A 4-Month-Old Girl with Persistent Respiratory Distress and Multiple Admissions to the Pediatric Intensive Care Unit Due to a Congenital Bronchogenic Cyst

Patient: Female, 4-month-old  
Final Diagnosis: Bronchogenic cyst  
Symptoms: Respiratory distress • wheezing  
Medication: —  
Clinical Procedure: —  
Specialty: Pediatrics and Neonatology  
Objective: Congenital defects/diseases  
Background: Bronchogenic cysts are foregut duplication cysts that result in congenital cysts of the tracheobronchial tree. They can be fatal especially, if they present in early infancy. Persistent respiratory distress is the most frequent reported presentation. Histologically, bronchogenic cysts are unilocular fluid-filled lesions that are composed of respiratory epithelium. This report presents the case of a 4-month-old girl with persistent respiratory distress and multiple admissions to the Pediatric Intensive Care Unit (PICU) due to a congenital bronchogenic cyst.

Case Report: We describe a 4-month-old girl with persistent respiratory distress, intermittent choking, and recurrent PICU admissions. The patient was managed as a case of bronchiolitis, which led to ineffective treatment numerous times. Radiological work-up revealed unusual findings of asymmetrical hyperinflation. Bronchoscopy, which was performed to clear the airway and retrieve a possible foreign body, showed a non-pulsatile mass compressing the entry of the main bronchi with more pressure on the left main bronchus. Bronchogenic cyst was suspected and confirmed with high-resolution computed tomography (HRCT). Surgical intervention was performed, with no reported complications.

Conclusions: This report has shown that in neonates presenting with respiratory distress and no signs of infection, a broad differential diagnosis including congenital cysts should be considered. As in this case, lung and thoracic imaging are required to confirm the diagnosis. We also highlight the need to involve subspecialties to avoid potential delay in diagnosis or exposing patients to unexpected complications.

Keywords: Bronchogenic Cyst • Bronchoscopy • Intensive Care Units

Abbreviations: CT – computed tomography; MRI – magnetic resonance imaging; VATS – video-assisted thoracoscopic surgery; PICU – Pediatric Intensive Care Unit; HRCT – high-resolution computed tomography

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**Background**

Bronchogenic cyst is a rare benign congenital malformation that occurs due to development of buds in any part of the tracheobronchial area early in pregnancy [1]. The cyst’s location is determined by the embryological stage at which the defect occurs. They can be found in the mediastinum, pulmonary parenchyma, and, rarely, the abdominal area and skin [1]. The incidence rate is about 1/42,000 to 1/68,000 individuals, but the epidemiology is unclear [2]. Prenatally, bronchogenic cysts can be identified as unilocular fluid-filled cysts using ultrasonography [1]. In infancy, compression of the adjacent structures can lead to respiratory distress with recurrent cough and wheezing, whereas adults usually experience recurrent infections. However, especially in adults, bronchogenic cysts can be completely asymptomatic [1,3]. Bronchogenic cysts rarely cause symptoms in newborns, but respiratory impairment and stridor are possible manifestations [4].

Bronchogenic cysts can be diagnosed most accurately by histological examination. Cysts are usually solitary lesions composed of respiratory epithelium, fibrous tissue, smooth muscle fibers, and mucous glands. They contain air, fluid, or both [5]. Other diagnostic tools are chest radiography, computed tomography (CT), and magnetic resonance imaging (MRI); however, these modalities are not specific enough for diagnosis. Chest X-rays can be useful to reach the diagnosis in 77% of patients [5]. Subcarinal cysts are more visible and easier to localize, given their shape and consistency and their adjacency to other structures on CT scan compared to plain radiographs [3].

As soon as bronchogenic cysts are identified, surgical resection is indicated. Asymptomatic patients are also advised to undergo surgical intervention, as untreated intrathoracic cysts often result in mortality [6]. Afterward, a precise tissue examination can be performed to confirm the diagnosis [3]. Unresected cysts have a higher risk of infection, respiratory distress, and, rarely, recurrent pneumothorax and hemoptysis [3]. This report presents the case of a 4-month-old girl with persistent respiratory distress and multiple admissions to the Pediatric Intensive Care Unit (PICU) due to a congenital bronchogenic cyst located in the subcarinal area.

**Case Report**

A 4-month-old girl was admitted to the general pediatric ward with history of shortness of breath since birth. Past medical history revealed several previous hospital visits with episodes of persistent respiratory distress that started at 1 month after birth. The baby was managed with bronchodilators and normal saline nasal drops on several occasions without any response. She required multiple visits to the PICU for respiratory support and wheezy chest. The baby was a product of a twin pregnancy that ended with cesarean section with a birth weight of 2.4 kg. She was discharged on the fourth day in good condition. The baby had history of occasional choking but never had cyanotic spells or noisy breaths, and there was no history of atopy in the family.

The patient appeared well, with a respiratory rate of 52 breaths/minute. Oxygen saturation ranged from 87% on room air and 98% on oxygen 0.5l/m. The baby had normal growth parameters. Auscultation revealed reduced breath sound on the left side, predominant wheezy chest on the right side, and scattered crackles bilaterally were also noticed. Apart from tachycardia, normal heart sound was heard, and the rest of examination results were normal.

The initial blood work-up was within normal limits, and a viral screen was negative. Her initial atrial blood gas readings revealed mild respiratory acidosis (pH 7:32, PCO\textsubscript{2} 42, HCO\textsubscript{3} 25 mmHg). The chest X-ray was very alarming and puzzling to the team, as there was significant bilateral hyperinflation, which was worse on the left side (Figure 1). The initial clinical impression included introduced foreign body aspiration, for which flexible bronchoscopy of the lower airways was performed, but no foreign body was seen. However, a dome-shaped carina with obliteration of the sharp angle and significant external non-pulsatile compression on both entrance of the main bronchi, especially the left side, were noticed (Figure 2). A prefusion
scan was performed and showed decreased perfusion in the left side. Additionally, echocardiography was conducted to rule out any congenital cardiac abnormalities, which was normal. Eventually, a CT scan of the chest demonstrated a subcarinal swelling 1×1 cm causing compression on the left main bronchus (Figure 3). The bronchoscopy and CT scan combined confirmed the diagnosis of bronchogenic cyst.

The patient was initially admitted as a case of wheezy chest for evaluation. Bronchiolitis was excluded due to persistent respiratory distress and negative virology results. In addition, there were no prodromal symptoms such as runny nose, blocked nose, and fever, or history of sick contacts. An introduced foreign body in the airway was suspected due to her history of contact with other children at home. Airway anomalies, silent aspiration, and Macleod syndrome were all kept as potential differential diagnoses. Eventually, the radiological findings were consistent with typical mediastinal bronchogenic cyst; therefore, the patient underwent video-assisted thoracoscopic surgery (VATS). Surgical intervention was conducted through right thoracotomy approach. The cyst was identified in the subcarinal area, anterior to the esophagus, but connected to the inferior aspect of the carina and subcarinal bronchial wall tissue. It was carefully dissected with no complications of air leak or bleeding. A biopsy was taken and showed a cystic lesion lined by ciliated pseudostratified columnar epithelium surrounded by a fibromuscular wall containing cartilage and mucous glands, consistent with the diagnosis of bronchogenic cyst. No intra- or postoperative complications were reported. The patient’s symptoms subsided, and no recurrence was detected during subsequent visits.

Discussion

This child presented with persistent respiratory distress and wheezy chest that were not responding to bronchodilators. The causes of recurrent respiratory distress in children were investigated; she underwent flexible bronchoscopy and CT scan of the chest, which revealed a bronchogenic cyst located in the subcarinal area. Surgical intervention was conducted, and pathologic assessment confirmed the diagnosis of the bronchogenic cyst. This case highlights the importance of considering a broad differential diagnosis in infants with persistent respiratory distress, including congenital lung malformations.

Bronchogenic cysts are rare congenital malformations, accounting for less than 15% of all primary mediastinal malignancies [7]. They account for approximately 71% of all intrathoracic foregut cysts [8]. Cystic lesions can be a result of abnormal budding of the foregut or the tracheobronchial tree, and manifest differently depending on their location [1]. After the first effective surgical intervention of bronchogenic cysts, they were grouped according to their anatomical distribution into paratracheal, carinal, hilar, paraesophageal, and miscellaneous [3]. Bronchogenic cysts usually have a thin-walled lesion that is filled with mucus and consists of respiratory epithelium cells. Other important histological characteristics include seromucous glands, smooth muscle, and distinctly hyaline cartilage [1]. Ribet et al reported that bronchogenic cysts were
symptomatic in 70.8% of affected children [7]. Symptoms are either secondary to airway compression, as in our case, or are due to infection [7]. Some cases are asymptomatic and are usually discovered accidentally, even in adulthood [1]. The cyst’s location can have an impact on how it behaves. Wheeze and recurrent pneumonia are symptoms of a bronchogenic cyst at a bronchial site [9]. Hyperinflation of the distal lung may occur in case of partial airway blockage resulting in a ball-valve effect. However, when the obstruction is complete, distal atelectasis can be seen [9]. A similar presentation to our case was reported in an 8-month-old girl who had recurrent cough and wheezing. Eventually, a bronchogenic cyst located in the middle mediastinum that was compressing the carina and the proximal part of the right and left main bronchi was confirmed by CT scan of the chest [10]. However, a different presentation was reported in a 6-month-old infant who was noticed to have stridor since birth, associated with respiratory distress. An CT scan of the chest eventually revealed a middle mediastinum cyst, ventral to the tracheal bifurcation and dorsal to the right pulmonary artery, compressing the trachea and the main bronchi [11].

Bronchogenic cyst can be diagnosed antenatally or in the neonatal period, where the cyst is often radiopaque due to fluid accumulation [1,3]. Maurin et al conducted a retrospective study describing the approach to diagnosis and management of bronchogenic cysts. Some participants were diagnosed antenatally by ultrasound followed by MRI. All patients had a CT scan at 1 month of age, which was repeated at 1 year before the surgical intervention was performed. Three out of 8 children were discovered to have different types of malformations postnatally, indicating that there is a high probability of an antenatal diagnosis error. These findings support the need for a postnatal CT scan to provide a more precise diagnosis [12]. A well-defined rounded lesion can be detected on a plain chest X-ray. It can be useful to establish the presence of a cyst depending on several features. Small cysts, for example, can be difficult to spot in the presence of a large cyst. Similarly, large thin-walled cysts can be misdiagnosed as pneumothorax or lobar emphysema [3]. Asymmetrical hyperlucency on left side were reported to be the main finding on chest X-ray, as presented in our case and in other similar cases [10,13]. Lung hyperinflation can be explained by different mechanisms. Bronchial obstruction caused by a cyst can lead to hyperinflation as a result of collateral ventilation along the pores of Kohn [14,15]. In our case, a ventilation/perfusion scan confirmed the non-functioning nature of the hyperinflated left lung, in which ventilation can be impaired due to delayed emptying of the alveoli in the affected side, while perfusion often is diminished due to markedly attenuated blood vessels on the affected side. CT scans can show a well-demarcated homogeneous mass with water density [10,13]. Also, MRI can detect high-intensity protein-rich elements [6]. Most of the published cases similar to ours had normal echocardiogram findings, although 1 study described a case of a newborn with respiratory distress after birth and had abnormal echocardiogram findings. Following further investigations, a bronchogenic cyst arising from the trachea was found to be compressing the left atrium and pulmonary artery branches [16]. The differential diagnosis of bronchogenic cyst is broad and depends on the location of the cyst. For example, when the bronchial branches are affected, the cyst tends to be filled with air or can be of mixed type, which can be misdiagnosed as lung abscess, carcinoma, or tuberculosis [1,7]. It was reported that 84% of bronchogenic cysts are located in the mediastinal region, which, on the other hand, tends to be round and contains fluid that causes airway compression and raises a different spectrum of differential diagnoses [1,17]. Due to the asymmetry and hyperlucency of the left lung on radiographical images, the work-up in this patient was aimed to exclude the possibility of a foreign body introduced by other sibling, as this baby is still too young to be exploring her surroundings. However, the patient’s characteristics, such as her age and a history of recurrent respiratory distress, raised the suspicion of congenital lung anomaly as a possible cause. Bronchiolitis was excluded by the history of recurrent respiratory compromise and negative prodromal history of flu illness. Additionally, a history of intermittent choking along with physical examination findings of scattered crackles suggested a possibility of aspiration pneumonia; third-generation cephalosporin was commenced, and no improvement was seen. Acquired chest infection was also ruled out due to normal virology results. Swyer-James-Macleod syndrome was one of the possible differentials, as it is a rare complication of bronchiolitis obliterans with hyperlucency finding in the chest X-ray [18]. A comparable case had similar findings on flexible bronchoscopy, which showed a dome-shaped carina, with external non-pulsatile compression [10]. A CT scan was scheduled, and based on the CT scan findings and bronchoscopy images, a diagnosis of bronchogenic cyst was made.

Surgical removal of a bronchogenic cyst is highly advised to avoid unwanted consequences. Maurin et al found that cysts varied in terms of growth acceleration, with some increasing in size while others maintaining the same diameter over the first year of life, possibly due to mucus secretion [12]. The cyst can increase in size, thus affecting adjacent structures, causing respiratory compromise, rupture, and infections [1,17]. Particularly, bronchogenic cysts that are small and localized in the crania, as in our case, require an immediate surgical intervention to minimize the risk of complications [1]. Additionally, cysts that are attached to other structures carry a higher risk of complications, as they are difficult to excise [1,3]. A review of the literature revealed use of various surgical techniques, including sclerosing material injection, transbronchial or percutaneous aspiration, and mediastinoscopy, but a larger sample size is required to assess their effectiveness [19].
Conclusions

This report has shown that in neonates who present with respiratory distress and have no signs of infection, a diagnosis of congenital lung disease should be considered. As in this case, a high index of suspicion is required and an extensive work-up and multidisciplinary approach is essential to confirm the diagnosis. Surgical excision was reported to be therapeutic in most similar published papers and follow-up is critical to assess any potential deterioration. We report this case to alert pediatricians to unusual causes of respiratory distress and to enhance involvement of subspecialties with bronchoscopy services in the evaluation of such patients.

Declaration of Figures’ Authenticity

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