Case Report

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Pathology Section

Congenital Pulmonary Airway Malformation, Stocker Type I: A Case Report

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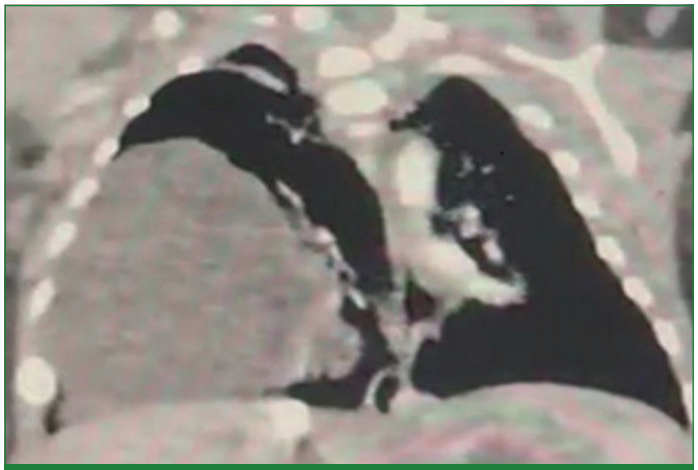
ABSTRACT

Congenital Pulmonary Airway Malformation (CPAM), formerly known as Congenital Cystic Adenomatoid Malformation (CCAM), is a rare developmental dysplastic lesion of the foetal tracheobronchial tree and a rare cause of neonatal respiratory distress. Here, authors present a case of term, male newborn baby, who was antenatally diagnosed to have cystic lesion in right lung. Soon after birth, baby developed respiratory distress in the form of subcostal retractions for which Contrast-Enhanced Computed Tomography (CECT) was done and it showed a well-defined hypodense lesion occupying the right lung predominantly the middle and lower lobe. At the age of one month and 22 day, right posterolateral thoracotomy was performed, and the specimen was histopathologically diagnosed as CPAM type I. Frequent Respiratory Tract Infections (RTI) are major concerns in these patients. When a child having recurrent episodes of RTI, CPAM can be considered as one of the underlying causes. Malignancy and frequent airway infections are major concerns in these patients. Surgical excision is recommended to make a definite diagnosis and exclude hidden malignancies and is also the treatment of choice. CPAM is rare for its presentation, early diagnosis and treatment favours good prognosis. The present case report emphasises the importance of considering CPAM in the differential diagnosis of neonatal respiratory distress and recurrent childhood RTIs, highlighting the role of imaging and histopathological findings in achieving a definitive diagnosis.

Keywords: Adenomatoid malformation, Developmental anomaly, Macrocysts, Neonatal respiratory distress

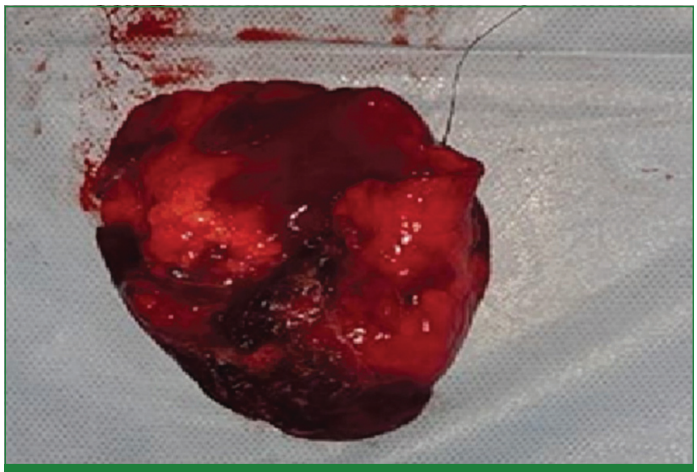
CASE REPORT

A term, male newborn baby antenatally diagnosed to have cystic lesion in right lung developed respiratory distress soon after birth, in the form of subcostal retractions with Downes' score of 2/10. Chest X-ray revealed a homogeneous opacity in the right hemithorax involving middle and lower lung field. CECT thorax showed a well-defined hypodense lesion involving middle and lower lobe of right lung with a differential diagnosis of duplication cyst or CPAM was reported [Table/Fig-1]. Earlier antenatal scan at 28 weeks showed right lung macrocyst. At 31 and 34 weeks scan, CPAM Volume Ratio (CVR) was 1.46 cm^2 (cyst size 3.9×4.5×5.5 cm) and 1.49 cm^2, respectively. No hydrops was identified. Right lower lobectomy of lung was performed at the age of 52 days based on the present complaints and imaging studies.



[Table/Fig-1]: CECT well-defined cystic lesion involving right middle and lower lobe.

The histopathological analysis was performed. The specimen had grey brown bosselated appearance measuring 4×4 cm. Cut surface showed multiloculated 4×2 cm cyst filled with blood. Lung parenchyma surrounding the cyst and hilar structures appeared normal [Table/Fig-2]. Under microscopy, variably sized cysts were seen with some having papillary projections [Table/Fig-3a]. Small cysts were lined by ciliated cuboidal to pseudostratified columnar epithelium with interspersed alveolar type spaces. Large cysts were lined by flattened cuboidal epithelium with areas of haemorrhage and congestion [Table/Fig-3b]. Adjacent alveoli showed haemorrhage and congestion [Table/Fig-3c]. Hilar structures were normal. It was diagnosed as CPAM, Stocker type I. Postoperative recovery was supported with regular clinical follow-up and imaging, ensuring satisfactory lung expansion and favourable long-term prognosis.

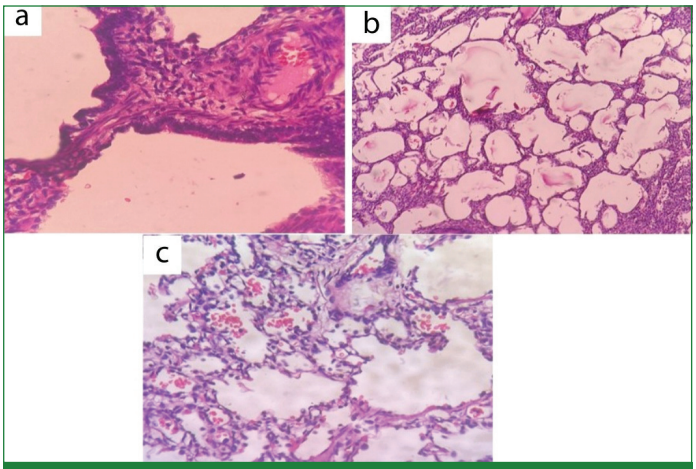


[Table/Fig-2]: Lobectomy showing bosselated surface.

DISCUSSION

The CPAM were first identified in 1949 by Ch'in KY and Tang MY as distinct and rare lesions in stillborn or premature newborns with anasarca [1]. CPAM, earlier termed as CCAM, are the most common form of congenital parenchymal lung malformations and have an estimated incidence of 1:25,000-1:35,000 births [2]. About 15-

50% of cases of congenital cystic lung diseases are reported to be CPAM [3]. Majority of CPAM lesions are asymptomatic; however, if symptomatic, they typically present with recurrent infections and/or respiratory distress. Kirsten Rat Sarcoma Viral Oncogene Homolog (KRAS) mutations are confirmed in CPAM, with other tumour-related genetic factors under study [3].



[Table/Fig-3]: a) Cystic spaces lined by flattened cells H&E (4X); b) Cystic spaces lined by columnar epithelium H&E (40X); c) Adjacent alveoli with haemorrhage and congestion H&E (10x).

The CPAM can present as an incidental lesion, remain undetected during early life, and later be complicated by mucinous adenocarcinoma due to the presence of mucinous cell clusters and KRAS mutations within the lesion. Respiratory distress remains the most common clinical manifestation of symptomatic CPAM in the neonatal period, especially when the lesion is large enough to produce mass effect and mediastinal shift [4]. Histologically, CPAM is divided into large-cyst and small-cyst variants. Large-cyst (macrocystic) CPAM shows unilocular or multilocular cysts greater than 2 cm lined by ciliated respiratory epithelium with frequent mucinous cells, corresponding to Stocker type I. Small-cyst (microcystic) CPAM demonstrates compact irregular microcystic spaces and bronchiole-like tubular structures, with mucinous epithelium less commonly seen [3,5].

The modified Stocker classification (types 0-4) provides further refinement. Type 0 involves the tracheobronchial region, affects all lobes and is typically incompatible with life. Type I is the most common, characterised by single or multiple large cysts lined by ciliated pseudostratified epithelium with mucinous cells and cartilage [3]. Type II arises from terminal bronchioles and presents as multiple small cysts giving a sponge-like appearance. Type III is a solid-appearing microcystic lesion arising from distal bronchioles and alveolar ducts, usually presenting in utero or at birth with severe respiratory distress. Type IV originates from the distal acinar region and presents as large peripheral thin-walled cysts associated with pleuropulmonary blastoma [6].

The differential diagnosis of congenital cystic lung lesions includes pulmonary sequestration, bronchogenic cyst, and Congenital Lobar Emphysema (CLE). Pulmonary sequestration receives systemic arterial supply from the aorta and lacks communication with the tracheobronchial tree [5]. Bronchogenic cysts typically show ciliated pseudostratified columnar epithelial lining with cartilage, bronchial glands, smooth muscle, and neural elements. CLE results from defective bronchial cartilage and shows hyperinflated alveoli without discrete cyst formation [5].

In developed settings, most CPAMs are diagnosed during routine mid-trimester anomaly ultrasonography. Prognostication during foetal evaluation is often guided by the CVR, where high values are associated with increased risk of hydrops, and lower initial CVR values predict more favourable outcomes [2,7]. Postnatal imaging with chest X-ray and Computed Tomography/Magnetic Resonance Imaging (CT/MRI) is essential for accurate mapping of the lesion and surgical planning. Surgical excision (often lobectomy) remains the gold standard of management both for symptom control and prevention of long-term complications such as recurrent infections and malignant transformation [4].

When compared with previously published neonatal cases of CPAM type I, the present case demonstrates several similarities and distinctive features. Like most published reports, the lesion in our patient was detected antenatally, respiratory distress developed shortly after birth, and definitive management was achieved through lobar resection with histopathological confirmation of type I CPAM [8]. However, unlike reports complicated by pneumothorax, hydrops or emergency ventilation requiring immediate surgery, the present case involved a predominantly middle and lower lobe lesion on the right-side that remained clinically stable until elective surgical excision at one month and 22 days of age. The smooth postoperative recovery and favourable outcome further reinforce existing literature that early recognition, careful perinatal monitoring and timely surgical intervention offer excellent prognosis in CPAM type I and successfully prevent long-term infectious and malignant sequelae [9].

CONCLUSION(S)

The CPAM is a rare developmental anomaly of the lung that presents with a spectrum of clinical manifestations. Early detection and prompt management are vital in ensuring optimal outcomes for affected individuals. Surgical resection remains the mainstay of treatment for symptomatic cases, with a good prognosis expected following complete excision. Long-term follow-up is essential to monitor for potential complications and to ensure continued respiratory health. Further research and case studies are warranted to enhance our understanding of CPAM and improve the management of this congenital lung disorder.

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