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Case Report

Paediatrics Section

Congenital Pulmonary Airway Malformation in a Two-month Old Infant: A Case Report

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ABSTRACT

Congenital Pulmonary Airway Malformations (CPAM) are rare clinical conditions that affect the lungs. They are characterised by the formation of large, multiloculated, cystic structures due to the overgrowth of the terminal bronchioles. This report presents the case of a two-month-old female infant who was referred to the pediatric emergency ward with a diagnosis of aspiration with collapse of the left lung. The patient exhibited progressively worsening respiratory distress accompanied by fever. The infant had a history of breast milk aspiration and had been admitted to the hospital multiple times due to respiratory distress since birth. Even after discharge, she required moist oxygen therapy at home. Upon presentation, a chest X-ray showed left lung collapse, and her respiratory rate was measured at 70 breaths/min. Other vital parameters and laboratory examinations were within normal limits. A high-resolution Computed Tomography (CT) scan of the thorax confirmed the diagnosis of CPAM. The patient was placed on ventilator support and received antibiotic treatment. Once stabilised, she underwent right postero-lateral thoracotomy with right middle and lower lobectomy under general anesthesia. Following surgery, she was discharged in stable condition on the seventh postoperative day, with her respiratory distress resolved. The child continues to receive regular follow-up care, including immunisations and appropriate nutritional support.

Keywords: Bronchioles, Respiratory distress, Thoracotomy, Ventilator

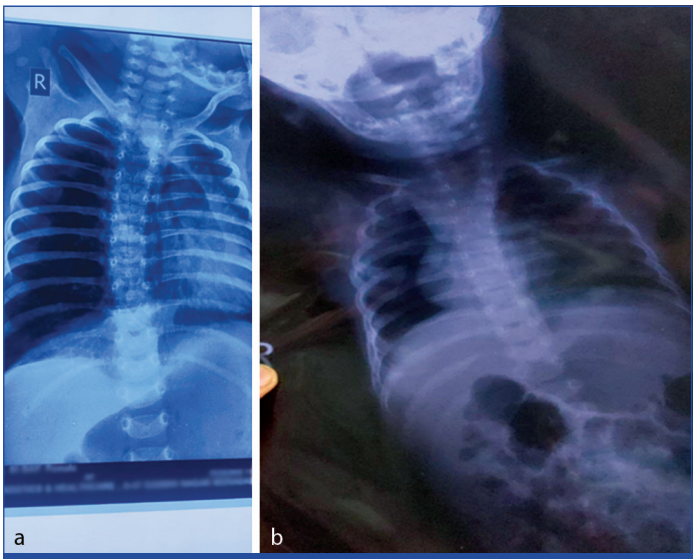
CASE REPORT

A two-month-old female infant was brought to the pediatric emergency ward of a tertiary care institute in Uttarakhand, India, with a chief complaint of persistent respiratory distress over the past six days, which had been gradually worsening. The respiratory distress was accompanied by low-grade fever (100°F). There was no significant antenatal history.

The infant was born at term via spontaneous vaginal delivery, weighing 2.465 kg at birth. Shortly after birth, she developed neonatal hyperbilirubinemia and was admitted to the Neonatal Intensive Care Unit (NICU) for two days. Four days after discharge, she presented with cyanosis, excessive crying, and episodes of up-rolling of the eyes. She was diagnosed with aspirational pneumonia and received Continuous Positive Airway Pressure (CPAP) ventilation in the NICU for two days. After seven days of hospitalisation, the baby was discharged in stable condition with home oxygen support. However, one month later, she experienced cyanosis again and was admitted to a local private hospital with a diagnosis of aspirational pneumonia. She stayed in the hospital for five days and was discharged with oxygen support.

For the next 25 days, the infant appeared well. However, she then developed low-grade fever and persistent respiratory distress, which gradually worsened. She was taken to a local hospital, where she was diagnosed with pneumonia and left lung collapse. Subsequently, she was referred to our institution with a prescription for antibiotics {injection (inj.) Piperacillin-Tazobactam 270 mg TDS} and moist oxygen support (2 L/min via nasal prongs).

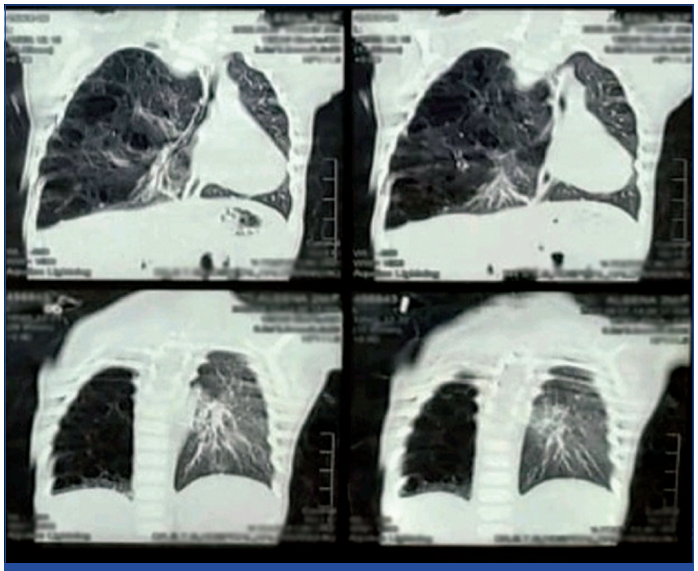
Upon admission, the child had no fever, a heart rate of 108 bpm, respiratory rate of 70/min, and SpO_2 of 100% on moist oxygen support (6 L/min). Breath sounds were positive on the right side of the chest but absent on the left side. A chest X-ray revealed collapsed left lung [Table/Fig-1]. Laboratory examinations showed a hemoglobin level of 8.3 g/dL, total leukocyte count of 3700/cc, and a platelet count of 3.5 lakh/cc. Additionally, serum glucose level was 73 mg/dL, urea 14 mg/dL, creatinine 0.3 mg/dL, total bilirubin 0.7 mg/dL, and alkaline phosphatase 404 IU/L. Serum levels of sodium, potassium, and ionised calcium were 132 mg/dL, 5.5 mg/dL, and 1.1 mg/dL, respectively.



[Table/Fig-1]: a) Posteroanterior view of the chest skiagram of the patient; b) Chest X-ray shows overinflated right lung with mediastinal shift to the left. Hyperlucent right hemithorax with contralateral mediastinal shift, with collapse of left lung.

A provisional diagnosis of aspirational pneumonia with left lung collapse was made. Due to severe respiratory distress, the patient was placed on CPAP ventilation with Fraction of inspired oxygen (FiO_2) 50%. She was advised nothing per oral and prescribed intravenous antibiotics (injection piperacillin-tazobactam 270 mg TDS) and amikacin (intravenous 40 mg diluted in normal saline over 20 minutes OD). The baby also received intravenous fluid regimen of second-instar nymphs on day 5 (N2D5) at a rate of 70 mL over six hours and salbutamol nebulisation every four hours. A high-resolution computed tomography scan of the thorax revealed diffuse hyperinflation with emphysematous changes in the right lung field, along with the attenuation of vascular structures and multiple air-filled cystic spaces (0.7×0.7 cm) in the right middle and lower lobes. The scan also showed retrosternal herniation, mediastinal shift to

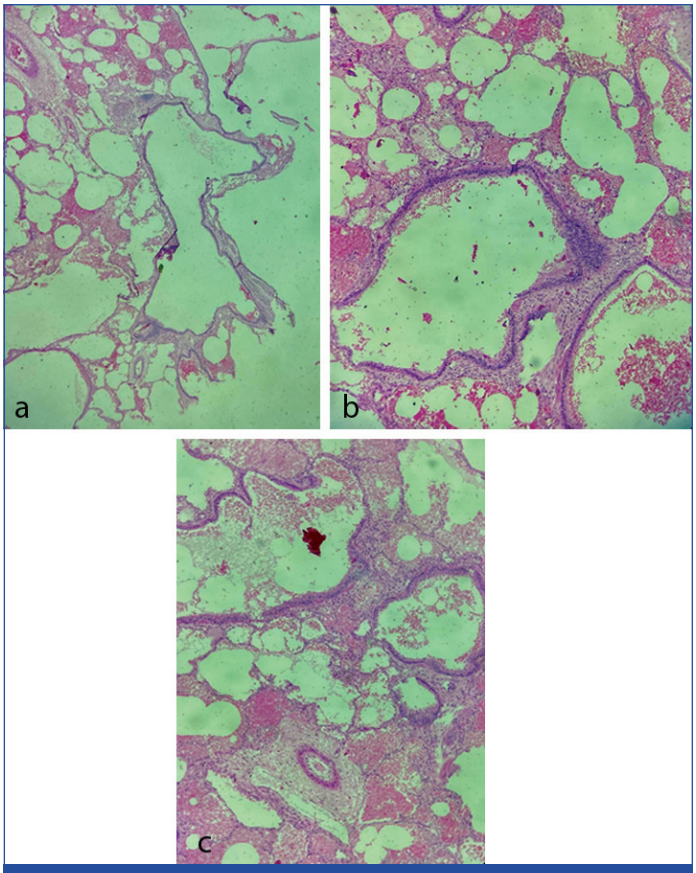
the left, and collapse of the left lung [Table/Fig-2]. These findings were consistent with the diagnosis of congenital lung malformation, specifically CPAM. The patient's treatment continued as per the earlier regimen, and her respiratory distress subsided by the third day of admission, at which point she was referred to a paediatric thoracic surgeon.



[Table/Fig-2]: High-resolution computed tomographic scan of the thorax showing multiple cystic lesions in the right middle and lower lung lobes of the patient.

The patient underwent right posterolateral thoracotomy with right middle and lower lobectomy under general anesthesia. From postoperative day 0, the infant was resumed on oral feeding via nasogastric tube and remained on oxygen support. One unit of packed red blood cells was transfused on postoperative day 1 without any adverse reactions. The intercostal drain and oxygen support were removed on day 7, and the child was discharged with stable vitals, no respiratory distress, fully tolerating oral breastfeeding, and with a SpO_2 of 98% on room air. The thoracotomy wound had healed by the time of discharge. Unfortunately, further follow-up of the patient could not be conducted at the study institution.

The histopathology of the resected lung tissue confirmed the presence of multiple variable-sized cysts lined by adenomatous columnar epithelium with areas of hemorrhage and congestion, consistent with CPAM [Table/Fig-3a-c].



[Table/Fig-3]: a) Multiple sized cysts lined by columnar epithelium with congestion, (20x, H&E stain); b) Cystically dilated bronchioles (40x, H&E); c) Cystically dilated bronchioles filed with haemorrhagic fluid, (40x, H&E stain).

DISCUSSION

CPAMs result from abnormalities in bronchiole branching and development. It is believed that an imbalance between cell proliferation and apoptosis during organogenesis contributes to the development of CPAM [1]. Pathologically, CPAM is classified into five types: Type 0 (acinar dysplasia), Type 1 (macrocytic, lined by pseudostratified epithelium, good prognosis), Type 2 (microcystic, poor prognosis), Type 3 (cuboidal epithelium, poor prognosis), and Type 4 (adults, macrocysts) [2]. In most cases, symptoms of CPAM present immediately after birth or during the neonatal period, as seen in the patient in this study.

Respiratory distress is the most common presentation, occurring in approximately 80% of cases [3]. Other case reports notably by Chan IC et al., and Sime H et al., have also noted significant respiratory distress as the presenting complaint [3,4]. Similarly, the patient in this study presented with persistent and worsening respiratory distress.

In well-equipped healthcare institutions, CPAM is often diagnosed during antenatal ultrasonography [5-7]. However, this was not the case for the patient in this report. Possible reasons for this could include poor resolution of the ultrasound equipment, lack of expertise of the sonographer, or the ultrasound being performed at a later stage of pregnancy when the sensitivity to detect CPAM decreases [8]. Additionally, the diagnosis of CPAM in this case was further complicated by multiple episodes of breastmilk aspiration pneumonia. This delayed the provisional diagnosis of CPAM after birth. However, it is also possible that the presence of the lesion itself contributed to the multiple episodes of milk aspiration.

In symptomatic cases like the infant in this report, surgical management is recommended with a good prognosis. The patient experienced significant relief of respiratory distress and improved after undergoing right middle and lower lobectomy to remove the malformed lung tissue.

CONCLUSION(S)

In conclusion, this case highlights the importance of considering CPAM in infants with multiple episodes of postnatal respiratory distress that do not respond to conventional treatment. If CT thorax findings are consistent with the diagnosis, thoracotomy should be considered as the management of choice for these infants.

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