Left Lung Aplasia with Patent Ductus Arteriosus in a Term Newborn Baby: A Case Report

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Article title: Left Lung Aplasia with Patent Ductus Arteriosus in a Term Newborn Baby: A Case Report.

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Abstract

Pulmonary aplasia is very rare congenital anomaly identified by the absence of lung parenchyma or vessels with the presence of basic bronchus. This malformation mostly accompanies other anomalies, and chest computed tomography is a useful diagnostic tool. We present a rare case of left lung aplasia with patent ductus arteriosus in a term neonate who had respiratory distress. The clinical features were observed during the first week of life, and the diagnosis of these anomalies was established by chest x-ray, thoracic computed tomography, and echocardiography. Therefore, further imaging is crucial to make the final diagnosis in term newborns who exhibit respiratory distress and total lung opacity on chest x-ray. Since the index case of lung aplasia was associated with congenital heart disease and had strong parental consanguinity, a genetic basis may have been involved in the parthenogenesis of pulmonary aplasia.

Keywords: Neonate; Pulmonary aplasia; Patent Ductus Arteriosus.

Introduction

Pulmonary aplasia is a congenital anomaly characterized by impaired growth and development of the lung parenchyma, airways, and vessels. It is a very rare congenital malformation with an estimated prevalence of 1-3.4 per 100,000 live births.[1,2,3] There are three types of congenital underdevelopment of the lungs. Type 1 or lung agenesis is distinguished by the complete absence of the lung tissue, bronchi, and blood vessels. Type 2 or lung aplasia is identified by the absence of pulmonary parenchyma or vessels, presence of basic bronchus each of which ends in a blind pouch. Type 3 or lung hypoplasia is characterized by the existence of varying amounts of bronchial tree, pulmonary parenchyma, and supporting vasculature.[3, 4] Imaging investigations, particularly a chest CT scan, are useful to diagnose lung aplasia.[5] The clinical manifestations range from asymptomatic to various complaints such as feeding problems, respiratory distress and recurrent

chest infections that mostly appear during the neonatal period or early childhood. More than half of cases result in death within the first five years of life. However, the manifestations might appear in adulthood or may remain asymptomatic throughout the life in very rare situation.[5,6] Bilateral lung agenesis or aplasia is fatal, while patients with right lung agenesis have a worse prognosis than those with left lung agenesis.[4]

Case Report

A one-day old male neonate weighing 2.7 kg was born at 38 weeks of gestation to a 27-year old primigravida mother by vaginal delivery at a local hospital. The mother was in good health during pregnancy and did not receive any teratogenic drugs or radiation. The parents were double third-degree relatives, which denoted a strong parental consanguinity. The baby had no respiration and cry at birth with an Apgar score of 4/10 and 6/10 at the 1st and 5th minutes of life, respectively. He was referred to our department at 12 hours of live because of respiratory difficulty and a suspected diagnosis of neonatal sepsis. On arrival, he had fever, no sucking, granting and less movement. On physical examination, there were sluggish primitive reflexes, fever (rectal temperature of 39ºC), tachypnea (respiratory rate of 95/min), heart rate of 171/min, subcostal and intercostal retraction, nasal flaring, diminished breathing sound on the left side, and peripheral cyanosis with oxygen saturation of 84%. There were no meconium stained mucus membranes and skin, as well as no visible deformities in the newborn baby. Blood investigations revealed hemoglobin of 15.8 gm/dl, total leucocyte count of 21700/mm^3 (Polymorphs 76%, lymphocytes 20%, eosinophils 2% and monocytes 2%), platelet count of 126000/mm^3, and C-reactive protein of 6mg/dl. Abnormal clinical findings, leukocytosis and elevated level of C-reactive protein were used for the diagnosis of neonatal sepsis. The chest X-ray showed homogenous opacity on the left lung with hyperlucency of right lung and mediastinal shift to the left side. [Figure 1] Patient was put on

intravenous fluids, oxygen, and antibiotics consisting Cefotaxime and Ampicillin intravenously, during the first and second days. In addition to the mentioned management, we start expressed breast milk by nasogastric tube and add sodium and potassium to the intravenous fluid on the third day. On the ford day of live, contrast enhanced computed tomography (CT) of the chest showed a left lung parenchymal loss, hyperlucent right lung, evidence of pulmonary vasculature and mediastinal shift toward the left side [Figure 2, 3] , all are diagnostic evidences of left lung aplasia or type 2 congenital lung underdevelopment. On the fifth day of life, the doppler echocardiography revealed a 4mm PDA with mild pulmonary artery hypertension (40mmHg) [Figure 4]. The patient was diagnosed as a case of left lung aplasia, PDA, mild pulmonary artery hypertension and neonatal sepsis. Therefore, diuretic therapy, restriction of maintenance fluid, and three doses of oral ibuprofen were recommended as medical care of PDA together with the management of neonatal sepsis. On the 8th day of live, he was stable with good breastfeeding and no need for respiratory support, as well as no changes were observed in the second chest x-ray [Figure 5]. The second doppler echocardiography showed the closure of PDA after the full course of medical management [Figure 6].

Discussion

Pulmonary aplasia is a very rare congenital malformation that can be developed unilaterally or bilaterally. Bilateral condition is extremely rare and incompatible with life.[4-7] This malformation mostly occurs during the embryonic and pseudo-glandular stages of lung development (5-17 weeks of gestation).[1] Although the main cause of this anomaly is unknown, genetic, vitamin A deficiency, mechanical, and teratogenic factors have been implicated in the pathogenesis of pulmonary agenesis.[1, 2, 5, 7] Melo et al. discovered a genetic basis for this anomaly with an autosomal recessive inheritance pattern.[9] According to the evidences of chest x-ray and thoracic computed tomography,

the index case is left lung aplasia or type 2 congenital lung underdevelopment. Diagnostic findings of thoracic computed tomography were loss of left lung parenchyma, hyperlucent right lung, mediastinal shift to the left side, and visible left lung vasculature and distally blinded main bronchus. Up to date, the majority of reported cases of lug agenesis were not accompanied by patent ductus arteriosus and were detected beyond the neonatal period; these are significant distinctions from our case.[2-7] The current case report revealed a strong parental consanguinity because they had double third-degree relations. This finding straightens the genetic basis of the mentioned anomaly, as suggested by Melo et al.[9] The clinical features of the neonate presented in this case report was respiratory distress and signs of respiratory infection which are consistent to the literature.[5, 6] In the study of Thomas et al., 50% cases of lung aplasia were accompanied by congenital heart disease, and the prognosis of all four patients was good in the medium term, and there was no regular oxygen requirement.[2] In our case, along with lung aplasia, patent ductus arteriosus was detected initially and was closed after the medical management. The patient had a stable condition on full enteral feeds without any respiratory support after one week of hospitalization. These findings are similar to the study of Thomas et al.[2]

Conclusion

The manifestations of lung aplasia may be observed as early as in the first week of life. Hence, for term neonates with respiratory distress and complete lung opacity on chest x-ray, further imaging is required to evaluate such malformation. The index case of lung aplasia was associated with congenital heart disease and had a strong parental consanguinity. Therefore, in the pathogenesis of pulmonary aplasia, a genetic basis maybe implicated. Further analytic studies are needed to evaluate these issues. PDA was closed after the medical management; hence such care is highly

required. As neonatal sepsis accompanied the current case, the proper evaluation and management of infection is suggested for patients with lung aplasia.

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Conflict of Interest statement

The authors declare that they have no known financial conflicts of interest or close personal connections that might influence the findings of this study.

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Ethical Approval

This study was approved by the Department of Neonatology (Protocol no 5 dates 22/9/2022), Kabul University Medical Sciences.

Consent

Consent to publish the case report was obtained from the patients' guardians. The Helsinki Declaration was taken into consideration.

Guarantor

Mansoor Aslamzai

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