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Congenital Lobar Emphysema: Our Case Report in Gatot Soebroto Army Hospital

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Abstract
Emphysema is a disease of a lung condition due to airway and or alveolar abnormalities usually a chronic progressive lung disease caused by significant exposure to noxious particles or gasses that causes shortness of breath, but Congenital Lobar Emphysema is a rare malformation developmental of a lung, and it can cause severe respiratory distress in the newborn baby. Herein, we aimed to report our case of Congenital Lobar Emphysema.

Keywords: Congenital Lobar Emphysema, Newborn, Respiratory Distress

Case Report
A 3 days old female baby referred to our hospital with severe respiratory distress. At the previous hospital she was atermly delivered via spontaneous labor due to preterm premature rupture of membranes (PROM), APGAR 7/8, birth weight 2870 g, birth height 47 cm, thoracoabdominal breathing. We find an diminished and wheezed lung sound on her right chest. Her DOWNE score was 3/4, and she had been given CPAP with Flow 7 PPEP 7 FiO^2 30-50% for her respiratory distress, but not adequately helping. Her chest xray and CT showed hyperaeration and hyperinflation of right lung, with herniation of right lung to the left, opacities on her left lung, that indicated Congenital Lobar Emphysema. After several discussions, we decided to perform Wedge Resection. intraoperatively, we found emphysematous lungs at medial and superior lobes of the right lung. We resected a part of the medial and superior lobes of the right lung. She gained better at her breathing, her pneumonia deceased, and she came home after 7 days post surgery

Figure 1. Chest Xray pre-operation

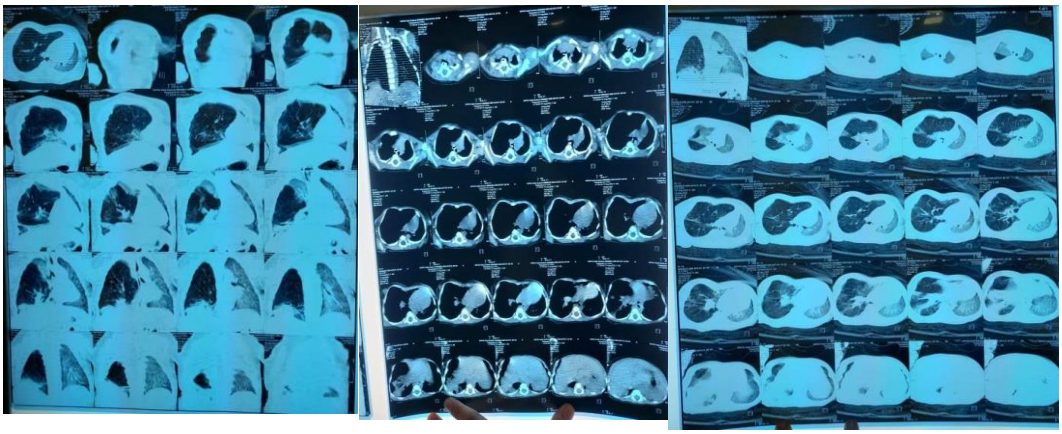

Figure 2-1. Chest CT Pre-operation

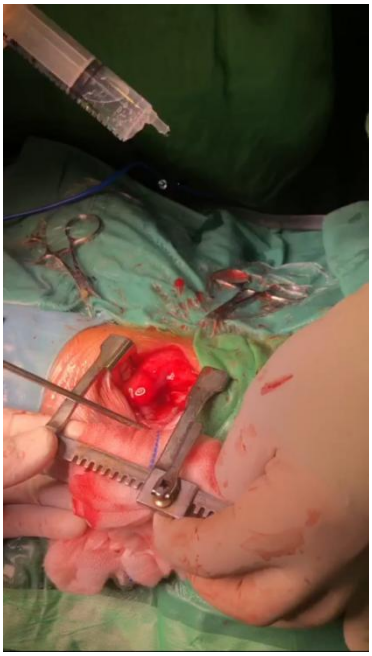

Figure 2-2. Intra Operation

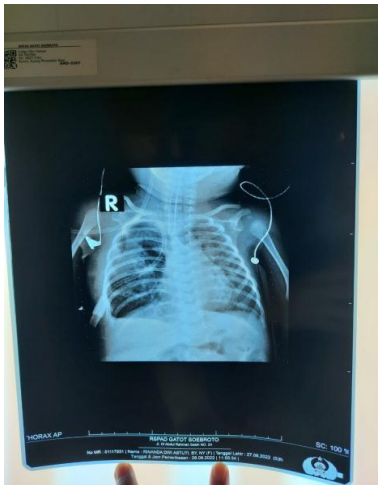

Figure 2-3. Chest X-ray Post Operation

Discussion

Congenital Lobar Emphysema, or congenital lobar overinflation, or infantile lobar emphysema[1-3], is a rare congenital malformation with a prevalence of 1 in 20,000 to 1 in 30,000[2]. We have a female patient in our hospital, but it is more common in males, and the male to female ratio is 3:1. One-third of cases are symptomatic at birth and nearly all of them are diagnosed in the first 6 months of life.[7-9] The most common observation of this disease is left upper lobe involvement (43%), followed by right middle lobe (32%) and right upper lobe (21%) involvement, similar with in this case at her right upper lung lobe. Lower lobe involvement (2%) is the rarest form.[7-11] In the literature, more than one lobe and bilateral involvement have been described. There are two types of CLE leading to hyperinflation of the lobes of the lung: 1) Intrinsic CLE, more like in this case, happens when there is an obstruction from something inside the lung, such as an abnormal airway tree or bronchial cartilage acting like a one-way valve, 2) Extrinsic CLE

happens when there is compression of the airway by a nearby structural abnormality (such as a bronchogenic cyst) that causes pressure on the airways[1].

CLE has some differential diagnosis:

a) Pneumothorax. They have similarities in their clinical presentations with respiratory distress, hyperresonance and decreased breath sounds on the affected lung, and the chest x-ray will show that the mediastinum deviates to the contralateral lung in tension pneumothorax but CLE has no compression to the diaphragm unlike pneumothorax[22]
b) Pneumonia. Pneumonia present with fever, cough, chest retractions, and could lead to cyanosis and the affected lung collapsed with consolidation. Pneumonia improves with antibiotics and did not have hyperinflation at the affected lung[23]
c) Congenital Cystic Adenomatoid Malformation. They have similarities in ultrasound finding which is highly echogenic mass but CLE has devoid cystic lesion compared to CCAM[21]
d) Congenital Diaphragmatic Hernia. Both of this diseases have similarities with hyperlucent on the affected lung, but CHD has gas-filled loops of bowel in the chest[24].

About 20% of CLE may associated with other congenital anomalies especially cardiac anomalies[4], such as Patent ductus arteriosus, atrial septal defect, ventricular septal defect, total pulmonary venous return anomaly, and tetralogy of Fallot are the most common congenital cardiac defects seen with CLE[14,16-20] . This patient has middle to large secondary ASD, right atrium dilatation, left pulmonary artery stenosis, which may be corrected after she fully recovered. Vascular abnormalities such as pulmonary arterial sling anomalies and abnormal pulmonary venous return anomalies may cause CLE[12-15]

Early management of CLE improves outcome and avoid life-threatening complications. Management CLE divided into two categories, asymptomatic and symptomatic patients. If the patient is asymptomatic, the parents should be guided about conservative management. There should be regular follow-up and observation, and the parents should be advised of the steps that should be taken if the patient starts to show symptoms. If the patient has severe symptoms, parents should be guided about the possibility of surgery (lobectomy) as well as its prognosis and lifestyle changes accordingly. The parents should be provided with educational material about the disease. Flow charts, diagrams, and videos should be used to explain each and every aspect of the disease, treatment, and lifestyle changes until the parents fully understand everything and are satisfied with the provision of care[5]. In this patient, because of her severe respiratory distress, was decided to perform lobectomy.

For better screening for this congenital disease, prenatal evaluation can be very useful, from USG until MRI prenatally. Neonatal echocardiography also should be performed to evaluate any congenital heart disease[24]. This patient did not have proper antenatal care during her pregnancy due to economic matters

Conclusion

A good clinical and radiological understanding is required to diagnose this rare & dangerous anomaly which may mimic other causes of respiratory distress. Radiologic investigations as adjunct are very helpful in making the diagnosis. Early identification and proper management can lead to good improvement of the patient's life

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