Indian Journal of Thoracic and Cardiovascular Surgery (September–October 2023) 39(5):547–550
https://doi.org/10.1007/s12055-023-01555-y

CASE REPORT

Congenital pulmonary airway malformation in a 5 month old boy, complicated by pneumonia

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Received: 30 March 2023 / Revised: 17 May 2023 / Accepted: 2 June 2023 / Published online: 3 July 2023

Abstract

Congenital cystic lesions of the lung are rare. The most common congenital anomaly among them, involving the lower respiratory tract, is congenital cystic adenomatoid malformation (CCAM), currently referred to as congenital pulmonary airway malformation (CPAM). They may be incidentally detected on prenatal or postnatal imaging. They usually present within two years of life, rarely in adults. CPAMs may be asymptomatic at birth. Some may opt for observation alone due to lack of evidence on the incidence of long-term complications. However, rarely has a CPAM remained asymptomatic throughout life and complications eventually develop. Pneumonia is most common, which is not amenable to medical treatment alone. CPAMs are notorious for their known malignant potential and they may also lead to pneumothorax, hemoptysis and hemothorax. Computed Tomography Thorax is the investigation of choice. Surgical resection is known to be safe and is the mainstay of treatment. For patients who are diagnosed prenatally, surgery is recommended at 3 to 6 months, so that compensatory lung growth can occur. A five-month old boy from Bhutan, with cough, intermittent fever, respiratory distress and a history of recurrent lower respiratory tract infections, was diagnosed with CPAM, complicated by pneumonia and underwent surgery at our centre, following optimal medical management viz. antibiotics and supplemental oxygen. The post-operative course was relatively uneventful and he was discharged from hospital in 7 days.

Keywords Congenital cystic adenomatoid malformation · CCAM · Congenital pulmonary airway malformation · CPAM · Pneumonia · Surgical resection

Introduction

Congenital cystic lesions of the lung are rare. The most common congenital anomaly among them, involving the lower respiratory tract, is congenital cystic adenomatoid malformation (CCAM), currently referred to as congenital pulmonary airway malformation (CPAM). It was first reported as a distinct disease entity by Ch'In and Tang in 1949. The incidence of CCAM has been reported to be from 1 in 11,000 to 1 in 35,000 live births [1], with a higher incidence reported in the mid-trimester, due to spontaneous resolution. CCAMs are classified into five types [2] – type 0 CCAM arises from the trachea/bronchus, it is the rarest type and lethal at presentation; type 1 CCAM is the most common (50–70% cases), arising from the distal bronchus or terminal bronchiole; type 2 CCAMs (15–30%) arise from terminal bronchioles; type 3 CCAMs (5–10% cases) arise from acinar-like tissue; and type 4 CCAMs (5–15% cases) have an alveolar origin with malignant potential and have been known to be associated with pleuropulmonary blastoma. CCAMs are usually isolated and sporadic, but type 2 CCAM is different, with a majority having associated anomalies (cardiac/renal/gastrointestinal/musculoskeletal). It was proposed that the nomenclature of this lesion "CCAM" be changed to "CPAM" since the lesions described as cystic are present in only 3 of the 5 types and "Adenomatoid" only in one type (Type 3). CPAM more accurately fulfills all five types of this classification. Pathogenesis of CPAMs include abnormal proliferation of tissues, airway obstruction, and dysplasia or metaplasia of normal tissues. The pseudo-glandular phase of lung development is usually affected. The vascular supply for CPAMs is

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pulmonary, presentation is usually unilateral, more in males and a tracheo-bronchial connection is present. The presentation is usually in the first two years of life, with respiratory distress, though it has been reported in adults [3, 4], sometimes complicated by pneumonia.

Case report

A five-month old boy from Bhutan, weighing 5.6 kg, presented with cough, intermittent fever, respiratory distress and a history of recurrent lower respiratory tract infections. A Contrast Enhanced Computed Tomography (CECT) Thorax revealed a well defined, large. multi-loculated cystic lesion with enhancing solid component, measuring 6.4cmX4.8cmX8.8 cm involving the left lower lobe. It was suspected to be CPAM, exerting a mass effect with a mild contralateral shift of the trachea and cardia and inferior displacement of the left hemidiaphragm. Following a routine pre-operative work-up, broad-spectrum antibiotic coverage and supplemental oxygen therapy for a week, informed consent from both parents, the patient was taken up for surgery. In the right lateral position, a muscle-sparing posterolateral thoracotomy was done through the left 6^{th} intercostal space, the left lung mass was approached and the mass was found to be located in the left lower lobe, measuring 10cmX8cmX10cm, adherent to the parities laterally and the diaphragm below. The mass was completely resected out and the left lung checked for leaks and repaired with interrupted 5–0 polypropylene sutures. The resected lung mass was sent for histopathology examination (Fig. 1), which showed CPAM. The patient made a relatively uneventful recovery, and was discharged from hospital in 7 days.

Discussion

The major differential diagnosis for CPAM is bronchopulmonary sequestration (BPS). BPS may be detected on prenatal ultrasound and usually appears as a well-defined, solid and homogenous mass. The difference between BPS and CPAM is that BPS has no connection to the tracheobronchial tree and that it receives its blood supply from the systemic circulation via an anomalous systemic artery instead of the pulmonary circulation, as in CPAM. Other diagnoses to consider when diagnosing CPAM include congenital diaphragmatic hernia, bronchogenic cyst, congenital lobar emphysema, and pneumatoceles [5]. Cystic fibrosis, primary ciliary dyskinesia (PCD) and Williams Campbell syndrome (WCS) may be rarer differentials. The pulmonary histopathology in case of congenital lobar emphysema is usually normal, with mild alveolar dilatation without destruction of the alveolar septa. PCD is a genetic disorder, causing abnormalities in the structure or function of the cilia, with mucus accumulation in the lungs. Therefore, in such cases, bronchial dilation is secondary to mucus accumulation. The congenital form of WCS is a cystic lung disease associated with a deficiency of cartilage in the bronchial tree, diagnosed by excluding other common causes of bronchiectasis. Bilateral involvement of the lung and the association with other congenital anomalies (usually cardiac) is often observed [6].

There is still no consensus as regards the optimal management of asymptomatic CPAMs. The CONNECT consortium (the COllaborative Neonatal Network for the

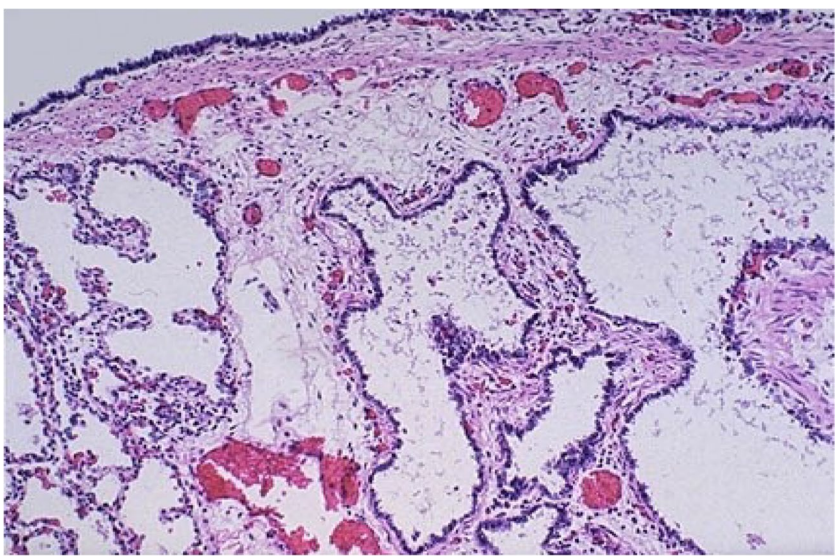

Fig. 1 Histopathology picture of the resected lung mass, suggestive of a Congenital Pulmonary Airway Malformation (CPAM), showing proliferation of the terminal respiratory structures forming cysts, which are lined with ciliated pseudostratified epithelium, with polypoid projections of mucosa and small cartilage foci in the wall of the bronchus

first European CPAM Trial)-an international collaboration of specialized caregivers, underlines a set of outcome parameters, which include respiratory insufficiency, surgical complications, mass effect and multifocal disease. These outcome parameters have been incorporated in the CONNECT trial, a randomised controlled trial which aims to compare between the conservative and surgical management of asymptomatic CPAMs [7]. In this trial, children will be followed up for 5 years and it aims to put to rest the debate between the 'wait and watch' versus aggressive surgical management for asymptomatic CPAMs.

CPAMs may be incidentally detected via routine prenatal ultrasound or postnatal imaging. They may be asymptomatic at birth. Some may opt for observation alone due to lack of evidence on the incidence of long-term complications [8]. However, rarely has a CPAM remained asymptomatic throughout life and complications eventually develop [8]. Pneumonia is most common, which is not amenable to medical treatment alone in form of antibiotics or supplemental oxygen. CPAMs are notorious for their known malignant potential and they may also lead to pneumothorax, hemoptysis and hemothorax and therefore qualify for early resection. For patients diagnosed prenatally, surgery is recommended at 3 to 6 months, so that compensatory lung growth can occur. The post-operative course is usually uneventful and long-term follow-up has shown adequate respiratory function. Even in cases of CPAM complicated by pneumonia, surgery may be performed safely after the clinical and laboratory parameters have improved following antibiotic therapy and supplemental oxygen, even in the absence of complete radiological resolution [9]. Surgery is the mainstay of treatment, and often resection of the lung mass is accompanied by segmentectomy, lobectomy, bilobectomy etc. depending on the anatomy of the lesion. The surgical approach has undergone changes over the years with thoracoscopy producing similar results as thoracotomy, with lesser pain, better cosmesis and relatively shorter hospital stay being the added advantages over the traditional thoracotomy approach [10]. However, for lesions bigger than 5 cm in size, previous lung infections and pleural thickening and inability to tolerate single lung ventilation, thoracotomy may be the preferred approach by the surgical team, as was the case here [10].

Out of the different types as per the Stocker classification [2], Type 1 reportedly has the best prognosis. In case of type 2, the prognosis depends on the severity of the associated anomalies. Type 3 often has hypoplasia of an entire lobe, producing complications like pulmonary hypertension. Type 4 has an excellent prognosis with surgical resection but it also has a strong malignant potential, with a few reported cases of pleuropulmonary blastoma [5].

Conclusion

CPAM is a rare congenital condition, which usually presents early, up to 2 years of life, and is often detected incidentally on routine prenatal or postnatal imaging. Sometimes they may present in adults. The usual presentation is cough, respiratory distress and recurrent lower respiratory tract infections. Computed Tomography Thorax is the investigation of choice. Surgical resection is known to be safe and is the mainstay of treatment. For patients who are diagnosed prenatally, surgery is recommended at 3 to 6 months, so that compensatory lung growth can occur.

Acknowledgements We gratefully acknowledge Dr. Subhasis Saha, MCh, Consultant Pediatric Surgeon, AMRI Hospital, Mukundapur, Kolkata, India.

Funding None.

Data availability The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethical committee approval Not applicable, it being a case report.

Human and animal rights statement All procedures involving human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent and consent for publication Written, informed consent was obtained from the patient's parents for the management of this case and the use of clinical data and images relevant to this case, for publication.

Competing interests The authors declare that they have no competing interests.

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