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Antenatal Diagnosis and Surgical Management of Cpam: A Case-Based Approach

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ABSTRACT

Introduction

Congenital Pulmonary Adenomatoid Malformation (CPAM, formerly Congenital Cystic Adenomatoid Malformation (CCAM) is an uncommon but clinically relevant malformation of the lung. The incidence of CPAM is estimated to range from 1:25,000 and 1:35,000 live births, representing 25% of all diagnosed congenital pulmonary malformations.

Case presentation

A 30 year old antenatal mother at 21 weeks of gestation came for anomaly scan and was diagnosed with mixed type, left CPAM with midline shift to right side, CPAM volume ratio 0.475. The child was followed up and presented as a 5-month-old male with antenatally diagnosed CPAM (Type 1) of the left lung presented with a 2-month history of intermittent cough and cold. Born at term via LSCS to a mother with gestational diabetes, he had transient neonatal respiratory distress. Clinical examination revealed reduced air entry in the left upper zones. Imaging showed a growing cystic lesion with mediastinal shift. At 4 months, CT confirmed a 4.8 × 4.4 × 4.5 cm Type 1 CPAM. Elective thoracotomy with left upper lobectomy was performed. Histopathology confirmed CPAM Type 1. Postoperative recovery was uneventful, with full lung re-expansion and discharge on POD 7.

Discussion

This case emphasizes the necessity of the multidisciplinary approach that includes the pediatrician, radiologist,obstretician, surgeon and pathologist in order to achieve the best outcome. Early detection, follow up, appropriate surgical resection and adequate histopathological assessment are essential in the management of Congenital Pulmonary Adenomatoid Malformation.

Conclusion

This case highlights the importance of early recognition, close monitoring, prenatal diagnosis, follow-up and proper treatment of Congenital Pulmonary Adenomatoid Malformation

Keywords: *Child, Congenital pulmonary Adenomatoid Malformation, Computed tomography, Congenital Cystic Adenomatoid Malformation*

1. INTRODUCTION

Congenital Pulmonary Adenomatoid Malformation (CPAM, formerly Congenital Cystic Adenomatoid Malformation (CCAM) is an uncommon but clinically relevant malformation of the lung.[1] It is characterized by hamartomatous proliferation of terminal respiratory bronchioles and with an associated reduction or complete absence of normal alveolar structures. The disordered proliferation is a result of non-functional lung tissue that is cystic

or adenomatous elements.[1] These lesions can be very diverse in terms of size, shape and clinical presentation, ranging from small and asymptomatic nodules to voluminous cystic lesions that can significantly occupy the thoracic space. The incidence of CPAM is estimated to range from 1:25,000 and 1:35,000 live births, representing 25% of all diagnosed congenital pulmonary malformations.[2] Although relatively uncommon, CPAM is the most common congenital cystic lung lesion diagnosed in neonates and infants. Its precise aetiology is unknown, but is thought to be due to abnormal branching morphogenesis during the pseudoglandular stage of lung development – during weeks 5-7 of gestation.[3] This disorganised development results in a mass of lung tissue that does not work but is composed of tissue that drains to the tracheobronchial tree.[4]

The risk of perinatal mortality for CPAM may be significant, particularly when there is a large lesion that results in mediastinal shift, compresses normal pulmonary parenchyma or evokes fetal hydrops.[5] However, with progress in antenatal imaging and neonatal intensive care, early diagnosis and treatment have become established that are associated with better perinatal outcomes. Most of these cases of CCAM are diagnosed prenatally by routine second-trimester ultrasonography, when CCAM may be visualized as an echogenic intrathoracic mass or as a lesion with recognizable cysts.[4] Based on the size and appearance of the cystic elements, CPAM has been categorized into macrocystic, microcystic, or mixed lesions; macrocystic lesions contain cysts >5 mm, microcystic lesions have cysts with CVR>1.6 is widely regarded as a cutoff with an elevated risk of establishing hydrops fetalis and thus determines the frequency of surveillance and the necessity of fetal therapy.[6]

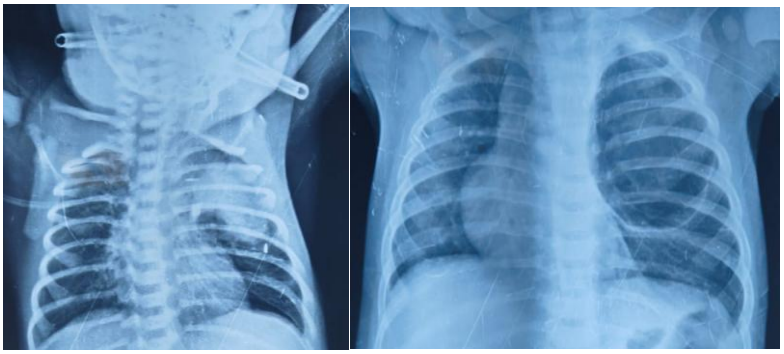
Many CPAMs are also asymptomatic postnatally and some will undergo spontaneous regression, particularly those diagnosed late in gestation.[4] However, on rare occasions an infected or large increasing lesion may cause respiratory distress, or recurrent pneumonia, feeding difficulties in the neonates and infants. Symptomatic lesions or those with radiological findings suggestive of complications should be further investigated by postnatal imaging such as CT and most often require surgical excision.[7] Surgical excision—most commonly lobectomy—is the gold standard for symptomatic CPAMs, or those at risk of complications such as recurrent infection, pneumothorax, or malignant transformation (e.g., pleuropulmonary blastoma or bronchioalveolar carcinoma). The decision of when to perform surgery is often made on an individual basis, in consideration of symptoms, tumor evolution, and risk evaluation.[8] Thoracoscopic resection is used in selectively in older children and in uncomplicated patients, which can provide the benefit of less postoperative pain and shorter hospital stay. The aim of this paper is to fill this gap using the description of a full term neonate with CPAM as well as the clinical course, imaging findings, surgical approach, and histopathological documentation. It underscores the value of careful antenatal and postnatal assessment, early surgical correction, and multidisciplinary care in the treatment of this rare though potentially lethal malformation.

CASE PRESENTATION

A 30 year old antenatal mother at 21 weeks of gestation came for anomaly scan and was diagnosed with mixed type, left CPAM with midline shift to right side, CPAM volume ratio (CVR) 0.475. On follow up CVR was 0.13, 0.17 at 31 weeks and 33 weeks of gestation respectively. The baby was born to a mother with GDM who was managed in pregnancy; lower segment cesarean section (LSCS) was done at term. The perinatal period was remarkable for transient respiratory distress in the immediate postnatal period, was managed conservatively requiring monitoring in the neonatal intensive care unit with no requirement for prolonged ventilation. The child was developmentally on target for expected milestones. The baby was growing well and was robustly nourished and moderately built. No history of multiple episodes of lower respiratory tract infections, ‘blue’ spells or failure to thrive were reported. There were no associated congenital anomalies or syndromic findings. The child appeared alert and playful on clinical examination. Vital signs were stable. Examination of the respiratory system showed reduced air entry at the left upper and mid zones with no adventitious sounds. Remaining systemic examination was normal. A 5 month old male child with the antenatal diagnosis of CCAM of the left lung was brought to paediatric outpatient department with history of intermittent cough and cold since last 2 months. At presentation there were no other symptoms like fever, feed intolerance, vomiting or respiratory distress.

Investigations

A chest X-ray taken at time of birth demonstrated a fluid-containing lesion in the left upper lung field, indicating a congenital lung lesion. Consecutive follow-up chest X-ray showed an increase in size of the lesion, with slight mediastinal shift to the right, and compression of the left lower lobe. At the age of 4 months, an ultra-low dose contrast enhanced computed tomography (CECT) of the thorax revealed a well-defined cystic lesion with fluid levels and several adjacent cysts, located in the anterior segment of the left upper lobe. The lesion was approximately 4.8 × 4.4 × 4.5 cm and was consistent with CPAM Type 1 (Stocker classification) having at least one large cyst greater than 2 cm in diameter. Because of the size and evolution of the tumor, and the possibility of infection, compression or malignant transformation, elective surgical resection was planned.



1A 1B

Figure 1A and 1B: Serial X-rays done from birth to pre-operative period at showed the lesion to be expanding with slight mediastinal shift to the right with compression of the left lower lobe

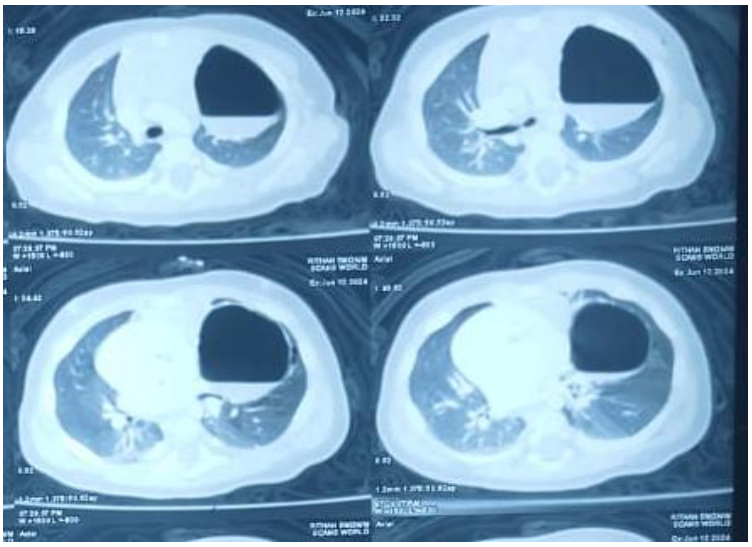


Figure 2: Ultra low dose contrast enhanced CT scan showed well defined cystic lesion with fluid level and surrounding cysts occupying left upper lobe anterior segment measuring 4.8x4.4x4.5cms- CCAM Type 1

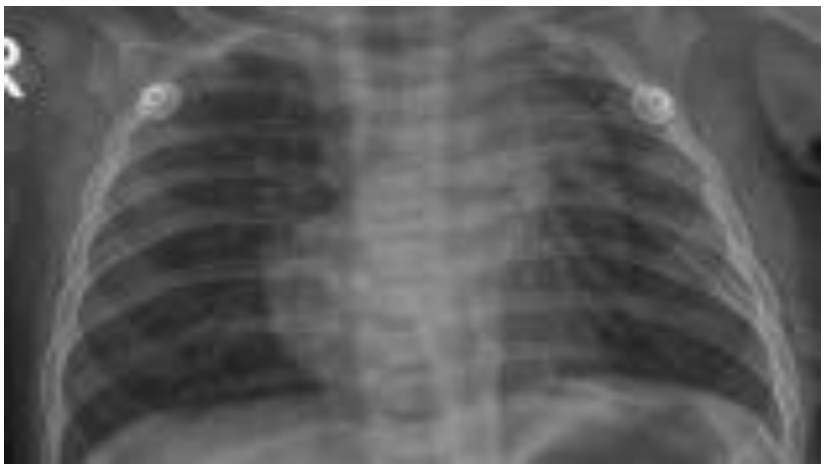


Figure 3: Postoperatively left lower lobe expansion was present

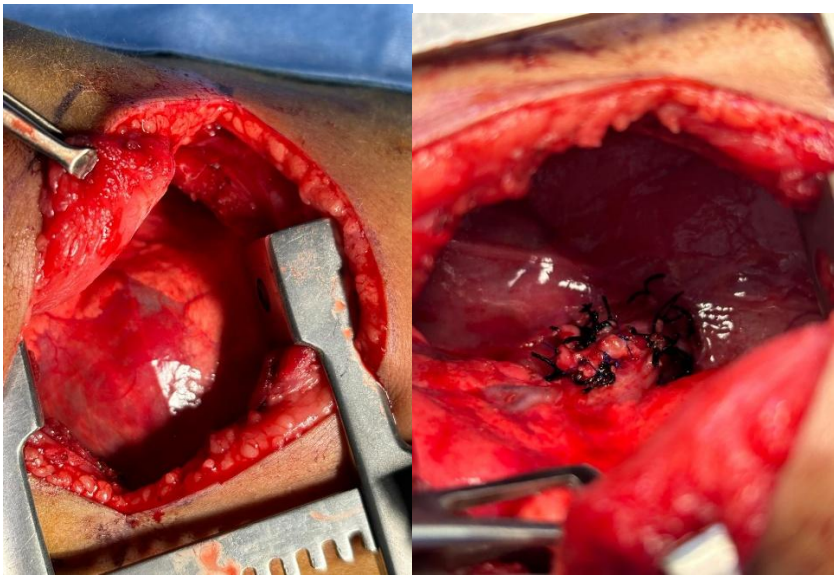


Figure 4: Intraoperative images showing the different steps

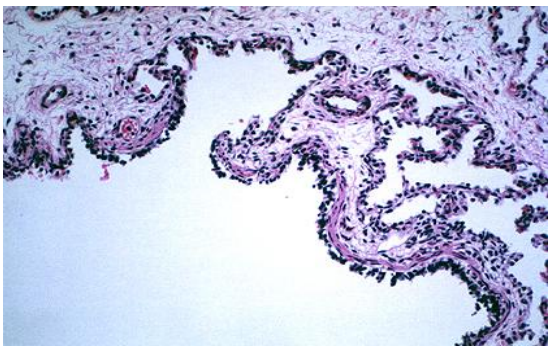


Figure 5: Histopathology slide of CPAM

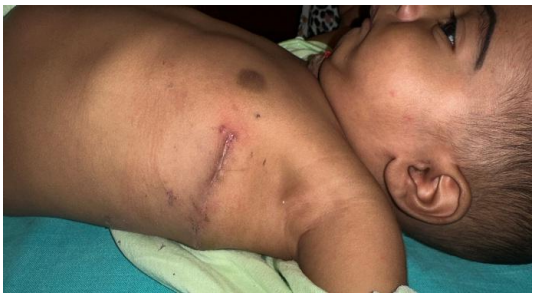


Figure 6: Postoperative image of the child

2. MANAGEMENT

The child was taken up for left thoracotomy with left upper lobectomy under GA. Intraoperatively, a large multiloculated cystic mass with turbid fluid, occupying the whole upper lobe was found; the size was approximately 8 × 4 cm. The lingula and lower lobe around this were collapsed secondary to mass effect. No pleural adhesion or intraoperative complications were observed. On gross and microscopic examination of the excised specimen, multifocal cystic spaces lined with ciliated columnar epithelium and having fibromuscular walls and rare mucous cells were present. No evidence of malignancy and atypia was observed. These features were compatible with CPAM Type 1. The patient had an uneventful postoperative course. Intraoperatively intercostal drain was placed and was removed on POD 4. The child was also successfully extubated and transferred to the postoperative ward for observation. Serial chest X-ray findings showed re-expansion of the left lower

lobe and disappearance of the mediastinal shifting. Histopathological examination revealed features that are suggestive of Congenital Pulmonary Airway malformation(CPAM) type 1. The patient was discharged home in a stable condition on POD 7 with regular follow-up and pulmonary rehabilitation being recommended.

3. DISCUSSION

This report describes a 5-month-old boy who was prenatally diagnosed with CPAM Type I in the left upper lobe and the presentation of an acute symptoms lasted approximately 2 months. The lesion grew in size, observed by serial imaging, and the development of clinical signs brought about the need for surgical treatment.

With the advent of routine prenatal ultrasonography, antenatal diagnosis of CPAM has become more prevalent. In the present case, the lesion was diagnosed prenatally, and surveillance was planned in the post-natal period.[7] However, lack of comprehensive pretreatment imaging characteristics, including the CPAM Volume Ratio (CVR), precludes evaluation of the growth dynamics of the lesion and potential fetal hydrops risk. However, the postnatal course was generally uneventful, with the child displaying mild respiratory symptoms until 5 months of ages.

The need for surgical resection was influenced by the lesion's progressive increase in size and the development of clinical symptoms. As management, resection of such lesions in CPAMs is usually required in symptomatic individuals and for lesions that have expanding growth and may be complicated with recurrent infections, pneumothoraces, or, although very rarely, malignant change. In our case, the child underwent left upper lobectomy by thoracotomy. Intraoperative findings revealed a cystic lesion, 8 × 4 cm in size, with turbid fluid in the left upper lobe, which resulted in the collapse of adjacent lung segments. Histopathological findings were compatible with CPAM Type I, large cysts lined by ciliated pseudostratified columnar epithelium.

The postoperative period was uncomplicated and, there was radiological evidence of re-expansion of the residual lung and resolution of mediastinal shift. This successful result is consistent with the results of the Bhende et al[9] who published the successful treatment of CPAM in infants by surgery, which showed the role of early intervention in preventing complications. Although CPAM Type I is thought to be associated with a favourable prognosis, malignant transformation in these lesions has been reported. Gopalaswamy et al[10]. reported one case of embedded CA with CPAM in a neonate with CPAM Type I, and emphasized the possibility of neoplastic transformation. While such cases are rare, this highlights the need for histopathological analysis of resected specimens and the thought of attempting surgical cure, even in asymptomatic cases. With respect to the surgical technique, although the performance of thoracotomy for this purpose is the traditional approach, it has also been successfully approached with less invasive methods such as video-assisted thoracoscopic surgery (VATS) in some cases.[11] Tsunezuka et al[12] detailed VATS lobectomy on a 1-year-old girl with CPAM Type II, which proves that minimally invasive methods are suitable for kids. Nevertheless, the surgical approach should be tailored to the patient's age, the size and location of the lesion and the surgeon's experience.

This case also emphasizes the necessity of the multidisciplinary approach that includes the pediatrician, radiologist, surgeon and pathologist in order to achieve the best outcome. Early detection, follow up, appropriate surgical resection and adequate histopathological assessment are essential in the management of Congenital Pulmonary Adenomatoid Malformation.

4. CONCLUSION

This case highlights the importance of early recognition and proper treatment of Congenital Pulmonary Adenomatoid Malformation. Prompt antenatal diagnosis allowed for planned postnatal surveillance and the risk of complication was avoided by early surgical therapy. The good postoperative outcome underscores the necessity of an interdisciplinary collaboration for the treatment of this type of congenital anomalies. In view of its potential (albeit rare) for malignant transformation, surgical resection and histopathological examination remains mandatory even in asymptomatic patients

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