A Rare Case of Congenital Pulmonary Airway Malformation in an Adult Patient: Case Report and Mini Literature Review

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Abstract

Congenital Pulmonary Airway Malformation (CPAM) is a rare developmental anomaly of the lung, usually diagnosed in infants but sometimes first detected in adults with non-specific respiratory symptoms. We report a 46-year-old non-smoking woman who suffered recurrent pneumonia and a persistent cavitary lesion, initially treated as a lung abscess. High-resolution CT revealed a large, septated cystic mass in the right lower lobe. Surgical resection was performed, and histopathology confirmed Type 1 CPAM complicated by secondary infection. Post-operative recovery was good, and imaging showed substantial improvement. This case underlines the importance of considering CPAM in adults with unexplained or recurrent lung infections. Surgical excision is both diagnostic and therapeutic in symptomatic or complicated cases.

Keywords: CPAM, adult, recurrent pneumonia, lung cyst, congenital lung anomaly

Introduction

Congenital Pulmonary Airway Malformation (CPAM), previously known as Congenital Cystic Adenomatoid Malformation (CCAM), is a rare developmental anomaly of the lung that results from abnormal branching morphogenesis of the terminal bronchioles during foetal development. This malformation leads to the formation of cystic, non-functioning lung tissue that does not participate in gas exchange.[1,2]

CPAM represents approximately 25% of congenital lung anomalies and is estimated to occur in 0.004% of all pregnancies.[1,2] It arises due to overgrowth of terminal bronchiolar structures and failure of normal alveolar development, leading to cyst formation with varying epithelial and stromal characteristics.[3]

CPAM was associated with high neonatal mortality, especially when large lesions caused mass effect leading to pulmonary hypoplasia, mediastinal shift, or foetal hydrops — a life-threatening condition in which abnormal fluid accumulates in two or more foetal compartments.[4] In such cases, survival rates were poor, and emergency postnatal interventions were often unsuccessful.

However, advances in prenatal imaging, particularly high-resolution foetal ultrasound and foetal MRI, have drastically improved early detection and monitoring.[4,5] These tools enable timely identification of at-risk foetuses, allowing for close surveillance and planning for delivery at tertiary centres with neonatal intensive care and paediatric surgical teams. In select cases with life-threatening lesions, prenatal interventions, such as steroid therapy or foetal surgery (e.g., thoracoamniotic shunt placement), may be considered to decompress large cysts and improve lung development.[4,5] Hence, neonatal outcomes have improved significantly, and survival rates for CPAM diagnosed prenatally or in the perinatal period now exceed 85–95% in many centres, particularly for lesions not associated with hydrops or other anomalies.[4,6]

Congenital Pulmonary Airway Malformation is classified into five types (0 to 4) based on their histological and pathological characteristics, according to the widely accepted Stocker classification system.[3] This classification provides insight into the embryologic origin, clinical features, and prognosis of each subtype. Type 0 is the rarest form, arising from the tracheobronchial tree. It presents with small, firm lungs and a solid appearance without cysts. Histologically, Type 0 lesions are lined by ciliated pseudostratified columnar epithelium and contain cartilage and submucosal glands like normal bronchial tissue. This type is usually incompatible with life due to severe pulmonary hypoplasia and extensive lung involvement.[3,6] Type 1 CPAM is the most common subtype, accounting for approximately 60–70% of cases.[3,4] Grossly, it consists of one or more large cysts greater than 2 cm in diameter, often multilocular. Microscopically, the cysts are lined by ciliated pseudostratified columnar epithelium and may contain mucus-producing goblet cells, with cyst walls composed of smooth muscle but typically lacking cartilage. Type 1 lesions generally have a good prognosis, and surgical resection is usually curative. However, there have been rare reports of malignant transformation, particularly into mucinous bronchioloalveolar carcinoma.[3,4,7]

Type 2 CPAM consists of multiple small cysts less than 2 cm, giving a spongy appearance.[3] Histologically, the cysts resemble bronchioles and are lined by cuboidal to columnar epithelium, with some smooth muscle in the walls. Type 2 is often associated with other congenital anomalies, such as renal or cardiac defects, and the prognosis depends heavily on these associated anomalies.[3,8] Type 3 CPAM appears as a bulky, solid mass with minimal cystic change and usually involves an entire lung lobe.[3] Microscopically, it consists of small bronchiole-like structures and alveolar ducts lined by cuboidal epithelium. Due to the mass effect and lack of cysts, this type often causes severe respiratory distress in neonates and has a poorer prognosis.[3,4] Type 4 CPAM features large, thin-walled peripheral cysts often greater than 10 cm in diameter, lined by flattened alveolar-type epithelium resembling distal acinar structures.[3,7] This type often presents later in childhood or adulthood and is associated with a higher risk of malignancy, especially pleuropulmonary blastoma (PPB). Because of this malignancy risk, surgical resection is typically recommended even for asymptomatic patients.[3,7]

Although often identified in infancy, some cases of CPAM may remain asymptomatic or undetected until adulthood, presenting with non-specific symptoms such as recurrent respiratory infections, chronic cough, dyspnoea, or pneumothorax.[1,7,8] In adults, CPAM can mimic conditions such as lung abscess, bronchiectasis, bronchogenic cysts, or even cystic neoplasms, leading to delayed or missed diagnosis.[1,7]

Surgical resection is the standard of care for symptomatic cases and is often advised for asymptomatic patients due to the risk of complications such as recurrent infections, air trapping, or malignancy.[4,5,7] Resection also provides a definitive diagnosis via histopathology. In adults, elective surgery is typically well-tolerated, with favourable outcomes.

In this report, we present the rare case of a 46-year-old woman who presented with recurrent pneumonia and lung abscess and was subsequently found to have Type 1 CPAM with secondary infection. Her case underscores the importance of considering congenital anomalies in the differential diagnosis of chronic pulmonary symptoms in adults.

Case Report

We present a 46-year-old lady, nonsmoker and doesn't have any medical background and no family history of lung diseases or malignancies. This patient was admitted multiple times with impression of community acquired pneumonia, then was seen and treated as lung abscess as she presented with a big cavity in superior and posteromedial segment of the right lower lobe of the lung with air fluid levels. She was treated with multiple antibiotics including amoxiclav and clavulanic acid, clarithromycin and later on clindamycin, then the patient was commenced on ciprofloxacin followed by piperacillin/tazobactam. However, after several antibiotic courses the cavity remained the same and was not changed.

The patient underwent Pulmonary Function Test which showed Forced Expiratory Volume in the first second (FEV1) 2.25 L (85%), Forced Vital Capacity (FVC) of 2.71 L (83%), FEV1/FVC of 96%, Total Lung Capacity (TLC) of 5.34 (120%) and DLCO of 9.01 (115%).

The patient underwent High Resolution Computed Tomography (HRCT) of the chest which showed: Large and septated air filled cystic structure arising within the left lower lobe and mainly by the right upper lobe superior medial segments, measuring 8.8 x 8.5 x 15.8 cm. The structure connects to the right lower lobe bronchi through multiple subsegmental bronchi. Fluid noted within the dependent aspect of the lesion giving an air-fluid level indicating some added infection. Findings are suggestive of congenital lesion with features that are favoring congenital pulmonary airway malformation CPAM, which is now infected [Figure 1].

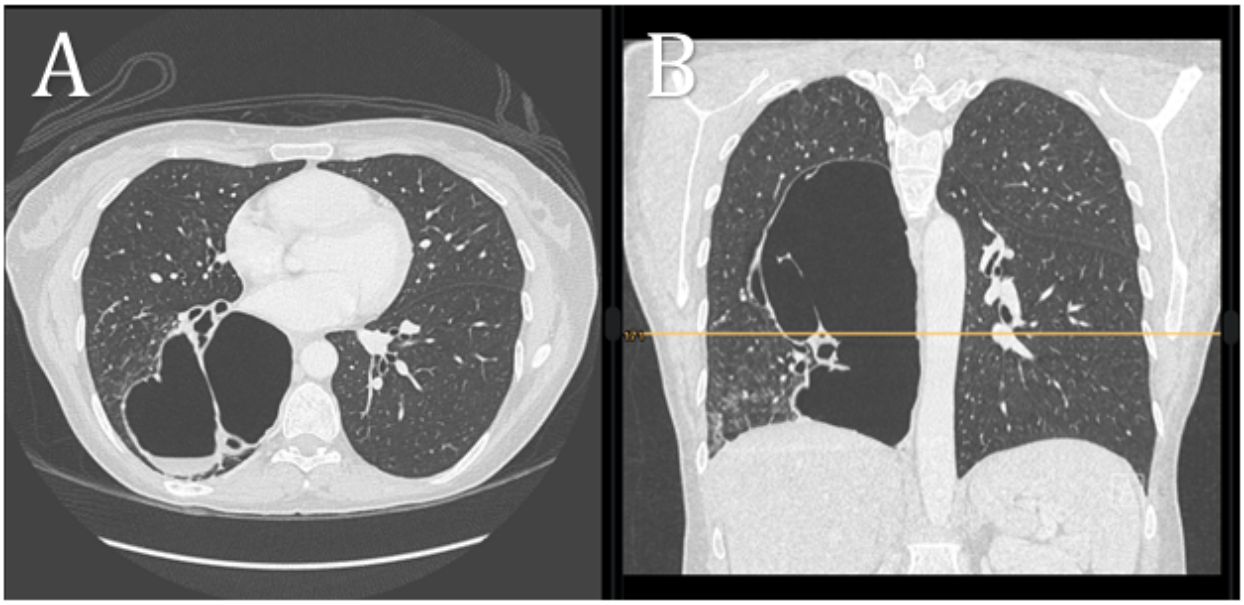

Figure 1: (Pre operation): Axial (A) and coronal (B) lung window showing a large right lower lobe multi-septated cystic structure with an air-fluid level. This structure connects to the right lower lobe bronchi. Findings are suggestive of congenital lesion with features that are favoring congenital pulmonary airway malformation CPAM.

She underwent right mini-thoracotomy excision of infected bronchogenic cyst and repair of multiple broncho-pleural fistulas. During the surgical procedure it was found to have pus filled bronchogenic cyst that had multiple blood supply from upper a lower lobes blood vessels. Following the excision of the cyst, the lung collapsed due to air leaks from multiple broncho-pleural fistulas which were repaired eventually.

The histopathology reported as the specimen grossly received as ruptured cyst in 2 pieces with smooth inner white surface. Microscopy shows a large cyst lined by ciliated respiratory-type epithelium with areas of epithelial denudation. The wall is composed of thick smooth muscle fibers with foci of mixed acute and chronic inflammation. Numerous smaller cystic spaces are seen within the wall and adjacent parenchyma, some lined by ciliated columnar and others by cuboidal epithelium. Foamy macrophages are noted within the cyst lumina. The histological diagnosis was Congenital Pulmonary Airway Malformation (CPAM), predominantly Type I with secondary inflammation present [Figure 2].

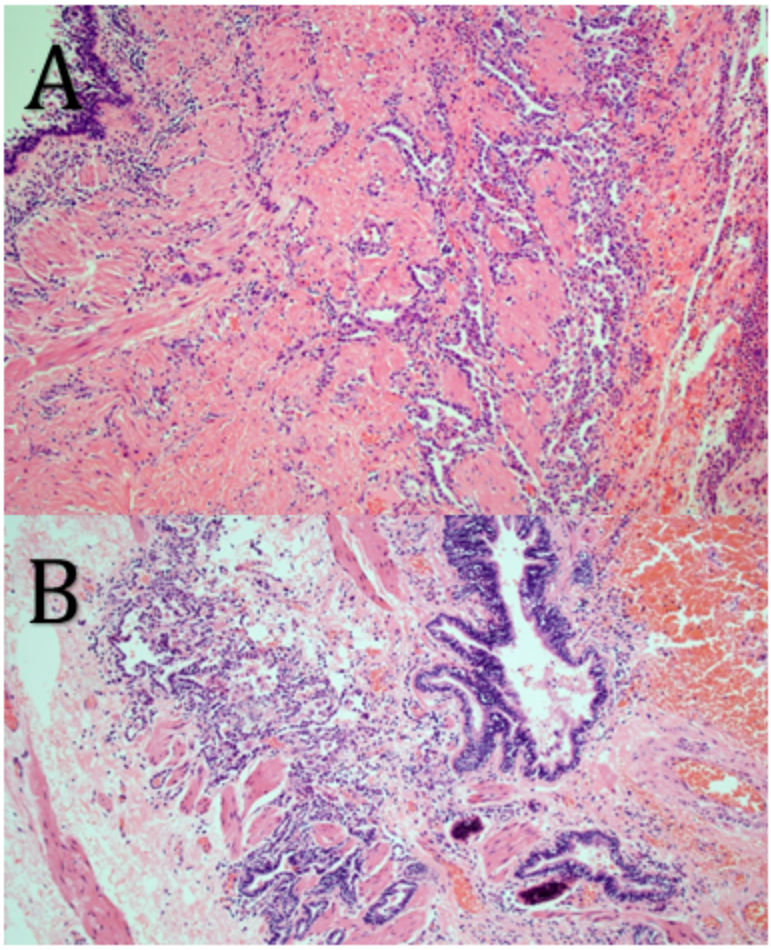

Figure 2: (a) HEx10: Larger cyst lined by ciliated respiratory type epithelium with acute and chronic inflammation. Prominent smooth muscle is the wall. (b) HEx10: Admixed with smaller cysts lined by respiratory type epithelium or cuboidal epithelium with pulmonary macrophages in the lumen. Smooth muscle fibers and mixed acute and chronic inflammation are noted in the wall.

The patient experienced mild cough and shortness of breath in the post-operative period, and the repeated CT scan showed significant interval reduction in size of the right lower lobe multi-lobulated cyst in-keeping with histopathology proven congenital cystic adenomatoid malformation [Figure 3].

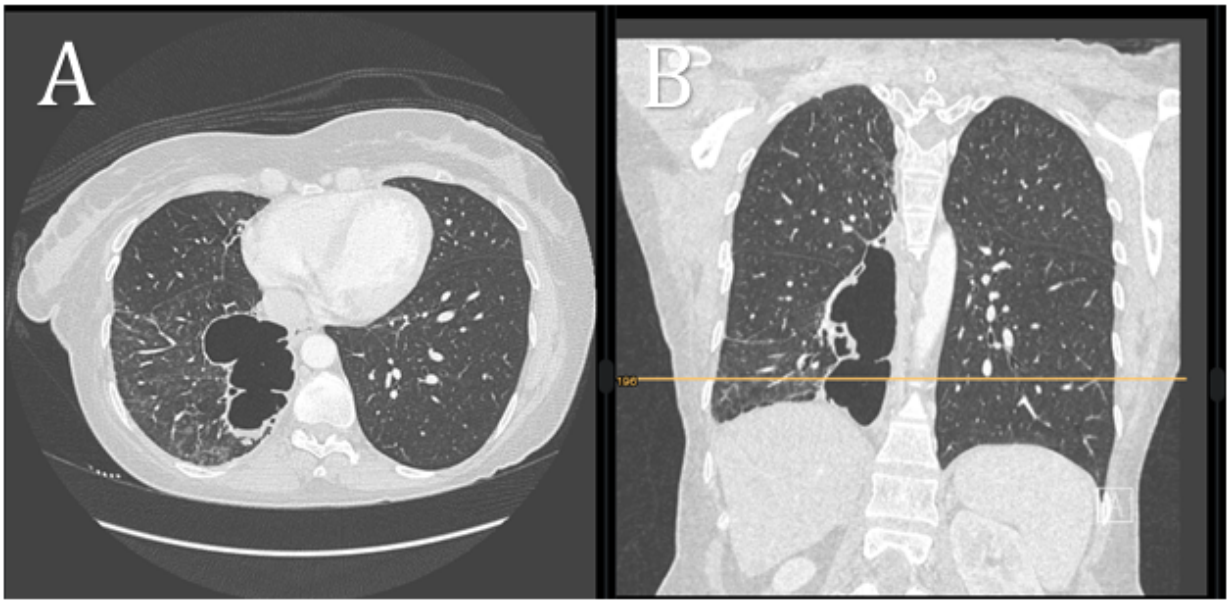

Figure 3: (Post operation): Axial (A) and coronal (B) lung window showing interval reduction in size of the previously seen large right lower lobe multi-septated cystic structure.

Discussion

CPAM is one of the known congenital lung diseases and it happens because of abnormalities during embryogenesis which leads to anomalous bronchial morphogenesis. This disease can be detected antenatally, and the presentation of the patients can vary from being asymptomatic or to have severe respiratory distress.[4] When diagnosed during infancy, it is often associated with significant morbidity, including respiratory failure and recurrent infections. However, the presentation in older individuals can be quite subtle, often leading to a delay in diagnosis. In adulthood, CPAM can present with symptoms like recurrent pneumonia, chronic cough, or hemoptysis, which can easily be misattributed to other more common lung diseases.[7]

The presentation of CPAM in adulthood is particularly challenging because of the diagnostic overlap with other lung conditions. For instance, Liu et al. reported an 18-year-old female with a history of recurrent respiratory infections who was eventually diagnosed with CPAM. Surgical resection led to complete resolution of her symptoms.[8] This is consistent with our case, where the patient was initially misdiagnosed with recurrent community acquired pneumonia and lung abscess, later found to have an infected congenital pulmonary malformation.

Confirming the diagnosis of CPAM in adults requires a high index of suspicion, as the symptoms and imaging findings often mimic a wide range of other pulmonary pathologies. The most common differential diagnoses include infected bullae, bronchogenic cysts, necrotic neoplasms, and cystic bronchiectasis.[1-3] Radiological imaging, particularly high-resolution computed tomography (HRCT), is essential for identifying the lesion and raising suspicion for CPAM. CT findings typically show multilocular cystic lesions of varying sizes, with or without air-fluid levels indicating secondary infection.[1,7] However, these features are not pathognomonic.

In our case, HRCT revealed a large, septated air-filled cystic lesion arising in the right lower lobe with features suggestive of secondary infection. Although these findings were suggestive of CPAM, they were not definitive.

Pulmonary function testing (PFT) may help assess the functional impact of the lesion, though it is not diagnostic. In our patient, lung function was preserved, and no obstructive or restrictive defect was observed—supporting surgical fitness but not contributing significantly to the diagnosis.

Histopathological examination remains the gold standard for confirming CPAM, especially in adults. In our case, the histological findings included large cysts lined by ciliated respiratory epithelium, smaller adjacent cysts, smooth muscle in the wall, and foamy macrophages within the lumina, consistent with Type 1 CPAM. The presence of chronic inflammation was indicative of secondary infection.

Given the risks of recurrent infection, progressive lung damage, and rare but documented risk of malignant transformation—especially into bronchioalveolar carcinoma or pleuropulmonary blastoma in types 1 and 4—surgical resection is both therapeutic and diagnostic.[8,9] In our patient, right mini-thoracotomy and cyst excision led to complete symptom resolution and radiological improvement, confirming the effectiveness of surgery.

Treatment is based on the clinical presentation and severity of the condition. In most symptomatic cases, such as those with recurrent lung infections or respiratory distress, surgical removal is necessary. Smaller, asymptomatic cases are typically monitored over time. Some evidence suggests that spontaneous regression can occur in cases detected through prenatal ultrasound.[7]

Conclusion

CPAM, though typically diagnosed in early life, can rarely present in adulthood with recurrent infections or atypical lung findings. This case underscores the importance of including congenital lung anomalies in the differential diagnosis of persistent pulmonary symptoms. Imaging may suggest CPAM, but histopathology remains definitive. Surgical resection is the treatment of choice, offering both diagnosis and cure. Early recognition and multidisciplinary management can prevent complications and improve outcomes, even in adults with delayed presentation.

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