

Congenital Lobar Emphysema : a case Report

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Abstract

This case is about a neonate with respiratory distress and primary diagnosis of Pneumothorax who had Congenital Lobar Emphysema. This case was the basis to review the topic of Congenital Lobar Emphysema (CLE) and the corresponding literature, considering that this is an uncommon disease and although congenital lobar emphysema is rare, clinical awareness of this condition is important for early diagnosis and effective surgical treatment. Due to the severity of respiratory symptoms, congenital lobar emphysema often requires surgical treatment in the early stages of life.

Key words: Neonate, Congenital Lobar Emphysema, Respiratory Distress

Introduction

Respiratory diseases are one of the common causes of NICU mortality (1, 2). These diseases are divided to two major groups: acquired and congenital. Congenital lung developmental syndromes show themselves with primary pulmonary hypertension (PPHN) and respiratory distress. These malformations are congenital lobar emphysema and thoracobronchial tract cystic anomalies like sequestration and bronchogenic cysts. Congenital lobar emphysema is a very rare congenital cystic malformation of the lung that can cause acute respiratory distress in early life (1, 3).

This case report is about a 27-days-old girl who was transferred to Emam Reza hospital NICU after being diagnosed with Pneumothorax in an outlying hospital emergency department and at least has been operated because of congenital lobar emphysema.

Patient Introduction:

A 27 –days-old girl who was 1800 grams, was transferred to Emam Reza hospital NICU, and has been treated with oxygen mask.

Infant has been born by cesarean section with history of PROM and 4 days chorioamnionitis in this center. At first she was 1580 grams and because of grunting and tachypnea admitted in NICU. In the 3d day she has an apnea attack and ventilated, after 3 days she became better and extubated. At the day 14th she was discharged from NICU with good health.

First diagnosis of her situation was sepsis, asphyxia and apnea.

After 13 days she was transferred to NICU again because of respiratory distress. In first step of work up Pneumothorax has been diagnosed, and although

we suspected to lobar emphysema and other cystic malformations. Computed tomography scan of the chest revealed congenital lobar emphysema, so infant referred to Shykh hospital for surgical management



Figure 1



Figure 2

DISCUSSION AND CONCLUSION:

Congenital lobar emphysema is characterized by over inflation of pulmonary lobe (gas trapping) and is caused by localized bronchial obstruction (1, 2). The disease may result in severe respiratory distress in early infancy.

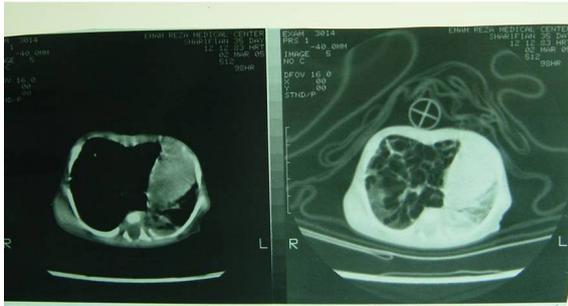


Figure 3



Figure 4



Figure 5

Over distension of the airspaces within a pulmonary lobe is associated with air trapping and compressive changes in the remainder of the lung (2). Mediastinal shift away from the increased volume results in compression of the contra lateral lung (1, 2).

CLE almost always involves one lobe, with 47 % rates of occurrence in left upper lobe (2). In some cases bilateral involvement happened with sever respiratory distress, which needs urgent surgical management (3).

CLE is most often detected in neonates or with in uterus ultrasound; however, less severely affected patients may present in infancy or childhood (4, 5).

Other Problems to be considered: Bronchial mucous plug with associated hyper aeration
Extrinsic bronchial compression, Agenesis/hypo genesis of contra lateral lung
Bronchial

hypoplasia with air trapping peripherally
Congenital cystic adenomatoid malformation , Pneumothorax (3, 6).

CX R finding are: A large, hyper lucent lung with attenuated but defined vascularity is observed. Compressed remaining lung on that side, flattened hemi diaphragm, and widened intercostal spaces also are seen. An involved lung is seen herniated across the anterior midline (7, 8).

CT scan can provide detail about the involved lobe and its vascularity, as well as about the remaining lung (1).

CT scan shows a hyper lucent, hyper expanded lobe (attenuated but intact pattern of organized vascularity) with midline substernal lobar herniation and compression of the remaining lung. Usually, the mediastinum is significantly shifted away from the side of the abnormal lobe (9, 10).

CLE has a good prognosis after medical and surgical management (11, 12).

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