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Primary Congenital Pulmonary Hypoplasia

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Abstract: ***Primary congenital pulmonary hypoplasia is a rare but lethal disease found in newborns causing respiratory distress. Hypoplasia is said to be primary when it occurs in the absence of any fetal or maternal disorders affecting lung development. Here is a case report of a term male newborn presenting with respiratory distress requiring ventilatory support followed by prolonged oxygen dependency. X-rays were showing homogenous opacities in right mid and lower zone for which CT was done and it came out to be pulmonary hypoplasia. It can be diagnosed antenatally using ultrasonography and MRI.***

Keywords: Primary congenital pulmonary hypoplasia, alveoli, airway generation

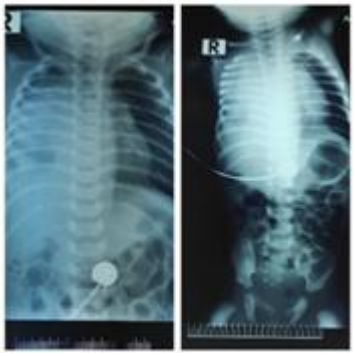
1. Introduction

Primary congenital pulmonary hypoplasia is a condition where there is decrease in the number of alveoli and airway generations unrelated to an underlying disorder. Usually, Pulmonary hypoplasia occurs commonly in association with congenital diaphragmatic hernia, oligohydramnios (mostly related to renal dysfunction), pleural effusions with fetal hydrops, skeletal dysplasias, malformations of the central nervous system and neuromuscular diseases, which affect the lung development. However, pulmonary hypoplasia may also be idiopathic or related to other syndromes and congenital anomalies - e.g., multiple pterygium syndrome/fetal akinesia-hypokinesia sequence (autosomal recessive) /scimitar syndrome and trisomy 21. Pulmonary hypoplasia is a rare but usually lethal disease. The incidence of congenital pulmonary hypoplasia may range from 9 to 11 per 10, 000 live births, and its mortality rate as reported by previous studies is 71 to 95%. We report a full-term male neonate who presented with respiratory distress on day two of illness.

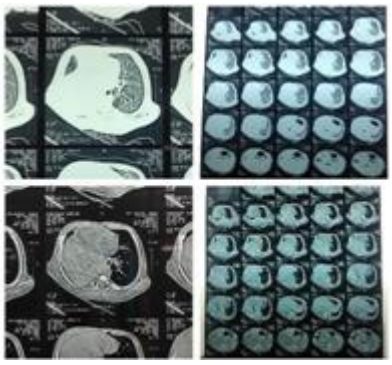
2. Case Report

A G3P1L1A1 mother with nil significant antenatal history delivered a male baby on 28.09.22 at 6.06 am at term gestation of 38 weeks two days with birth weight of 2.67 kgs through normal vaginal delivery. APGAR at 1 min was 8/10 and at 5 min was 9 /10. Inj. vitamin k was given. On day 2 of life, (28 hours of life) baby had respiratory distress, (air entry was decreased on right side), for which the baby was kept on CPAP. But in view of not maintaining saturation, on the same day baby was intubated and was on mechanical ventilation for 24 hours. Later settings were decreased and baby was extubated and kept on CPAP, with minimal settings. Initial repeat x-rays showed homogenous opacity in right mid and lower zone. But baby was having persistent respiratory distress. So on post natal day 18 CT chest was done which showed hypoplasia of the right lung and mild hyperinflation of left lung with no other congenital anomaly. Baby was kept on oxygen inhalation with prongs and is oxygen dependant since then.2D ECHO was done which showed mild TR with mild PAH, small OS-ASD, small right pulmonary artery. During the course of illness baby developed shock and sepsis, for which the baby was treated with antibiotics and inotropes and was weaned off.

At present baby is on oxygen inhalation with nasal prongs at 1L/min and on direct breastfeeding.



Chest XRAY



CT CHEST

3. Discussion

Usually babies with pulmonary hypoplasia present with respiratory distress or respiratory failure based on the severity of hypoplasia. The most important part in the treatment of congenital pulmonary hypoplasia is ventilatory support, HFOV, and even extracorporeal membrane oxygenation if needed. The prognosis depends on the size of the lungs if primary and the underlying cause if secondary and the development of pulmonary artery hypertension which has a bad prognosis.

In conclusion, primary congenital pulmonary hypoplasia should be highly suspected in neonates who present with immediate respiratory distress and small lung volume despite advanced respiratory support. USG and MRI (even antenatally), lung to body weight ratio, radial alveoli count are usually done to confirm the diagnosis. When they are not feasible or inconclusive CT can also be done. Mortality rate is high and even if they survive morbidity is more like chronic lung problems-e.g., reduced lung capacity, recurrent chest infections and impaired growth. So these babies after

discharge should have regular follow up. Severe pulmonary hypoplasia and pulmonary arterial hypertension are associated with reduced survival in babies with congenital diaphragmatic hernia and other associated anomalies. So 2D ECHO should be done for the babies at regular intervals.

Other differential diagnosis includes congenital pneumonia, early onset sepsis, congenital pulmonary airway malformation.

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