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Case Report

Congenital pulmonary airway malformation presenting with neonatal respiratory distress

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ABSTRACT

Congenital pulmonary airway malformation (CPAM) is a developmental lung abnormality that may present with respiratory distress in the neonatal period. Assessment requires correlation of clinical findings with the lesion's anatomy and effect on ventilation. A six-day-old female neonate had tachypnea and subcostal retractions beginning soon after birth. She was born at 40 weeks by normal vaginal delivery with a birth weight of 2.3 kg. Her respiratory rate was 78 breaths/min, heart rate 148 beats/min and oxygen saturation 94% while receiving oxygen through continuous positive airway pressure (CPAP). Examination revealed reduced left-sided chest movement, mild rightward tracheal deviation and diminished air entry over the left lower zone. Computed tomography showed multiple cystic air spaces in the left lung, compatible with CPAM. The complete blood count was normal, C-reactive protein was negative and blood culture was sterile. Respiratory distress improved with CPAP, oxygen, intravenous fluids and intravenous antibiotics. A congenital lung lesion should be considered when neonatal respiratory distress is accompanied by focal chest findings. Initial clinical improvement with supportive care requires subsequent assessment to plan surveillance or resection according to symptoms and lesion anatomy.

Keywords: Congenital pulmonary airway malformation, Congenital cystic adenomatoid malformation, Neonate, Respiratory distress, Continuous positive airway pressure

INTRODUCTION

Congenital pulmonary airway malformation (CPAM), previously termed congenital cystic adenomatoid malformation, comprises developmental abnormalities of the distal airways and lung parenchyma. The lesions contain cystic or apparently solid components with abnormal airway architecture; their morphological variability forms the basis of pathological classification.[1,2] The term CPAM accommodates this broader spectrum more accurately than the older designation, because all lesions do not share an adenomatoid appearance.[2]

The clinical spectrum extends from an antenatally detected, asymptomatic lesion to respiratory compromise in the neonatal period. In a symptomatic newborn, the important questions concern the lesion's extent, its effect on adjacent lung and its vascular supply. Prenatal and postnatal imaging can identify a congenital lung malformation, but imaging labels do not always correspond to the final pathological diagnosis; CPAM, bronchopulmonary sequestration and hybrid lesions may overlap.[3]

This report describes a term female neonate with respiratory distress beginning soon after birth, a cystic left pulmonary lesion and initial improvement with conservative treatment. It highlights the role of focal examination findings and imaging in diagnosis, and the distinction between short-term stabilization and subsequent management of the lesion.

CASE REPORT

A six-day-old female neonate was evaluated for fast breathing and subcostal retractions that had begun soon after birth. She was born at 40 weeks of gestation by normal vaginal delivery, had a birth weight of 2.3 kg and cried immediately after birth. The maternal history included hypothyroidism treated with medication for six months, which the mother had discontinued herself during pregnancy. The family history was noncontributory.

On examination, the respiratory rate was 78 breaths/min, heart rate was 148 beats/min and oxygen saturation was 94% while she was receiving oxygen through continuous positive airway pressure (CPAP). Tachypnea and subcostal retractions were present. The chest was symmetrical, with reduced movement on the left side, mild rightward tracheal deviation and diminished air entry over the left lower zone. The first and second heart sounds were normal, with no murmur. The abdomen was soft and nontender, and neurological examination showed normal tone and reflexes.

The complete blood count was normal, C-reactive protein was negative and blood culture was sterile. Liver and renal function tests were normal, and random blood glucose was 147 mg/dL. Chest ultrasonography and echocardiography were advised; their results were not documented.

Chest radiography and computed tomography formed part of the assessment (Figure 1). Computed tomography demonstrated multiple cystic air spaces within the left lung, compatible with a congenital pulmonary airway malformation. A histopathological subtype was not established. Initial treatment consisted of oxygen through CPAP, intravenous fluids and intravenous antibiotics. The respiratory distress improved with this conservative management.

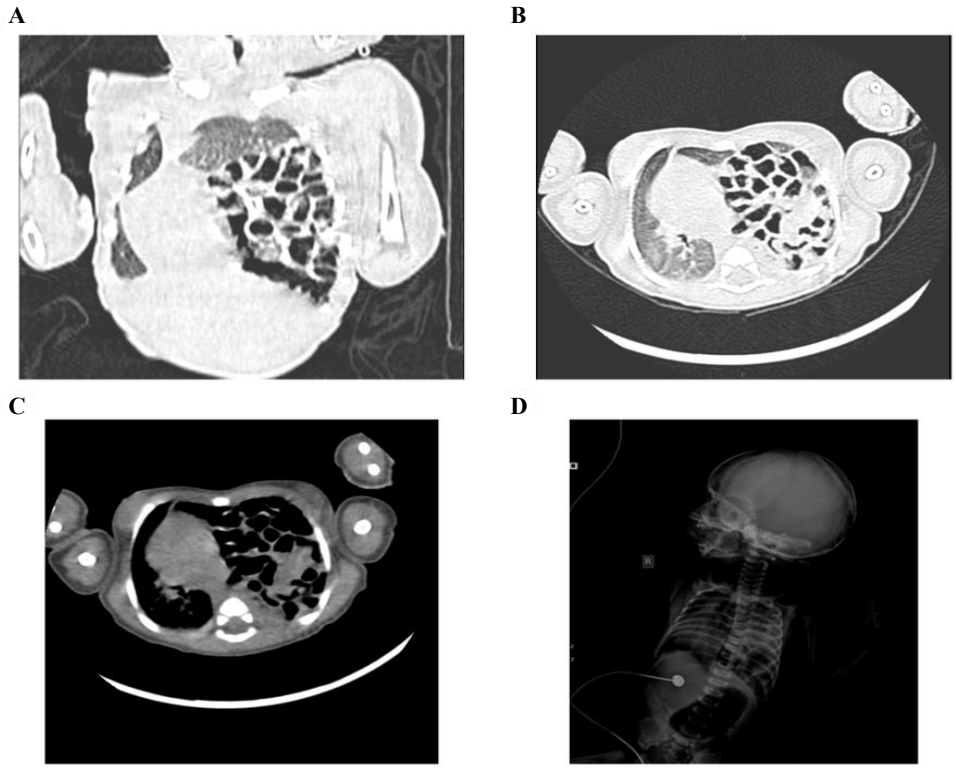


Figure 1: Imaging during the neonatal assessment (A) Coronal and (B and C) axial chest computed tomography images showing multiple cystic air spaces in the left lung (D) Chest radiograph obtained during the same assessment.

DISCUSSION

The focal respiratory findings in this neonate supported a structural pulmonary cause for the respiratory distress. Reduced air entry over the left lower zone and rightward tracheal deviation were consistent with the cystic left pulmonary lesion seen on computed tomography. Expanding cystic lesions can impair ventilation by compressing adjacent lung and displacing mediastinal structures.[1,3] The normal complete blood count, negative C-reactive protein and sterile blood culture provided no laboratory evidence of infection in the reported assessment. The clinical and imaging findings therefore supported CPAM as the explanation for the presentation.

Pathological classification and imaging descriptions should be distinguished. In Stocker's original morphological series, type 1 lesions contained relatively

large cysts, type 2 lesions contained multiple smaller bronchiolar-like cysts, and type 3 lesions were bulky with predominantly microscopic airway spaces.[1] Consequently, a radiological description such as macrocystic does not establish a type 2 pathological diagnosis. A cyst-size label alone is insufficient for subtyping; the epithelial lining, intervening tissue and the distribution of abnormalities must also be assessed.[1,2] As no histopathological findings were available in this case, the diagnosis remains based on the clinical presentation and imaging, without assignment of a definitive pathological subtype.

Antenatal detection allows serial assessment of lesion growth, mediastinal displacement and hydrops, with counselling and delivery planning at an appropriate centre. The CPAM volume ratio was developed to relate ultrasound-estimated lesion volume to fetal head circumference. In the original prospective study, larger ratios were associated with a higher risk of hydrops, although lesion morphology and a dominant cyst also influenced the course.[4] Prenatal risk assessment and postnatal clinical assessment therefore address related but different questions; neither an antenatal label nor apparent antenatal regression establishes the final anatomical diagnosis.

Postnatal cross-sectional imaging is important for defining the affected lobe, cystic architecture, remaining lung and vascular connections. Demonstration of an aberrant systemic feeding artery suggests sequestration or a hybrid lesion and has direct implications for operative planning. In a surgical cohort comparing imaging with pathology, CPAM, sequestration and hybrid lesions all contributed substantially to the final diagnostic spectrum, illustrating the limits of assigning a specific malformation from a single imaging modality.[3] The differential also includes congenital lobar overinflation, bronchogenic cyst and congenital diaphragmatic hernia, which are distinguished by their anatomical features.

Management of symptomatic neonatal CPAM begins with support of oxygenation and ventilation and paediatric surgical evaluation. Persistent respiratory compromise or substantial mass effect generally favours resection once the infant has been stabilized. The American Pediatric Surgical Association evidence review identifies lobectomy as the standard resection method, performed thoracoscopically or through thoracotomy according to the infant's condition, lesion anatomy and surgical expertise.[5] In the present case, CPAP, oxygen and supportive treatment were associated with initial improvement in respiratory distress. This response provides a basis for reassessment after stabilization; decisions about further imaging, surveillance and surgery require review of the lesion's anatomy and the subsequent respiratory course.

The management of an asymptomatic lesion remains debated. In a systematic review and meta-analysis, children who underwent surgery after developing symptoms had greater postoperative morbidity than those who underwent elective surgery before symptoms developed. However, the included studies were retrospective and subject to selection bias.[6] These findings support discussion of elective surgery and surveillance with the family, but do not establish a universal timing strategy. Evidence from asymptomatic cohorts is only partly applicable to an infant who has already required respiratory support, so subsequent planning should take the initial presentation into account.

Perioperative risk reflects the infant's clinical status as well as operative timing. In a multicentre surgical database study, preoperative oxygen dependence or ventilatory support was independently associated with increased morbidity. The study did not find a significant difference in morbidity between asymptomatic neonates and asymptomatic infants operated on later in the first year.[7] This distinction supports individualized planning and cautions against attributing risk to neonatal age alone. Follow-up should assess respiratory symptoms, feeding and growth, with further pulmonary evaluation when clinically indicated. Longer-term observations were not available for this infant, so the durability of the initial improvement cannot be assessed.

Histopathological examination also addresses important diagnostic alternatives. Type I pleuropulmonary blastoma can resemble a benign cystic lung malformation. A comparative study identified clinical and radiological features that favour CPAM or pleuropulmonary blastoma, but their overlap means that imaging cannot replace pathological assessment in a resected lesion.8 The possibility of diagnostic overlap should be separated from an assumption that every CPAM undergoes malignant transformation. Suspicious features or relevant personal or family history may warrant specialist assessment for a DICER1-associated disorder.[8] This case illustrates the value of linking the bedside examination to imaging when evaluating persistent neonatal respiratory distress. Maternal hypothyroidism is part of the antenatal history; the available information does not establish it as the cause of the lung lesion. The principal limitations are the absence of histopathological confirmation, precise treatment duration and longer-term outcome data. The documented improvement therefore describes initial stabilization with conservative care rather than a confirmed long-term outcome.

CONCLUSION

CPAM should be considered in a neonate with early respiratory distress, asymmetric air entry and tracheal deviation. In this infant, computed tomography supported the diagnosis and respiratory distress improved with CPAP, oxygen and supportive treatment. Subsequent neonatal and paediatric surgical assessment, with follow-up of respiratory health and growth, is needed to guide surveillance or resection.

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