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Lobectomy for acquired lobar emphysema months following newborn repair of congenital diaphragmatic hernia

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ABSTRACT

Chronic lung disease is a known morbidity of congenital diaphragmatic hernia repair. Although described as a diffuse lung process, we present a case of localized lobar emphysema in a child with a congenital diaphragmatic hernia repair successfully treated with lobectomy. We aim to describe this unique clinical manifestation, detail the intraoperative findings, and describe the successful postoperative course.

1. Introduction

Congenital diaphragmatic hernia (CDH) occurs in 1 in 2500–5000 infants [1]. Although the morbidity and mortality remains high, survival rates have increased over the past two decades [2–4]. The decrease in mortality is in large part due to advances in ventilator management and the adoption of permissive hypercapnia [5,6]. The increase in survival is most prominent for those infants with severe respiratory compromise [7]. Long term pulmonary function studies have shown that the respiratory status of these infants can improve as they reach adolescence and adulthood [6,8]. However, the intermediate stage during the child's younger years, can be filled with persistent pulmonary dysfunction and periods of respiratory decompensation [9–11].

Chronic lung disease (CLD) has been reported between 40 % and 50 % of infants with CDH and may require intermittent intensive ventilator rescue methods [12,13]. It is hypothesized that the pulmonary morbidities are a consequence of abnormal lung development which is aggravated by prolonged neonatal ventilation [5,13,14]. Retrospective studies utilizing computed tomography (CT) to measure lung density and volume have shown emphysematous-like changes throughout the affected lung in children with CDH compared to age-matched controls [14]. Of note, these changes are different from those seen in children with neonatal congenital cystic pulmonary malformations or large pulmonary sequestrations [14].

Lobar emphysema (LE) is a relatively rare disease in infants and children [15]. Patients with LE can develop significant ventilation-perfusion (VQ) mismatches resulting in hypoxia and hypercapnia [16]. We present the first report of a patient with ventilator dependent respiratory failure since repair of CDH who subsequently developed ipsilateral single lobe right upper lobar emphysema. Although a case report has been published describing ipsilateral emphysema in a patient with previously repaired CDH, this finding was diffuse and found throughout the lung [17]. In this case, the multidisciplinary team of pediatric pulmonology, pediatric surgery and pediatric intensive care were able to diagnose and treat this unique condition.

2. Case report

An 8 month old boy presented to our pediatric emergency department with an acute episode of hypoxia, increased work of breathing, and bilateral wheezing. His past medical history was significant for a previous 30 week 4 day preterm infant born with a right sided diaphragmatic hernia repaired at one week of age and chronic ventilator dependence. During his initial hospitalization, he did not require extracorporeal membrane oxygenation (ECMO) but was placed on the oscillator prior to surgery. The child underwent a primary repair of the diaphragmatic defect and a gortex patch closure of the abdomen. This patch was subsequently removed during an open gastrostomy placement. The patient received a tracheostomy at 4 months and was discharged from the hospital at 5 months. The child's discharge ventilatory settings were: SIMV FiO2 24 %, rate 20, PIP 34 PEEP 7, Ti 0.7 and PS 18 (SIMV = synchronized intermittent mandatory ventilation; Fi02 = fraction of inspired oxygen; PIP = Peak inspiratory pressure; PEEP = positive end-expiratory pressure; Ti = Tidal volume; PS = pressure support). Review of the patient's medical records showed four additional hospital admissions over the subsequent 3

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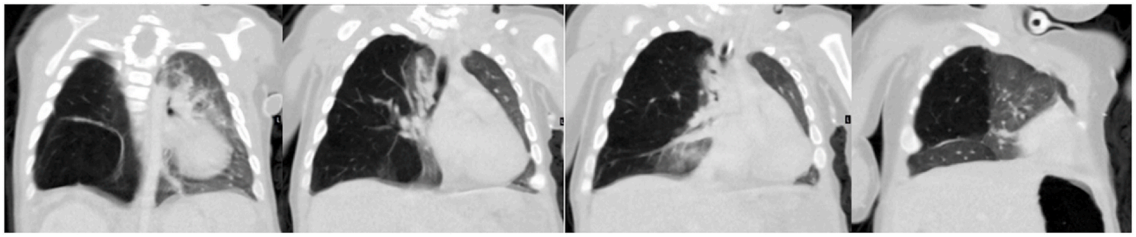

Fig. 1. Series from patient's preoperative CT showing hyperinflation and hyperlucency of the right upper lobe with a contralateral mediastinal shift resulting in left lung volume loss.

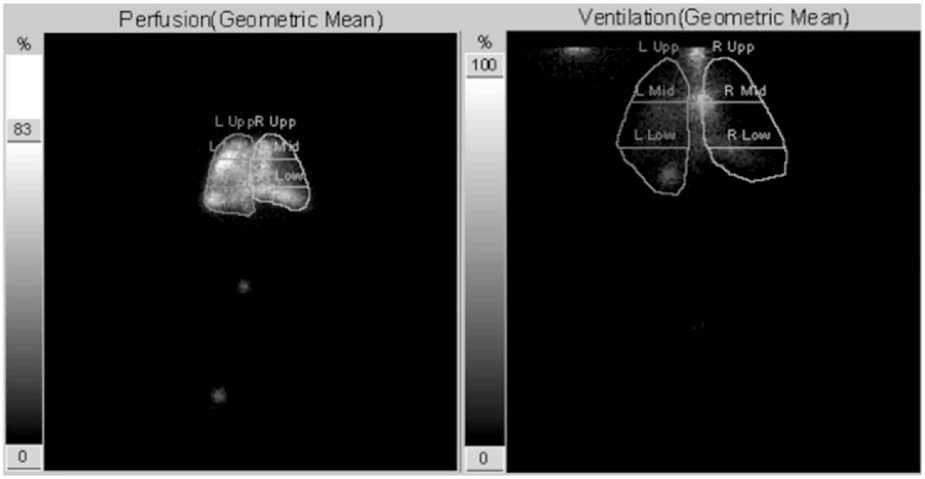

Fig. 2. Pre-operative ventilation perfusion study. There is a ventilation and perfusion defect involving the right apex. The differential quantitation of perfusion was 45 % to the right lung and 54 % to the left lung.

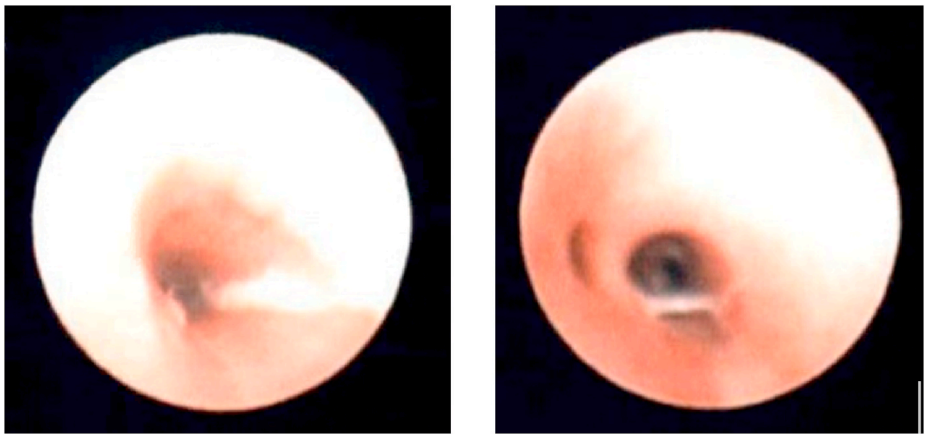

Fig. 3. Images from bronchoscopy performed preoperatively show bronchus intermedius leading into the right lower lobe and right middle lobe. No evidence of atresia, mucosal folds, or cysts affecting these two lobes of the right lung.

month period secondary to hypoxia and intermittent episodes of increased ventilator requirements. Radiographs during these hospitalizations showed a hyperinflated right hemithorax. Each time the child would eventually improve clinically with medical management and ventilator escalation.

During the last admission, the child underwent a CT scan of the chest which revealed hyperinflation and hyperlucency in the right upper lobe suggesting lobar emphysema (Fig. 1). A subsequent Ventilation/Perfusion (V/Q) scan showed normal and symmetric perfusion of the bilateral lungs. However, the right lung showed the upper third to have 12 % ventillation, middle third to have 23 %, and the lower third to have 14 % (Fig. 2). Overall, the right lung was contributing only 38 % of the ventilation, with the left lung contributing 62 %. Bronchoscopy revealed a normal carina and normal takeoff of the right upper lobe; however, there was bronchomalacia of that lobe and mucous plugging. These findings are consistent with lobar emphysema (Fig. 3). Of note, pulmonary function testing was not feasible due to the patient's age and current tracheostomy status. With an integrated treatment team including ICU physicians, pediatric surgery, and pediatric pulmonology the decision was made to proceed with the right upper lobectomy.

The child underwent a posterolateral right thoracotomy with single lung ventilation. The right upper lobe was very pale compared to the other lobes and the fissures were poorly developed, however, it accounted for the majority of the lung volume when inflated. With intermittent and controlled inflation of the lung, the emphysematous lobe was delineated. Dissection was started along the horizontal fissure as it extended posteriorly with sharp dissection and staple transection. The

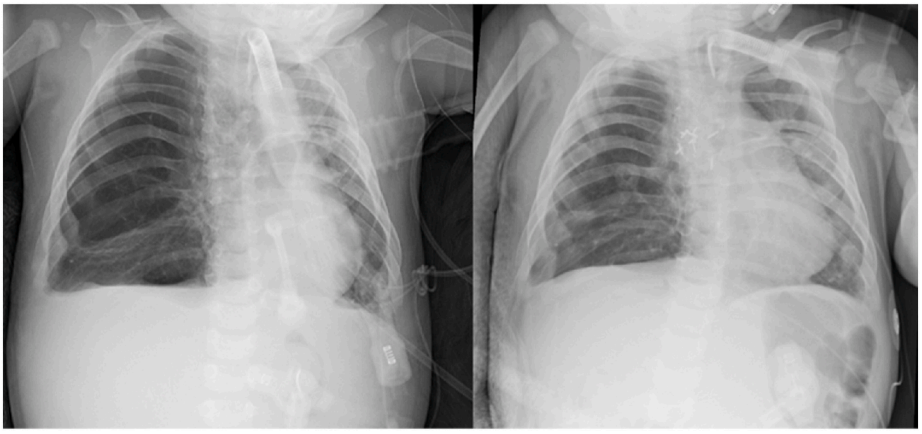

Fig. 4. Chest x-rays preoperative (Left) and post operative (Right). The hyperinflation and hyperlucency seen in the left image which is improved in the right image.

upper lobe bronchus and vascular supply were identified and controlled prior to ligation. Confirmation of the involved lobe was made with controlled released ventilation to the lung before the bronchus was ligated with a stapler. The vascular supply was then sealed and transected. A chest tube was placed and the thoracotomy closed.

The patient's postoperative course was uncomplicated and he was discharged on post operative day 12. Ventilatory requirements were significantly lower upon discharge. Upon presentation to the hospital he required a SIMV Fio2 60 %, PIP 30, PC 24, RR 44, iT 0.5. After surgery and at time of discharge the child was tolerating pressure support ventilation Fio2 30 %, PIP 20, PEEP 5. A preoperative and postoperative chest x-ray can be seen in Fig. 4; the initial lucency correlating to pulmonary hyperinflation has resolved radiographically. Pathology of the resected right upper lobe was consistent with lobular emphysema. Histologic sections show portions of disrupted lung tissue with pleural disruption and parenchymal hemorrhage. The alveolar spaces are expanded. Many alveolar spaces are filled with blood, and some contain aggregates of macrophages. No well-formed granulomas were appreciated. No significant acute inflammatory infiltrate is seen. The interstitial septae appear thickened with evidence of alveolar septal fibroplasia. There also appears to be mild peribronchial fibroplasia and pneumocyte hyperplasia.

3. Discussion

Current management of CDH has improved survival of the most compromised neonates. The majority of these CDH survivors have measurable lung disease and structural lung changes that lead to functional morbidity. The high oxygen requirements with prolonged and intermittent high pressure ventilation contributes to the development of chronic lung disease. Although long term studies report improvement of respiratory function as the child reaches the teenage years and young adulthood, there is a period of adaptation after the neonatal stage that can require intermittent intervention.

In children with CDH, the total number of alveoli are less than controls. Although the number of alveoli increases with time, the main manner by which the lung fills the chest cavity is by distension of the alveoli. Significant or prolonged ventilator pressures are thought to contribute to the pathogenesis of this emphysematous-like condition. Weis et al. demonstrated low-density areas in the ipsilateral lung compared to the contralateral lung consistent with diffuse emphysematous changes. This case is the first reported case of isolated ipsilateral right upper lobar emphysema with resulting chronic respiratory failure after repair of CDH.

Unlike congenital lobar emphysema (CLE), the lobar emphysema resulting from CDH is an acquired form of lobar emphysema, but results in similar pulmonary dysfunction. The pre-operative bronchoscopy and V/Q scan shows changes isolated to the right upper lobe with normal function of all other lobes. As seen in Fig. 3, the middle and lower lobes of the right lung were interrogated preoperatively and noted to be normal. Intraoperatively these findings were confirmed when the lung was inspected and the abnormal upper lobe identified. The pathology findings are consistent with LE including disrupted lung tissue with pleural disruption, expansion of alveolar spaces, and parenchymal hemorrhage.

The patient has experienced significantly improved pulmonary function as demonstrated by decreased ventilation requirements; at presentation requiring a volume control setting then post-operatively being transition to pressure support. This case demonstrates that resection of an abnormal lobe responsible for a V/Q mismatch can be safely performed and can benefit a child with a history CDH repair. Long term follow up will be needed to determine if these benefits will persist and to what degree the remaining lung compensates or develops emphysematous changes after resection.

Patient consent

Consent to publish the case report was obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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