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Secondary multicystic changes in the contralateral lung associated with Congenital Diaphragmatic Hernia

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ABSTRACT

In this report we present a case of extensive cystic changes of the lung that developed postnatally in an infant with right Congenital Diaphragmatic Hernia. The cystic changes occurred on contralateral side, in the setting of relatively low mean airway pressures. With gentle ventilatory techniques, Heliox, and intentional collapse of the affected lung, the cysts gradually resolved.

1. Introduction

Infants with Congenital Diaphragmatic Hernia (CDH) have multifactorial lung disease with associated poor oxygenation and ventilation. While mechanical compression from the herniated abdominal contents explains part of these issues, infants with CDH also have underlying abnormal lung parenchyma. CDH classically includes lung hypoplasia with an arrest in alveolar development which results in reduced gas exchange surface area, markedly thickened alveolar walls, and increased interstitial tissue [1]. Although more pronounced in the lung ipsilateral to the hernia, these same features can be recognized in both lungs [2]. The postnatal development of cystic changes in the lungs of patients with CDH is not reported in the literature. In this report, we present a case of secondary cystic changes in the left lung of a patient with right sided CDH.

2. Case report

Our case is a female infant born at 39 weeks to a 34 year old G1P0 mother with benign prenatal labs. Antenatal steroids were given once due to threatened preterm labor prior to 32 weeks. At 33 weeks, a right diaphragmatic hernia was noted on ultrasound, initially liver down. Follow up ultrasound at 36 weeks demonstrated liver and bowel in the right hemithorax. Apgars were 6, 6, and 7 at 1, 5, and 10 min, respectively. She was intubated after 50 s of CPAP, an enteric tube and PIV were placed, and she was transported to Rady Children's Hospital San Diego NICU.

She had significantly poor oxygenation and ventilation, initial blood gas: 7.06/94/55/-4 on 93% FiO2. Admission Chest radiograph (CXR) shown in Fig. 1. She gradually responded to High Frequency Oscillatory Ventilation (HFOV), inhaled nitric oxide, and Heliox (inhaled helium/oxygen mixture). Though she was close to meeting criteria for ECMO (Extracorporeal Membrane Oxygenation) on a few occasions, she did not ultimately require ECMO. The diaphragmatic hernia was repaired on DOL 20. Post-operative CXR is shown in Fig. 2. The right lobe of the liver was noted in the right hemithorax, however there was a reasonable amount of anterior diaphragm present, consistent with a Type C defect, and an 8 × 8cm Gore-Tex patch was placed. Her average mean airway pressure during mechanical ventilation was 7.5 cm H_2O, with a range of 6–11 cm H_2O.

Post-operatively, she continued to require iNO and heliox due to impaired oxygenation and ventilation. She developed a pneumothorax on the right and required a chest tube for 8 days. She was extubated and a trial of CPAP was done at 25 days of life, however at this time new cystic changes were noted in the left lung. Initially the cysts were small and detected on chest radiograph (Fig. 3). The cysts increased in size from DOL 26–28 and were confirmed on chest CT on DOL 30, shown in Fig. 4. The Chest CT showed extensive cystic changes of the left lung with the most central air-containing spaces surrounding the bronchovascular bundles/pulmonary interstitium. The majority of the left lung was involved with relative sparing of only the posterior medial left base. She was reintubated and underwent multiple cycles of HFOV on low mean airway pressures and heliox. Despite this, the cystic changes in the left lung continued to worsen, confirmed by repeat chest CT on DOL 51. Additionally, she developed a pneumothorax on the left that required a chest tube for 11 days. At this point, multiple discussions

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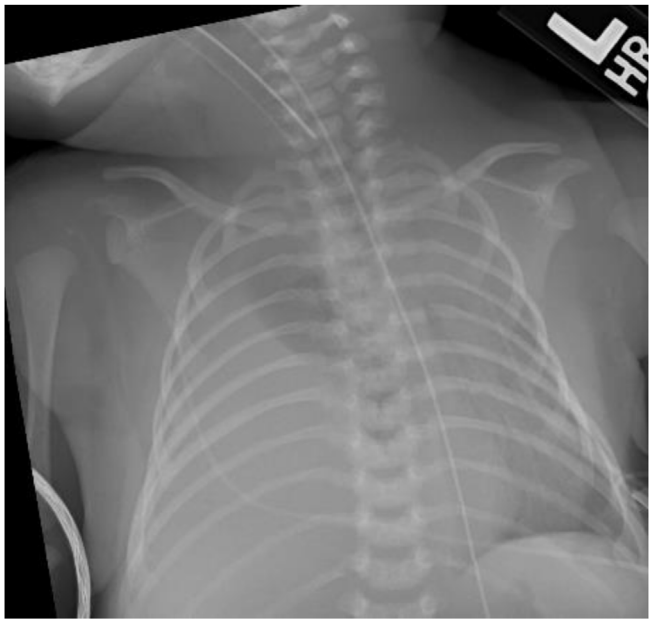

Fig. 1. Admission CXR.

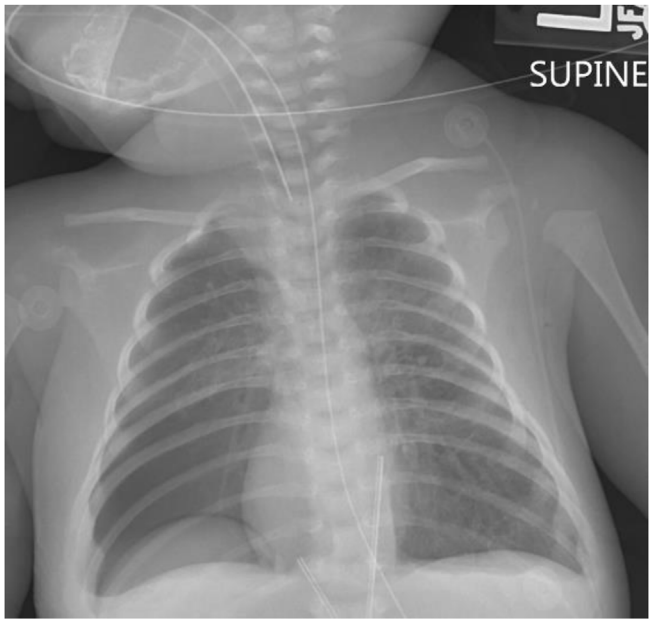

Fig. 2. CXR post-op CDH repair.

were had about the best course of action, as there was concern that the cysts would continue to worsen and cause further pneumothoraces. Left pneumonectomy was discussed, however there was concern that she would not be able to survive only with the hypoplastic right lung. To determine her ability to tolerate single lung physiology and as a potential therapeutic measure for the cysts, a trial of completely collapsing the left lung was undertaken. Multiple methods could be used to induce atelectasis of one lung, including selective mainstem bronchus

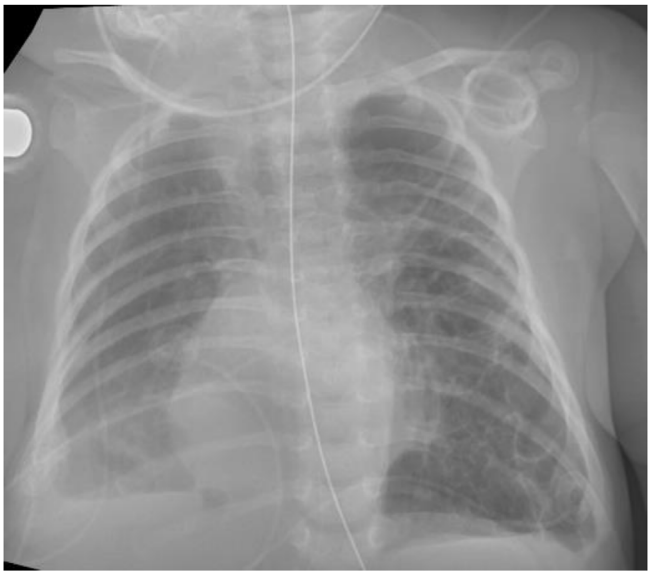

Fig. 3. CXR with cystic changes.

intubation, insertion of a temporary endobronchial blocker, and for neonates, prolonged lateral decubitus positioning. In order to avoid trauma to the airway, lateral positioning was chosen as the least invasive and most easily reversible intervention. She was placed left side down for 14 days to completely collapse the left lung and allow for healing (Fig. 5). Once the left lung was allowed to re-expand at DOL 67, cysts were no longer evident by CXR. Eventually, she was successfully extubated to CPAP at 3.5 months of age, and then gradually weaned to nasal cannula. She was discharged to home on oxygen at 6 months of age. No cysts were visible on CXR prior to discharge (Fig. 6).

She also developed significant pulmonary hypertension demonstrated by echocardiography. She transitioned from iNO to Sildenafil. She had a prolonged course of milrinone. A gastrostomy tube and fundoplication were performed at 3.5 months of age due to feeding difficulties. She had multiple infections throughout the admission, including bacteremia, pneumonia, urinary tract infection, and suspected endocarditis, all of which were successfully treated.

After discharge to home she was followed in Pulmonary and Cardiology clinics. She continues on home oxygen, baseline 0.5 L, 20 hours per day. The gastrostomy tube was removed when she reached feeding independence, just prior to 3 years of age. She has moderate to severe pulmonary hypertension currently on 3 medications. Outpatient CXR at 3 years of life demonstrated no cysts (Fig. 7). Her development is progressing well, and she is ahead in all areas except gross motor. She is bilingual and enjoys books.

3. Discussion

A literature search was conducted in January 2017: Pubmed, EMBASE, Web of Science, and Google Scholar were used to search Congenital Diaphragmatic Hernia and cystic lung, secondary cystic lung changes, acquired cystic lesions, and lung cysts. No citations were found that described the postnatal development of cysts in the lung for patients with CDH.

This is the first case that we are aware of in which such extensive cystic lung disease occurred postnatally in the lung contralateral to a

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