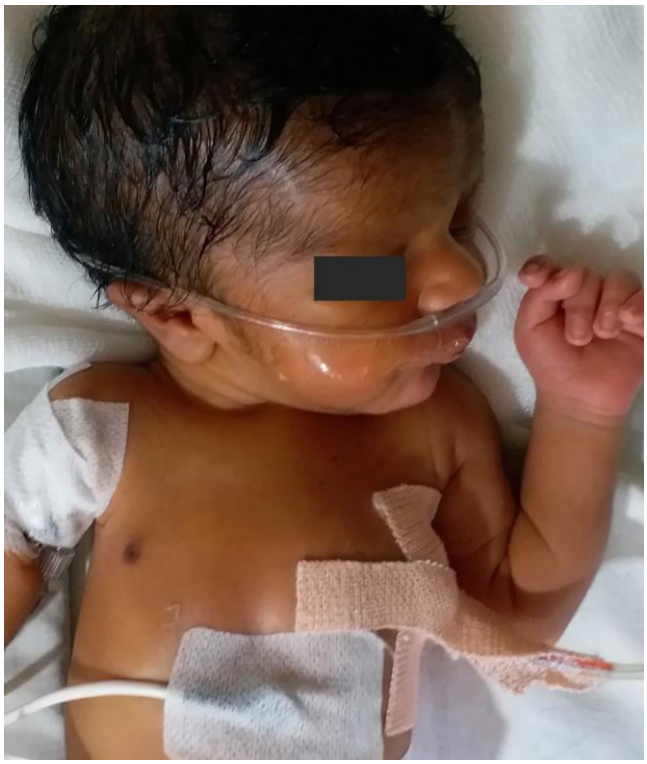

Figure 3: Clinical photo showing the intercostal tube in situ.

Monaldi divided the mal-development of the lung in four groups. Group I: No bifurcation of the trachea; Group II: Only rudimentary main bronchus; Group III: Incomplete development after the division of the main bronchus; and Group IV: Incomplete development of subsegmental bronchi and a small segment of the corresponding lobe. Pulmonary agenesis was initially classified by Schneider and Schawatbe and later modified by Boyden into three variants based on stage of development of lung bud (i) agenesis, in which there is complete absence of lung tissue, (ii) aplasia, in which rudimentary bronchus is present but no lung tissue is present, and (iii) hypoplasia, in which all the normal pulmonary tissues are present but are under-developed [5].

The etiology is not completely understood, but genetic, teratogenic and mechanical factors have been implicated. They are generally sporadic, with only a few reported as an

autosomal recessive pattern. They occur in both sexes with equal frequency and involve both lungs equally. There is a high incidence of associated cardiac, gastrointestinal, genitourinary, skeletal, central nervous system malformations and VACTERL anomalies [6]. The prenatal diagnosis of unilateral pulmonary agenesis is feasible by the total or partial absence of pulmonary parenchyma associated with an abnormal position of the heart within the thorax.

The presentation is not limited to neonatal period. Koseoglu et al. reported a 30-year-old man with right lung agenesis and left pulmonary bronchiectasis, identified during haemodialysis for chronic renal failure [7]. Musleh et al. reported a 55-year-old man with left lung agenesis discovered during mitral valve repair [8]. The most common complications in these patients are respiratory tract infections, which can lead to death. Signs of respiratory insufficiency are most commonly caused by kinking and trachea compression caused by heart rotation, mediastinal shift, and trachea curvature following pressure from dislocated aortic arch and truncus arteriosus [9]. Presently, contrast-enhanced CT Scan chest forms the standard investigation for diagnosis of lung agenesis, and pulmonary angiography and bronchoscopy are rarely required.

A diaphragmatic hernia and Swyer-James-MacLeod syndrome should be included in the differential diagnosis. No treatment is required in asymptomatic cases. Treatment is needed for lower respiratory tract infections. If postural drainage and antibiotics fail to resolve the infection, patients with stumps may require surgical removal of the stump. Corrective surgery of associated congenital anomalies should be undertaken. Various surgical corrections have been attempted to correct respiratory distress due to air trapping in normal lung and tracheal abnormality. Placement of inflatable prosthesis, aortopexy, and diaphragmatic translocation are reported [10]. These procedures provide recovery from respiratory distress by reducing heart rotation, mediastinal shift and relieving kink and tracheal compression as well as hyperinflation of lung parenchyma [9]. It is not recommended to use prophylactic surgery [6].

CONCLUSION

Congenital pulmonary agenesis is an extremely rare anomaly and may be asymptomatic throughout life or may present suddenly with severe respiratory distress. Prognosis depends on the severity of associated congenital anomalies and, the involvement of the normal lung in any disease process.

REFERENCES

1. Lee P, Westra S, Baba T, McCauley R. (2006). Right pulmonary aplasia, aberrant left pulmonary artery, and bronchopulmonary sequestration with an esophageal bronchus. Pediatr Radiol. 36: 449-452.
2. De Pozze, Brescia MA, Amermon EE, Sharma KK. (1960). Agenesis of the left Lung. Arch Pediatr. 77: 485-490.
3. Mardini MK, Nyhan WL. (1983). Agenesis of lung: Report of four patients with unusual anomalies. Chest. 87: 522-527.
4. Sadler TW. (2009). Respiratory system. In: Sadler TW, eds. Langman's medical embryology. 8th edition. MD: Lippincott Williams & Wilkins, 2009: 275-284.
5. Boyden EA. (1955). Developmental anomalies of the lungs. Am J Surg. 89: 79-89.
6. Maltz DL, Nadas AS. (1968). Agenesis of the lung: presentation of eight new cases and review of literature. Pediatrics. 2: 175-188.
7. Koseoglu N, Ucan ES, Cavdar C. (2005). Right lung agenesis and left lung bronchiectasis. Respir Med Extra. 1: 110-112.
8. Musleh GS, Fernandez P, Jha PK, Hasan R. (2004). Mitral valve repair in a 55year old man with left lung agenesis. Ann Thoac Surg. 77: 1810-1811.
9. Krivchenya DU, Dubrovin AG, Krivchenya TD, Khursin VN, Lysak CV. (2000). Aplasia of the right lung in a 4-year-old child: Surgical stabilization of the mediastinum by diaphragm translocation leading to complete recovery from respiratory distress syndrome. J Pediatr Surg. 35: 1499-1502.
10. Malcon MC, Malcon CM, Cavada MN, Caruso PEM, Real LF. (2012). Unilateral pulmonary agenesis. J Bras Pneumol. 38: 526-529.