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PULMONARY CYSTIC ADENOMATOID MALFORMATIONS IN CHILDREN: A 10-YEAR RETROSPECTIVE STUDY AND LITERATURE REVIEW

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ABSTRACT

Background: Congenital pulmonary airway malformation (CPAM) is a rare developmental anomaly. While antenatal imaging improves detection, optimal postnatal management remains debated.
Methods: We retrospectively reviewed 9 pediatric patients who underwent surgical treatment for CPAM between 2014 and 2024. Data included demographics, clinical presentation, imaging, histopathology, surgical approach, and outcomes.
Results: Two cases were diagnosed antenatally. The most common presentation was recurrent infection. All patients underwent thoracotomy and resection. Histopathology showed a predominance of type I CPAM. No mortality was reported.
Conclusions: CPAM requires early diagnosis and planned surgical intervention to prevent complications. Antenatal screening and postnatal follow-up are essential. Outcomes after surgery are excellent.

KEYWORDS

Pulmonary Cystic Adenomatoid children Congenital pulmonary airway malformation

MAIN ARTICLE

Introduction

Congenital pulmonary airway malformations (CPAMs), historically termed congenital cystic adenomatoid malformations (CCAMs), are rare anomalies of lung development characterized by abnormal bronchopulmonary tissue. These lesions represent approximately 25% of congenital lung malformations and occur in 1 per 25,000 to 35,000 live births.

CPAMs are typically unilateral and confined to a single lobe. They result from aberrant airway branching between the 5th and 17th weeks of gestation, leading to non-functioning lung tissue composed of cysts and abnormal airways. Stocker's classification divides CPAMs into five types based on histopathological features, with type I being the most common.

While many cases are now diagnosed prenatally via obstetric ultrasound, some may present postnatally with respiratory distress, recurrent infections, or remain asymptomatic. The optimal timing and approach to surgical resection remain under debate, particularly in asymptomatic patients.

In this study, we report a 10-year retrospective series of CPAM cases managed at our institution and provide a comprehensive review of the literature, focusing on diagnosis, treatment strategies, and outcomes.

Materials and Methods :

This is a retrospective, descriptive, single-center study conducted at the Pediatric Surgery Department of the Children's Hospital of Rabat (CHU Ibn Sina, Morocco). We reviewed the medical records of all children operated on for congenital pulmonary airway malformation (CPAM) between January 2014 and December 2023.

Inclusion Criteria:

All patients under 15 years of age who underwent surgical resection of CPAM confirmed by histopathological examination were included. Patients with incomplete records or uncertain diagnoses were excluded.

Data Collection:

For each patient, we collected the following variables:

- Demographic data: age, sex, and gestational age at birth
- Prenatal data: gestational age at diagnosis, type of anomaly suspected on antenatal ultrasound, and follow-up imaging
- Clinical presentation: respiratory distress, recurrent infections, or incidental discovery
- Radiological findings: prenatal and postnatal imaging (chest X-ray, thoracic ultrasound, CT scan)
- Surgical details: approach (thoracotomy or thoracoscopy), type of resection (lobectomy, segmentectomy), and intraoperative findings
- Postoperative outcomes: hospital stay, complications, need for intensive care, and duration of follow-up
- Histopathological classification: based on Stocker's system (type I to V)

Surgical Strategy:

All patients were operated on by experienced pediatric surgeons. The surgical technique was chosen based on the lesion's location, size, and symptoms. Most procedures were performed via posterolateral thoracotomy. When feasible, parenchyma-sparing resections were preferred. Chest drains were placed in all cases and removed based on clinical and radiological criteria.

Data Analysis:

All data were entered into a Microsoft Excel spreadsheet and analyzed descriptively. Given the small sample size, no statistical tests were applied. Outcomes were reported as absolute values or percentages.

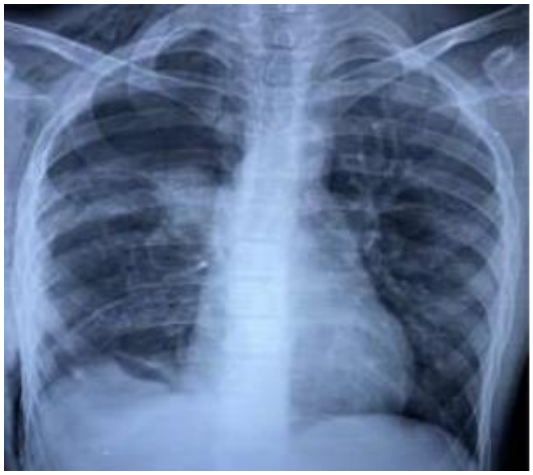


Figure 1:Standard chest radiograph of a 15-years-old child : We observe an area of increased radiolucency in the right upper lobe, suggestive of a giant lobar emphysema, which could be mistaken for a complete pneumothorax.

In the right middle and lower lobes, the radiographic image shows a pattern reminiscent of a "cluster of grapes," characteristic of bronchiectasis.

The right diaphragmatic dome appears altered and slightly elevated.

Between the right upper and middle lobes, an ovoid water-density opacity with irregular borders is visible, forming an acute angle with the mediastinum, which may correspond to an anomaly or mass requiring further investigation.

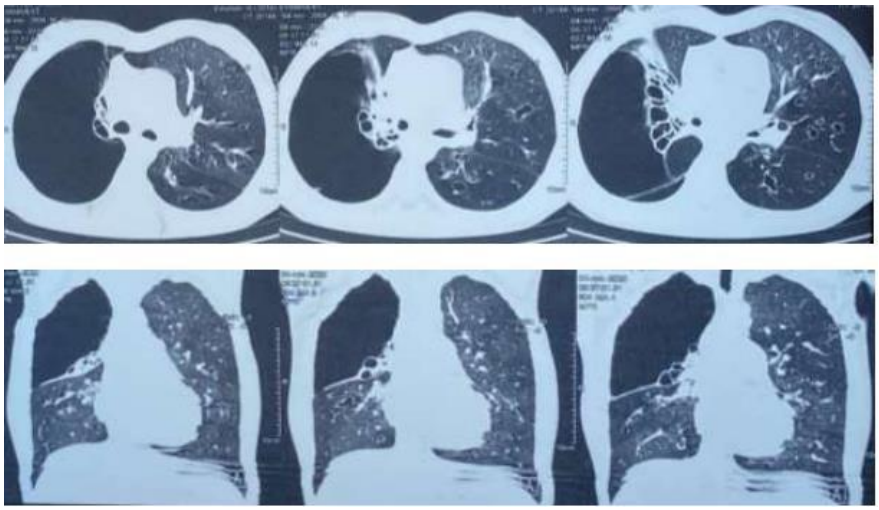


Figure 2:Chest CT scan of the same patient as in Figure 1. The right upper lobe shows a hyperlucent area consistent with giant lobar emphysema, which may mimic a complete pneumothorax. The middle and lower lobes display a "grape-like" cystic pattern characteristic of bronchiectasis. The right diaphragmatic dome is slightly elevated and deformed. Between the right upper and middle lobes, an ovoid, fluid-density lesion with irregular margins forms an acute angle with the mediastinum, suggesting a potential malformation or mass requiring further investigation.

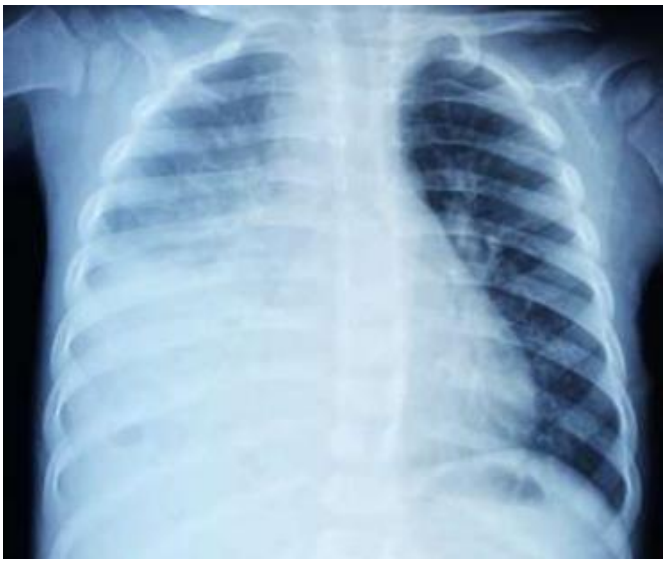


Figure 3:Standard chest radiograph of a 2-year-old child showing a homogeneous, well-defined opacity occupying the lower two-thirds of the right lung, associated with an air bronchogram. This appearance is suggestive of an acute lobar pneumonia (ALP).

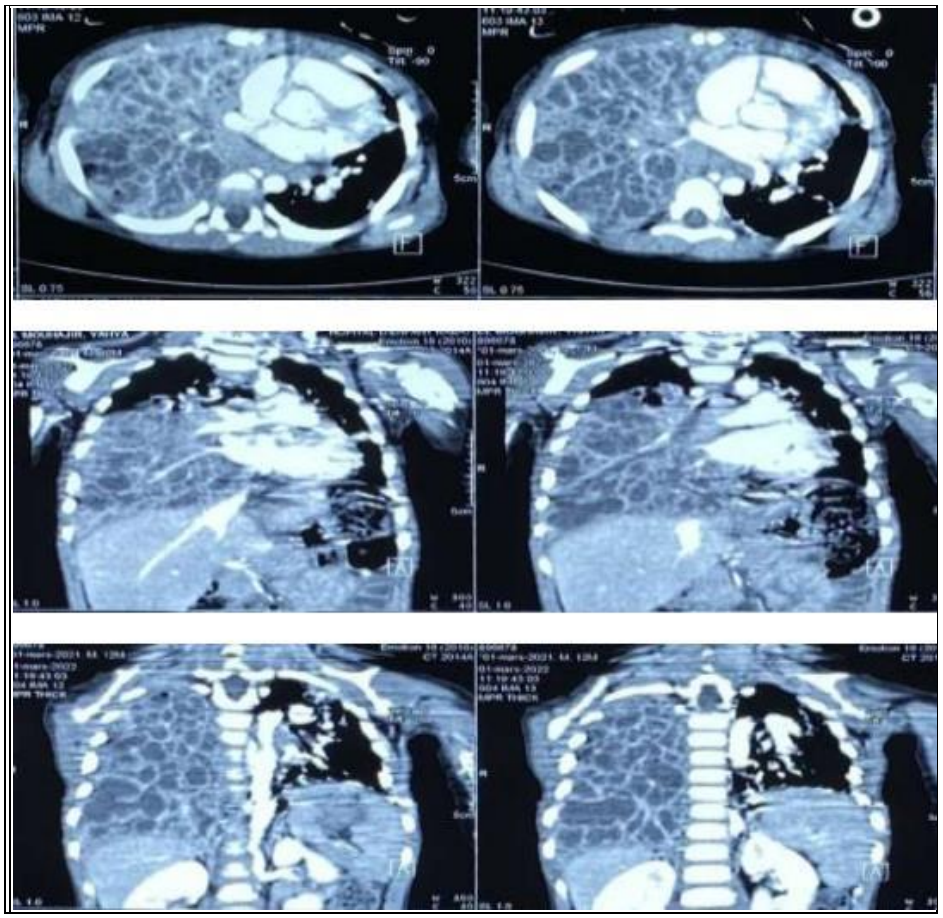


Figure 4:Chest CT scan of the same patient as in Figure 3, showing a multiloculated cystic formation involving the entire right lung. Both micro- and macrocysts are present, with thickened septated walls and fluid content. Contrast enhancement suggests secondary

infection. There is no evidence of ventilation impairment. A mediastinal shift is noted. These findings are highly suggestive of a type II congenital pulmonary airway malformation (CPAM), although the possibility of malignant transformation cannot be excluded.

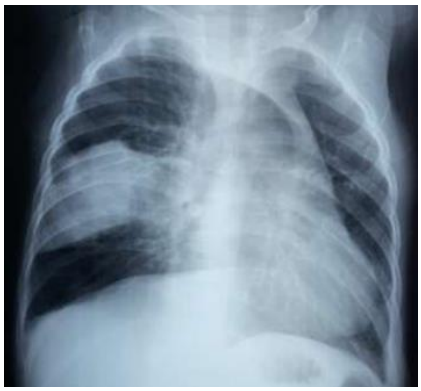


Figure 5.Standard chest radiograph of a 15-month-old patient showing a well-circumscribed, water-density opacity localized to the right middle lobe, without visible air bronchogram. The lesion is complicated by a mediastinal shift toward the left hemithorax and elevation of the right diaphragmatic dome.

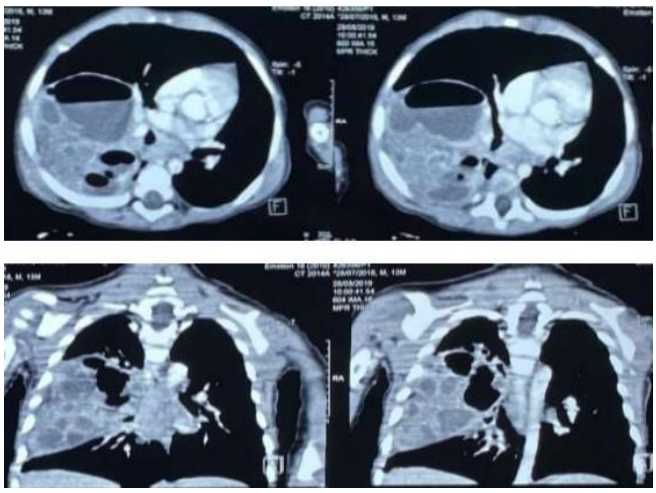
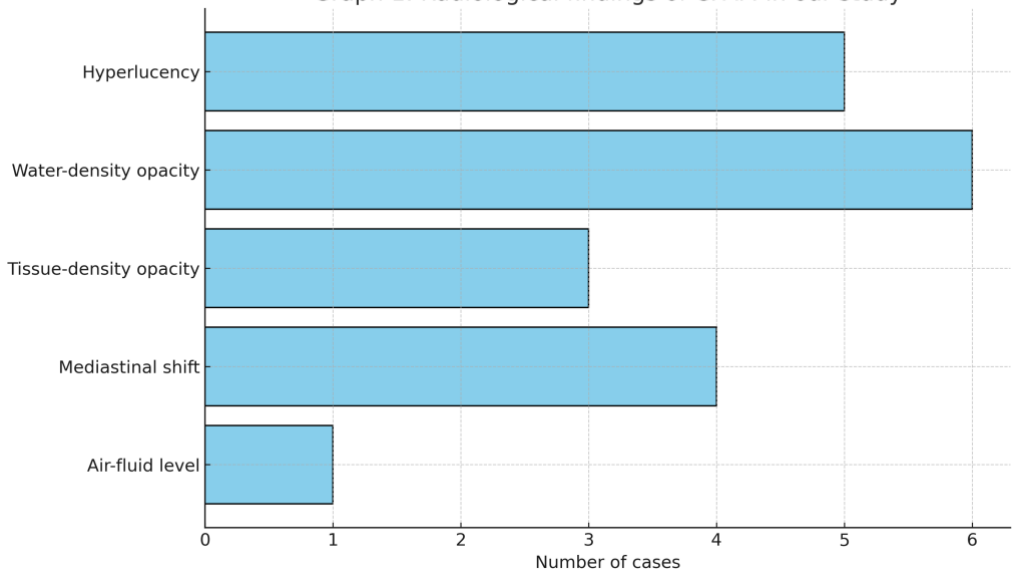


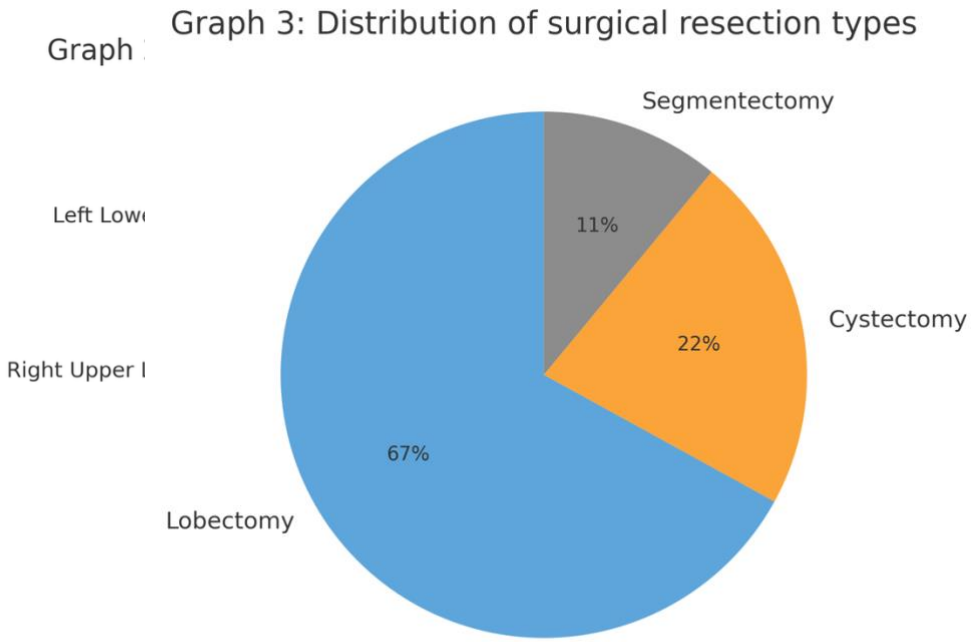
Figure 6.Chest CT scan of the same 15-month-old patient as in Figure 5, showing a multiloculated intraparenchymal cystic formation in the right middle lobe. The cysts are of variable size, with thickened walls enhancing after contrast administration. Air-fluid levels are visible within several cysts. These findings are highly suggestive of a superinfected type II congenital pulmonary airway malformation (CPAM).

Results

A total of nine patients were included (5 males, 4 females). The median age at diagnosis was 9 months. Two cases were diagnosed antenatally. Seven children presented with respiratory distress or recurrent infections. All underwent surgical resection via thoracotomy. Histopathological analysis revealed that type I CPAM was the most frequent, followed by type II. No intraoperative complications were noted. The postoperative course was uneventful in all patients. At a median follow-up of 24 months, all were asymptomatic and thriving.

Graph 1: Radiological findings of CPAM in our study





Discussion

Our 10-year experience with CPAM highlights the clinical variability of this malformation and the importance of surgical management. The majority of patients in our series presented after birth, with only two having antenatal diagnoses. This emphasizes the need for improved prenatal screening and follow-up.

Histologically, most lesions were classified as type I CPAM, which aligns with existing literature. Type I lesions are generally associated with good prognosis post-surgery. Type II lesions, less frequent in our series, may be associated with other malformations.

Thoracotomy remains the standard surgical approach in our setting. Although thoracoscopic techniques are gaining ground, they require equipment and expertise not widely available in all centers. Literature supports the safety and efficacy of video-assisted thoracoscopic surgery (VATS), particularly in elective cases.

The prognosis for CPAM is excellent with early diagnosis and resection. None of our patients experienced postoperative mortality, and all had favorable outcomes during follow-up. Delayed treatment increases the risk of infection and long-term pulmonary damage.

These results are consistent with international studies such as those by Langston et al. [1] and Morini et al. [2], which affirm the benefits of elective surgery before symptom onset. Antenatal detection, combined with multidisciplinary follow-up, optimizes surgical timing and minimizes complications.

Conclusion

Congenital pulmonary airway malformations are rare but potentially severe anomalies. Early detection—preferably antenatal—combined with elective resection allows optimal management. Histological analysis remains essential to confirm diagnosis and guide prognosis. Larger multicentric studies are needed to establish standard treatment algorithms in both symptomatic and asymptomatic patients.

ACKNOWLEDGEMENTS

The authors have no acknowledgements to declare and report no conflicts of interest.

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