A Case of Congenital Lobar Emphysema in the Middle Lobe

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ABSTRACT

Congenital lobar emphysema (CLE) is a rare cause of respiratory distress during the neonate period. It is characterized by overinflation of pulmonary lobe, most commonly the left upper lobe or the right middle lobe. We report a case of a 21-day-old baby with the severe respiratory distress. Diagnosis of CLE was made by computed tomography scan and chest X-ray and confirmed by histopathological studies. We would like to draw attention to this rare condition, and discussing their clinical features and management.

Key words:
Congenital lobar emphysema, middle lobe, respiratory distress

INTRODUCTION

Congenital lobar emphysema (CLE) is uncommon event in neonates. It is characterized by overinflation of pulmonary lobe. However, this affection can cause a severe respiratory distress with high level of mortality or result in serious morbidity and disability. The exact etiology of disease is not known. The prognosis depending of the precocity of management means that the clinicians must have a high index of suspicion for CLE in the context of respiratory distress in neonate period, and ask for imaging techniques.

CASE REPORT

A 3-week-old male baby weighing 2300 g was referred to our neonatal intensive care unit for asphyxia with respiratory distress not responding to medical management in regional hospital. His mother did not have any follow-up during her pregnancy. He was delivered as a full term with delayed cry and respiratory distress.

Physical examination revealed a tachypneic baby with flaring of alae nasi and sub costal retraction, the cyanosis was generalized (oxygen saturation spO2 was 70% in air). On examination of the respiratory system, decrease breath sound on the left hemithorax was noted. The cardiac auscultation was normal.

The chest X-ray showed hyperinflation on the right side, and right basithoracic opacity [Figure 1]. computed tomography (CT) scan of the thorax supported the X-ray [Figure 2]. There was hyperinflation on the right middle lobe with tracheal and mediastinal shift to the left side. The blood parameters were normal. On the echocardiography, there was no evidence of congenital cardiac anomalies.

After hemodynamic stabilization of the child, surgical intervention was performed. The right middle lobe looked emphysematous at time resection [Figure 3]. Post-operative chest X-ray showed expansion of the right upper and lower lobes, with no emphysema or mediastinal shift.

Histopathological examination of the excised right middle showed alveolar distension without fibrosis [Figure 4].

The child was discharged 7 days post-surgical intervention, and he was seen as an out-patient for follow-up. He had normal O_2 saturation in room air and he was feeding well.

DISCUSSION

CLE is a rare cause of respiratory distress during infancy and neonate period.[1] It is more common among males than females. The left upper lobe is most commonly affected but any lobe can be involved.[1,2] Although bilateral involvement has been reported.[3] In our case, the right middle lobe is involved.

It is due to hyperinflation of lung lobe, collapse of adjacent lobe, compression of controlateral normal lung and mediastinal shift.[1] The etiology of CLE is unknown. The underlying defect is often inadequad cartilaginous support

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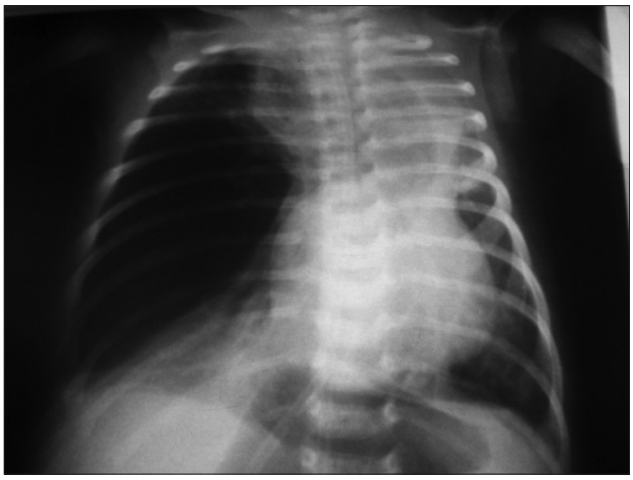

Figure 1: Chest radiography: Hyper clarity of right lung field and a shift of the mediastinum to the left

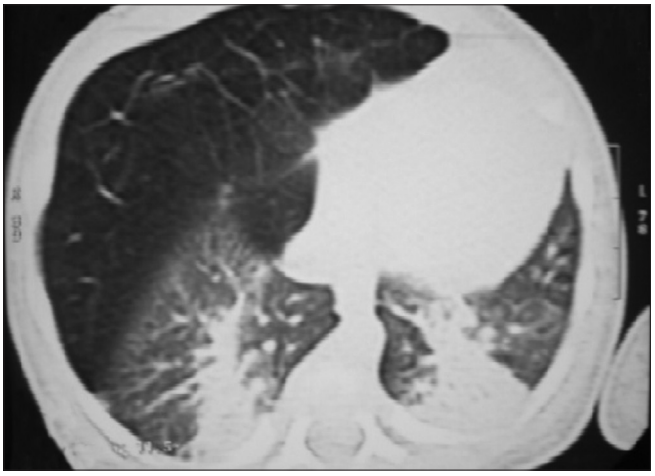

Figure 2: Pre-operative computed tomography scan: Emphysema lung middle lobe

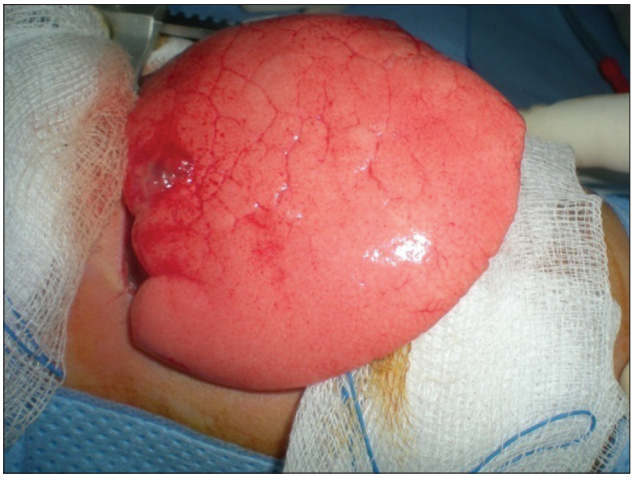

Figure 3: Gross specimen of the emphysamatous right middle lobe at time of intervention

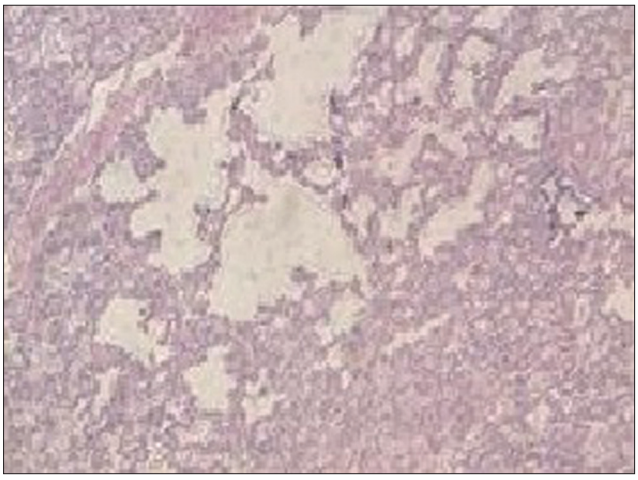

Figure 4: Histological section of the specimen. Structural anomaly of the bronchus with drainage emphysematous alveoli that appear collapsed after exsufflation

of the bronchus and air way collapse creating a "ball-valve" mechanism and air trapping during expiration. However, a definitive causative agent may not be identified in about 50% of cases.[1,2]

The severity of symptoms of CLE vary, depending on the size of the overdistended lobe, compression of lung tissue around it and the degree of displacement of the mediastinum.[4] Symptoms usually appear at birth in 33% of cases, such as our case, and 50% during the 1st month and most before 1 year of age.

It should be noted that the importance of this case is twofold: The middle lobe localization because the most commonly affected lobe is the left upper 42% followed by the right middle, and the importance of timely diagnosis avoiding confusion with the presence of pneumothorax, pneumomediastinum, because the initial management was in the regional hospital.

The diagnosis of CLE was made by postero-anterior and lateral chest X-ray, which shows the classical signs of lobar hyperinflation, mediastinal shift, and sometimes controlateral atelectasis. CT scan can provide details about the involved lobe and it vascularity as well as information about the remaining lung, excluding a vascular abnormality and other conditions that might be confused with CLE such as pneumothorax, diaphragmatic herniation, or cystic adenomatoid malformation.

In our case, unfortunately baby's mother did not have any follow-up during her pregnancy, and he was managed initially in regional hospital. However, the diagnosis antenatal is possible in the same situations.

Different congenital thoracic malformations are frequently detected on routine antenatal ultrasound. However, a definitive diagnosis cannot usually be established antenatally

with absolute accuracy because of either its low prevalence in utero or the increased echogenecity of the lungs, which could be too subtle to be appreciated in utero. The main fetal sonographic features of CLE include a bright echogenic lung with or without cystic lesions, a mediastinal shift, and the associated cardiovascular anomalies.[5] Magnetic resonance imaging should differentiate CLE from cystic adenomatoid malformation and bronchopulmonary sequestration because of the former's homogeneity intact lung structure with stretched hilar vessels.[6] The role of this imaging techniques pose a challenge to diagnosis and underscore the need for continued postnatal evaluation and management.

The management of CLE is usually resection of the affected lobe,[7] but in asymptomatic and mildly symptomatic patients, conservative approach is warranted.[8]

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