Congenital Lobar Emphysema- Exploring the Myth!

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Abstract: A case of congenital lobar emphysema is reported in an infant who was otherwise diagnosed to have viral bronchiolitis in the immediate admission.

Keywords: Infant, Respiratory distress, Lung malformation, CLE, Female, Lobe excision.

INTRODUCTION

Congenital lobar emphysema is a rare malformation of the lower respiratory tract characterised by hyperinflation of one or more of the pulmonary lobe [1, 2]. The prevalence is of 1 in 20,000 to1 in 30,000 live births [3]. Male sex seems to be more frequently affected than the female one (3:1) [4]. Privileged localisation is at the pulmonary left upper lobe (43%), followed by the right middle lobe (32%)and the right upper lobe (20%) of cases [5].

CASE REPORT

A 3- months old-female infant weighing 4.5 kg was brought to paediatric emergency room because of hurried breathing. On physical examination, the patient had respiratory distress with bilateral rhonchi at the chest auscultation. A diagnosis of bronchiolitis was made. The baby was managed with nebulised epinephrine and got admitted to the ward. After a transitory improvement, and inspite of multiple administration of nebulised epinephrine, the clinical picture deteriorated and FiO2 requirement increased. A chest radiogram showed hyperinflation of the pulmonary left upper lobe, with displacement of the mediastinum to the right side (Figure **1**). At the computed tomography, the above findings were accompanied by sub-segmental collapse of the left lower and right upper lobes (Figure **2**). There were no any other associated anamolies including cardia.

An excision of the emphysematous lobe, which measured 9.5 x 7 x 2 cm (Figure **3**), was performed.

At the histological examination, the sample showed lung parenchyma with dilated and over-distended, air filled alveolar ducts and alveoli; a picture was consistent with diagnosis of CLE (Figure **4**).

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DISCUSSION

In its classic, idiopathic form (that is, not secondary to extrinsic compression), CLE is thought to arise from obstruction of the lobar bronchus. Theoretically, name congenital lobar hyperinflation would be more appropriate because normally formed acini are seen with greatly over distended alveoli but without tissue destruction, this being in contrast to acquired pulmonary emphysema. The prevalence of CLE is of 1per 20,000-30,000 live births. The male sex appears to be more affected than female one in the ratio of 3:1. Usually, the clinical manifestation become apparent in the neonatal period, but in some cases they are delayed for as long as 5-6 months.

The pulmonary upper lobes are most frequently involved, the left upper lobe being the commonest site of localisation. The diseased lobe is essentially non-functional because of the dysventilation and the over – distension of the alveolar structures. Due to the displacement of the mediastinum and the consequent compression, also the function of the contralateral lung may be compromised [4].

In a number of cases, such as that here described, antenatal ultrasound is not able to disclose CLE; in some cases, images suggestive of pulmonary malformations may disappear during serial antenatal evaluations, while becoming again apparent at postnatal stage [5].

Typically, the chest radiogram demonstrates a radiolucent and the displacement of the mediastinum [6, 7]. CT is performed to confirm the diagnosis. Histology supports the radiologic evidences in distinguishing CLE from other congenital lesions of the lungs, such as lobar sequestration and cystic adenomatoid malformations. In most of the cases, the patients become symptomatic within six months of life, and usually present with respiratory distress, cyanosis and recurrent pulmonary infections. In a minor number of cases the lesion does not give symptoms and

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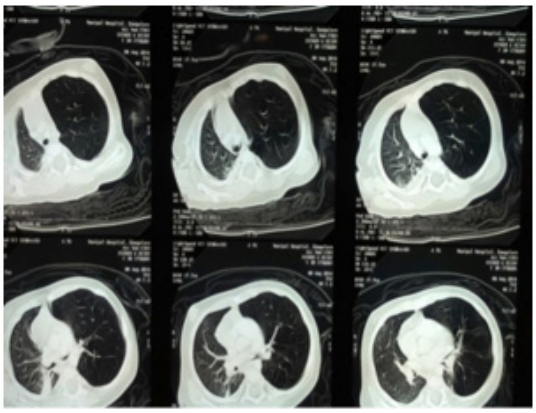
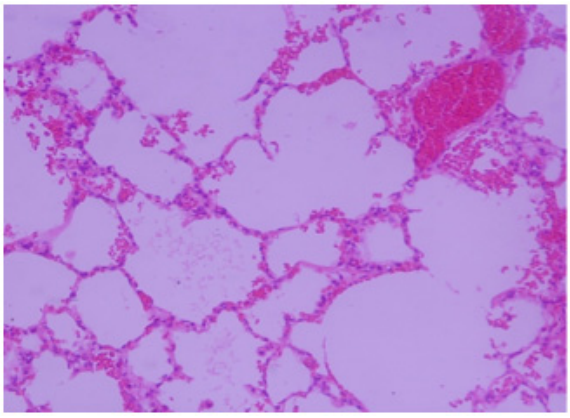
(1)



(2)

Figure 1: Hyperinflation of left upper lobe of Lung.
Figure 2: CT chest demonstrating hyperinflation of upper lobe of left Lung.

(3)



(4)

Figure 3: Specimen of Lung (L) upper lobe.
Figure 4: Microscopy demonstrating the lung parenchyma with dilated and over Distended airfilled alveolar space.

remains undiagnosed. CLE is necessarily managed by excision of the affected lobe. After surgery the prognosis is generally good, with compensatory pulmonary growth seen on the same side [8, 9].

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