

Congenital Lobar Hyperinflation - A rare anomaly misdiagnosed as bronchiolitis: A case report

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Abstract

Congenital Lobar Hyperinflation is an overinflation of one or more than one lobes of the lung and is caused by an abnormal development of broncho-pulmonary segment. We report a case of a 2-month-old female who presented in outpatient department with complaints of fever, cough and difficulty in breathing. This case was seen in January, 2018. Congenital Lobar Hyperinflation was diagnosed on chest x-rays and Computed Tomography (CT) scan. Immediate surgical excision (lobectomy) of the affected lobe was conducted.

Keywords: Congenital Lobar Hyperinflation, Congenital Lobar Emphysema (CLE) Congenital Heart Disease (CHD)

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Introduction

Congenital Lobar Hyperinflation, Congenital Lobar Emphysema or Congenital Lobar Overinflation is a postnatal over distension of one or more than one lobes of a histologically normal lung.¹ Multilobar presentation has been reported previously, however, it is a rare finding.² Some of the cases are autosomal dominant, while others can be sporadic in nature.³ It is either caused by an aberrant development of the broncho-pulmonary segment which leads to a deficiency of the focal cartilage of the tracheobronchial tree leading to a regional airway collapse or it may be caused by an external compression resulting from aberrant vessels. Prevalence is stated to be around 1 in 20,000 to 1 in 30,000 deliveries.⁴ Males are affected twice as commonly as females.⁵ In 30% of the cases, it is associated with Congenital Heart Disease.⁵ The over distended lung causes collapse of the ipsilateral lobe and leads to a mediastinal shift on the opposite side, resulting in lung hypoplasia. If the condition is not so grave, a conservative approach is preferred but if a severe entity

is diagnosed, surgical methods are opted.

We report a case of Congenital Lobar Hyperinflation in an infant with respiratory symptoms.

Case Report

A 2-month-old female presented at the outpatient department (OPD) with complaints of fever and cough for two days and breathing difficulties for a day at Sharif Medical City Hospital, Lahore in January, 2018. Fever was intermittent, low grade and was not associated with rigors or chills but was relieved by oral medications. However, cough and respiratory distress persisted despite treatment with antibiotics and bronchodilator nebulizers.

Patient was delivered 2 weeks preterm and had neonatal jaundice along with sepsis, which was treated with appropriate medication. Patient also had nasal flaring, subcostal recessions and respiratory distress. Her respiratory rate was 69 breaths/min, heart rate was 122 beats/min and blood pressure was 70/40 mmHg. Inspection of the chest revealed it to be elliptical in shape with intercostal and subcostal recessions, percussions showed hyper-resonance on both sides and bilateral rhonchi were found on auscultation. As a result of these findings, patient was treated as a case of bronchiolitis. Medications included nebulizers with bronchodilators and a 3rd generation Cephalosporin along with Amikacin. These were continued for 9 days but no improvement was seen. Chest X-ray revealed a left hypertransradiant hemithorax with an overinflated upper lobe; a retrosternal herniation with attenuation of the vascular marking and contralateral mediastinal shift was also visualized (Figure-1). Chest X-ray was repeated and a radiologist was consulted for comparison of both the x-rays. Computed tomography (CT) scan of the chest was advised due to a suspicion of Congenital Lobar Emphysema as both X-rays depicted an overinflated left upper lobe herniating retrosternally along with a contralateral mediastinal shift. Hence, CT scan confirmed the presence of a Congenital Lobar Hyperinflation (Figure- 2).

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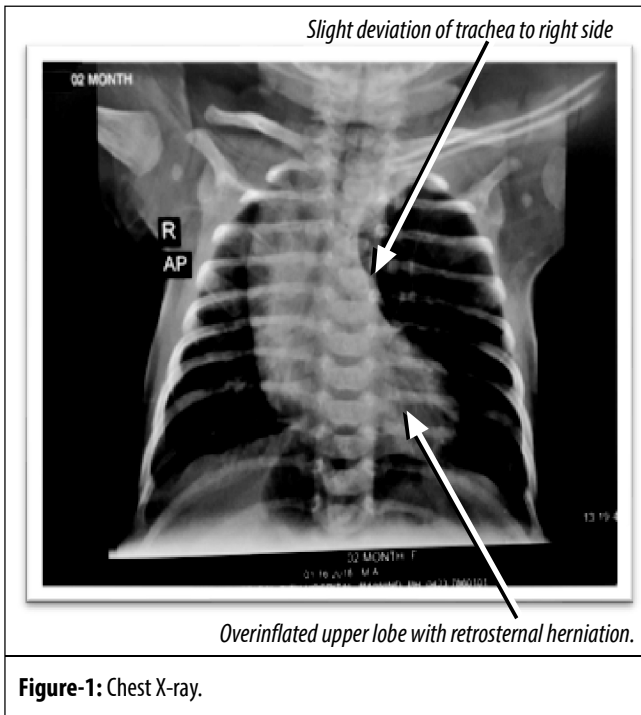


Figure-1: Chest X-ray.



Figure-2: Computed Tomography Scan.

After this, a paediatric surgical consult was obtained and an immediate lobectomy of the affected lobe was performed. Post-operative care was provided for 4 days after which the patient was discharged with marked improvement.

On post-operative follow-up at 11 days, no respiratory distress was noted. On examination her respiratory rate (52 breaths/min), blood pressure (84/49 mmHg) and axillary temperature (36.5C) was normal.

Patient was called for a follow-up after 2 months of surgery. On examination the patient had a normal feeding pattern with an adequate weight gain. No respiratory symptoms were seen and a decrease in the right shift of the mediastinum was observed after lobectomy. The respiratory rate, blood pressure and body temperature were normal.

Discussion

Congenital lobar hyperinflation usually manifests during the first 3 months of infancy. It involves an overdistension of one or more than one lobes of the lungs. There are two possible pathophysiological mechanisms which can provide an explanation for the condition.⁶ Firstly, an obstructive agent acts as a ball valve which allows the air to enter on inspiration but prevents its expulsion on expiration and secondly, it can be due to collaterals that bring in the air behind the obstruction. These collaterals consist of 'pores of Kohn' and other direct accessory broncho-alveolar connections called 'canals of Lambert'. Emerging data suggests that the minor transcription errors in the fibroblast growth factor 10, hedgehog signalling and Homeobox protein Nkx 2.1 pathways may lead to a localized anomaly of the bronchial cartilage resulting in Congenital Lobar Emphysema as these are responsible for the branching morphogenesis of the lungs.⁷

Clinical manifestations can be delayed for 5-6 months in 5% of the patients.⁸ In most of the prenatally diagnosed cases, patients usually do not show any symptoms at birth and occasionally, it may remain undiagnosed until the school age. Symptoms may vary from mild respiratory distress to acute respiratory failure with marked tachypnoea, severe recessions, nasal flaring, grunting and late cyanosis or it may also progress to pneumonia.⁹ Diagnosis is made on chest X-rays, which reveals an over distended affected lobe often accompanied by mediastinal shift, herniation and deviation of the trachea from midline. CT scan and Magnetic resonance imaging can also be

used to localize the vessel which may cause an extra luminal compression. This condition can be misdiagnosed as a cystic lesion of the lung for example a cystic adenomatoid malformation or a pneumothorax.

There is some controversy regarding the treatment choice for Congenital Lobar Hyperinflation.³ Surgical removal is the most common treatment modality with operative mortality rate noted to be around 3 to 7%.¹⁰ Endoscopic management of Congenital Lobar Emphysema has also been reported in the literature.¹¹ Some forms of conservative management are recommended in patients who feed properly and do not show any signs of respiratory distress. These measures include light ventilation procedures which if successful will result in fewer emergency surgeries. Antenatal diagnosis can be done with ultrasonography, which was not done in our case.¹²

While treating respiratory distress, positive pressure ventilation should be used with caution due to the tendency to undergo an auto-positive end-expiratory pressure. When Congenital Lobar Hyperinflation progresses to cause a mediastinal shift and worsening of symptoms, an open lobectomy is indicated.

Conclusion

There should be a high index of suspicion if an infant presents with respiratory distress and cystic lesion of the lung on a chest X-ray.

Informed consent was obtained from the parents of the patient for publication of the case report.

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Conflict of Interest: None to declare.

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References

1. Cockburn F, Carachi R, Goel KN, Young D. Respiratory System. In: Cockburn F, Carachi R, eds. *Children's Medicine and Surgery*. 1st ed. London, UK: Arnold, 1996; p.143.
2. Özçelik U, Göçmen A, Kiper N, Dogru D, Dilber E, Yalçın EG. Congenital lobar emphysema: evaluation and long-term follow-up of thirty cases at a single center. *Pediatr Pulmonol* 2003; 35: 384-91.
3. Nath MP, Gupta S, Kumar A, Chakrabarty A. Congenital lobar emphysema in neonates: Anaesthetic challenge. *Indian J Anesth* 2011; 55: 280-3.
4. Townsend CM, Beauchamp RD, Evers BM, Mattox KL. *Pediatric Surgery*. In: Arons CB, Adams AB, Adams C, eds. *Sabatton Textbook of Surgery*. 1st ed. New Delhi: Elsevier, 2016; p.186.
5. McIntosh N, Helms P, Smyth R, Logan S. The Newborn. In: McIntosh N, Helms P, Smyth R, Logan S, eds. *Forfar and Arneil's Textbook of Pediatrics*. 7th ed. China: Churchill Livingstone Elsevier, 2009; p.257-8.
6. Kumar V, Abbas AK, Aster JC. The Lung. In: Alpers CE, Anthony DC, Chang A, eds. *Robbins & Cotran Pathologic Basis of Disease*. 9th ed. Faridabad: Elsevier, 2014; p.678.
7. Sadaqat M, Malik JA, Karim R. Congenital lobar emphysema in an adult. *Lung India* 2011; 28: 67-9.
8. Kliegman RM, Stanton BF, Schor NF, Geme JS. Emphysema and Over inflation. In: Kliegman RM, Stanton BF, Schor NF, Geme JS, eds. *Nelson Textbook of Pediatrics*. 19th ed. Philadelphia: Elsevier, 2011; p.1461.
9. Brunnicardi FC, Andersen DK, Billiar TR, Dunn DL, Hunter JG, Matthews JB, Pollock RE. *Pediatric Surgery*. In: Brunnicardi FC, Andersen DK, Billiar TR, Dunn DL, Hunter JG, Matthews JB, Pollock RE, eds. *Schwartz's Principles of Surgery*. 9th ed. USA: McGraw-Hill, 2010; p.1418-9.
10. Andrade CF, da Costa Ferreira HP, Fischer GB. "Congenital lung malformations." *J Brasileiro de Pneumologia* 2011; 37: 259-71.
11. Phillipos EZ, Libsekal K. Flexible bronchoscopy in the management of congenital lobar emphysema in the neonate. *Canadian respiratory journal*. 1998;5:219-21.
12. Eber E. Antenatal diagnosis of congenital thoracic malformations: early surgery, late surgery, or no surgery? *Semin Resp Crit Care Med* 2007; 28: 355-66.