Congenital Lobar Emphysema: A Commonly Misdiagnosed Entity

Dr. Kundan Mittal, Dr. Gurpreet Kaur, Dr. Joginder Silayach, Dr Anupama Mittal

Department of Pediatrics, Pt. B.D.S PGIMS, Rohtak, Haryana, India

Abstract

Congenital Lobar Emphysema (CLE) is a rare cystic malformation of the lung which is often misdiagnosed and managed as pneumothorax. We are describing a case of a 45 days old baby who presented to the emergency department with sudden onset of respiratory distress. On the basis of the chest X-ray (CXR) pneumothorax was suspected and an intercostal drainage (ICD) tube was inserted. But clinical and radiological non-improvement even after placement of an ICD tube raised the suspicion of CLE which was later confirmed by CT scan. Despite the opportunity to diagnose CLE on a simple investigational modality like CXR, cases of CLE tend to be misdiagnosed as pneumothorax very commonly and a high index of suspicion is required by the treating paediatrician.

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Key Words: Congenital lobar emphysema, CXR-Chest X-ray, ICD-Intercostal drainage tube

INTRODUCTION

Congenital Lobar Emphysema (CLE) is one of the rare cystic malformations of the lung causing over aeration of one or more lobes of a histologically normal lung. With an incidence of 1 in 70,000 to 1 in 90,000 live births most of the cases present in the neonatal period as a life threatening respiratory distress due to compression atelectasis, mediastinal shift, hypoxia, and associated hypotension [2]. The diagnosis may however be delayed for as long as 5 to 6 months in 5% of the patients [1]. CLE is generally misdiagnosed and managed as pneumothorax of the lung. Herein we report a case of a 45 days old child with life-threatening respiratory distress who was initially diagnosed as pneumothorax and managed in emergency by placement of an intercostal drainage (ICD) tube which was subsequently proved to be a case of CLE.

CASE REPORT

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45 days old exclusively breastfed male child presented to the Paediatrics emergency department with the compliant of cough for last 2 days and acute onset of respiratory distress since last 6 hours. There was no history of fever, cyanosis or seizures. At the time of admission the baby had tachypneoa and tachycardia with intercostals retractions and subcostal in drawing. However the blood pressure, temperature and capillary refill time were found to be normal. The oxygen saturation (SpO2) on room air was 87% which improved to 91% with oxygen by mask. The trachea and the apex beat were shifted to the right side with a hyper-resonant percussion note and a decreased breath sounds on the left side. The blood gas analysis revealed a pH of 7.39, PO2 of 75.4 mm Hg, PCO2 of 41.1 mm Hg, SpO2 of 92.1% and HCO3 of 19.3 mmol. Bedside Chest X-Ray (CXR) revealed an increased hyperlucency of the left lung fields and shifting of the mediastinum to the right side [Fig.1]. A provisional diagnosis of pneumothorax was made and an ICD tube was inserted.

The patient's condition did not improve post ICD tube insertion. A repeat CXR done revealed a similar picture with non expansion of the lung. The persistent clinical and radiological findings even after the placement of an ICD tube raised the suspicion of CLE. An urgent CT scan of the thorax was done, which revealed a hyperexpansion of left upper lobe with compression of the left lower lobe and the right upper and lower lobes with shifting of trachea and mediastinum towards right side [Fig.2]. A final diagnosis of CLE of the left

upper lobe was confirmed. A surgical excision of the emphysematous lobe was done, after which the patient recovered completely. A post operative CXR revealed expansion of the previously collapsed lung lobes. Biopsy of the resected lobe revealed distension of the alveolar acini with the absence of cartilage.

DISCUSSION

A rare congenital cystic malformation of the lung, CLE is characterized by normal architecture with lobar over aeration associated with respiratory distress secondary to partial obstruction of the bronchus via the ball-valve effect. It is also known as congenital lobar over inflation or infantile lobar emphysema [4,5]. This disease is attributed to a congenital deficiency of the bronchial cartilage, an external compression by the aberrant vessels [6], a congenital Cytomegalovirus infection [7], bronchial mucosal flaps and kinking have been described as its possible etiologies. It has been reported that this disease is more common in male children, being usually unilateral and affecting the left upper lobe more often (43%), followed by the right middle lobe (32%), although a bilateral involvement is not unknown [8].

The affected lobe is non-functional due to over distention and air trapping. In addition, there is a compression atelectasis of the ipsilateral normal lung leading to respiratory compromise. With further distension, the mediastinum is shifted to the contralateral side, resulting in impairment of bilateral lung function. A radiolucent lobe and a mediastinal shift are revealed by a radiographic examination. This condition is often confused with pneumothorax of the lung and subsequently managed by the placement of an ICD tube in emergency situation. A series of 3 cases which presented with respiratory distress were mistakenly diagnosed as pneumothorax and an ICD was inserted in all the 3 cases, but later on, all turned out to be CLE [9]. In another series of 10 cases of CLE, a correct diagnosis was made only in 4 out of the 10 cases on chest radiograph before referral, even though the diagnosis could be arrived at in all the 10 cases on the basis of the initial radiographs [10].

The basic investigation in congenital lobar emphysema is a readily accessible investigation CXR. Chest radiograph shows mediastinal shift and hyperinflation that is often confused with pneumonia and pneumothorax, even resulting in wrong placement of a chest drain. But non-improvement of the distress and non expansion of the lung are eye openers for a treating paediatrician [3]. A careful examination of the X-ray film will provide a diagnostic

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clue showing the presence of bronchovascular markings in CLE which are absent in the cases of pneumothorax [11].

CT scan is another diagnostic modality which shows the abnormally narrowed bronchus, hyperinflated left upper lobe and the collapsed lower lobe. All 3 lobes of the right lung can be distinctly made out in the CT scan thereby excluding a hypoplastic right lung. In addition, the CT scan also excludes a vascular abnormality and other conditions that might be confused with CLE, such as pneumothorax, pneumatocoele, a diaphragmatic hernia or a cystic adenomatoid malformation [12].

CONCLUSION

In spite of the development of advanced diagnostic techniques, CLE is still a diagnostic dilemma for the treating pediatrician although a provisional diagnosis can be conveniently established on the basis of CXR. But it requires a high index of suspicion in children presenting with a sudden onset of respiratory distress. A possibility of Congenital Lobar Emphysema should always be considered before inserting an Intercostal drainage tube in a suspected case of pneumothorax.

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REFERENCES

[1] al-Salem, AH., Adu-Gyamfi, Y., Grant, CS. (1990) Congenital lobar Emphysema. Can J Anaesthe 37, 377-379.

[2] Cote, CJ. (1978) The anesthetic management of congenital lobar emphysema. Anesthesiology 49,296-298.

[3] Boas, S., Winnie, GB. (2007) Emphysema and Overinflation. In: Kliegman RM, Behrman RE, Jenson HB, Stanton BF. Nelson textbook of pediatrics 18th edition. Elsevier publishers, 1779-1780.

[4] Stocker, JT., Drake, RM., Madewell, JE. (1978) Cystic and congenital lung disease in newborn. Perspect Pediatr Pathol 4, 93-154.

[5] Landing, BH., Dixon, LG. (1979) Congenital malformations and genetic disorders of the respiratory tract (larynx, trachea, bronchi and lungs). Am Rev Respir Dis 120, 151-185.

[6] Absher, DR., Kriss, VM., Cotrill, CM. (1999) Lobar emphysema due to anomalous aortic origin of the left pulmonary artery. Cardiol-Young 9, 327-330.

[7] Carrol, ED., Campbell, ME., Shaw, BN., Pilling, DW. (1996) Congenital lobar emphysema in congenital cytomegalovirus infection. Pediatr-Radiol. 89, 900–902.

[8] Ozcelik, U., Gocmen, A., Kiper, N., Dogru, D., Dilber, E., Yalcin, EG. (2003) Congenital lobar emphysema: evaluation and long-term follow-up of thirty cases at a single centre. Pediatr Pulmonol 35, 384–391.

[9] Tempe, DK., Virmani, S., Javetkar, S., Banarjee, A., Puri, SK., Datt, V. (2010) Congenital lobar emphysema: pitfalls and management. Annals of Cardiac Anesthesia 13, 53- 58.

[10] Bappal, B., Ghani, SA., Chaudhary, R., Sajvani, MJ. (1996) Congenital lobar emphysema: a review of 10 cases. Indian-J-Pediatr. 63, 801-808.

[11] Guidici, R., Leao, LE., Moura, LA., Wey, SB., Ferreira, RG., Crotti PL. (1998) Polyalveolosis: pathogenesis of congenital lobar emphysema? Rev Assoc Med Bras 44, 99-105.

[12] Joazlina, ZY., Wastie, ML., Bruyne, JAD. (2005) Congenital lobar emphysema: a diagnostic challenge. JHK Coll radiology 8, 105-108.

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Figure 1. Chest X-Ray (CXR) reveals an increased hyperlucency of the left lung fields and mediastinal shift to the right side

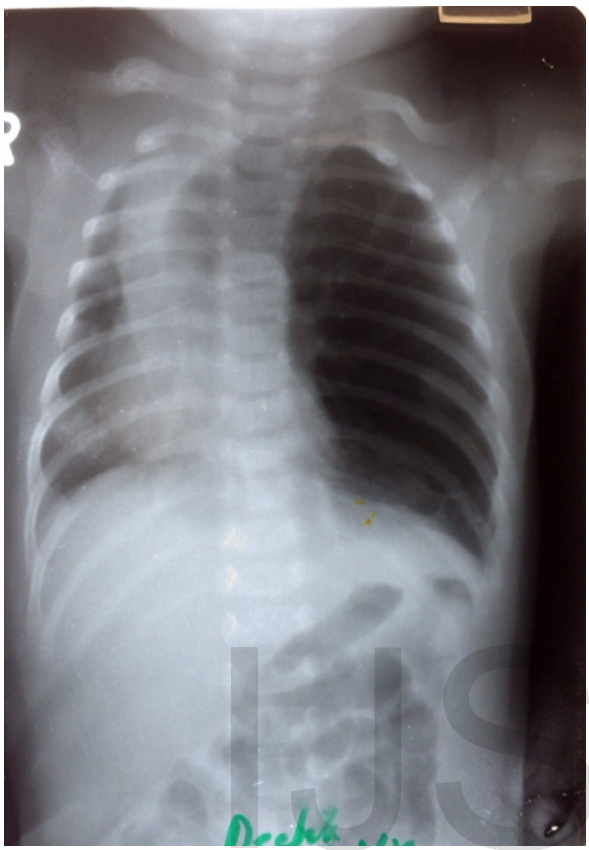

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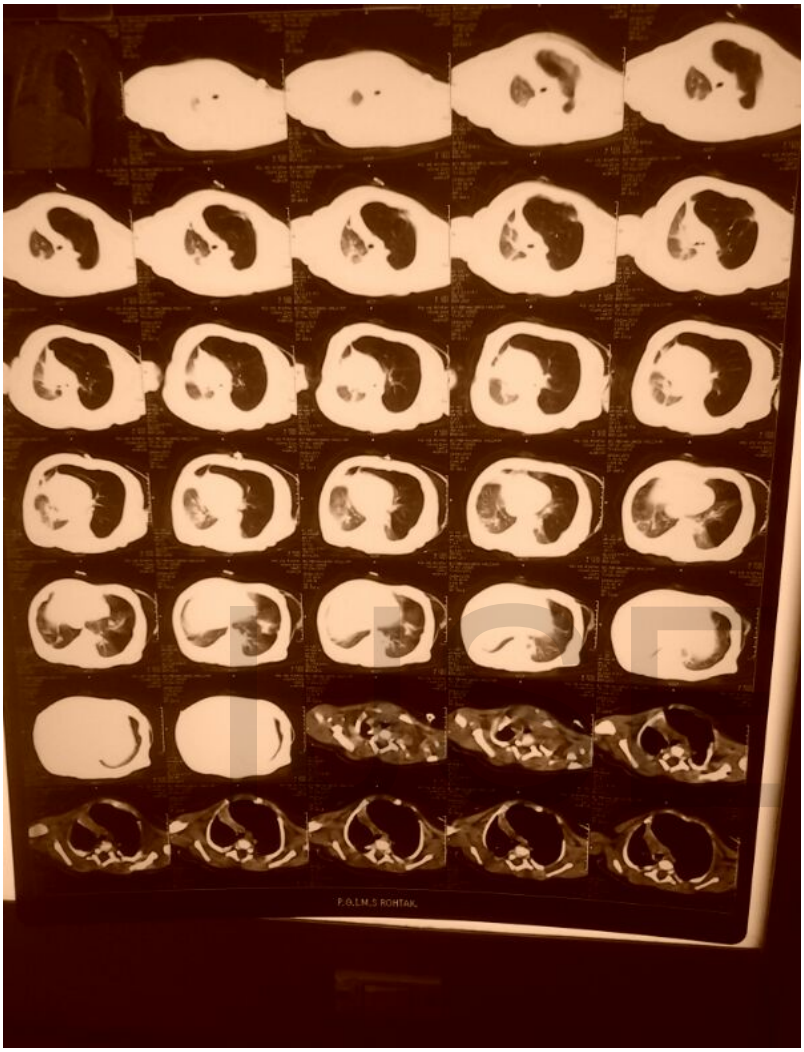
Figure 2. CT scan of the thorax revealed a hyperexpansion of left upper lobe with compression of the left lower lobe and the right upper and lower lobes with shifting of trachea and mediastinum towards right side



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