International Journal of Contemporary Pediatrics
Aditya K et al. Int J Contemp Pediatr. 2020 Apr;7(4):945-947
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: http://dx.doi.org/10.18203/2349-3291.ijcp20201004

Case Report

A case report of congenital lobar emphysema

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Received: 14 February 2020
Accepted: 20 February 2020

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ABSTRACT

Congenital lobar emphysema (CLE), also known as congenital alveolar overdistension, is a developmental anomaly of the lower respiratory tract that is characterized by hyperinflation of one or more of the pulmonary lobes. CLE is a rare congenital malformation with a prevalence of 1 in 20,000 to 1 in 30,000. We are reporting a 4-month-old boy presented with complaints of cough and cold for 3 days with history of similar complaints in the past at age of 1month and 2 months. Investigations revealed hyperlucency of left upper zone with tracheal shift and mild shift of the heart to the right. Elective Left Upper Lung Lobectomy was done and Appropriate supportive therapy given and child recovered well.

Keywords: Congenital lobar emphysema, Hyperlucency, Pulmonary lobes, Respiratory distress

INTRODUCTION

Congenital lobar emphysema (CLE), also known as congenital alveolar overdistension, is a developmental anomaly of the lower respiratory tract that is characterized by hyperinflation of one or more of the pulmonary lobes.[1,2] Other terms for CLE include congenital lobar over inflation, polyalveolar syndrome and infantile lobar emphysema.[3] CLE is a rare congenital malformation with a prevalence of 1 in 20,000 to 1 in 30,000.[4]

Progressive lobar hyperinflation is likely the final common pathway that results from a variety of disruptions in bronchopulmonary development. The most frequently identified cause of CLE is obstruction of the developing airway, which occurs in 25 percent of cases. Airway obstruction can be intrinsic, intraluminal or extrinsic, with the former being more common. This leads to the creation of a "ball-valve" mechanism, in which a greater volume of air enters the affected lobe during inspiration than leaves during expiration, producing air trapping. Additionally, bronchial atresia and pulmonary sequestration have been identified as a common finding in CLE.

CLE is characterized by overdistention of one or more lobes of the lung. This leads to compression of the remaining lung tissue and herniation of the affected lobe across the anterior mediastinum into the opposite chest, causing displacement of the mediastinum. The different pulmonary lobes are variably affected by CLE. The left upper lobe is affected most often (40 to 50 percent of cases). The distribution in right middle, right upper, and lower lobes is 25 to 35, 20, and 2 to 10 percent, respectively. CLE affecting multiple lobes is rare.[5-7]

Approximately 25 percent of cases present at birth, 50 percent by one month of age, and nearly all by six months of age. Progressive respiratory distress develops rapidly in some infants, while others have a more gradual, insidious onset and some may present as recurrent pneumonia and some may remain asymptomatic for years. Differential diagnosis includes 1. Pneumothorax or localised pulmonary interstitial emphysema 2. Space occupying chest lesion 3. Pulmonary growth

abnormalities 4. Swyer-James-Mcleod syndrome. Management can be done by resection of affected lobe or conservatively.

CASE REPORT

A 4-month male infant was brought to paediatric OPD with chief complaints of cough and cold for 3 days, rapid breathing with chest indrawing for 2 days, with past history of similar complaints at age of 1 month and 2 months. Antenatal and birth history was uneventful. Other history was not significant.

Physical examination

General examination was normal. There was no malnutrition. Tachypnoea was present, other vitals were stable. Respiratory examination showed mild tachypnoea, symmetrical chest expansion, Apex beat was felt at left 4th intercoastal space at mid clavicular line, bilateral subcoastal retractions, normal vesicular breath sounds with decreased breath sounds heard in upper zone of left lung with no adventitious sounds. Other systemic examination was normal.

Investigations

All the routine blood investigations were normal. Radiographic images of chest showed hyperlucency of left upper Zone with herniation of left upper lobe and tracheal shift and mild shift of the mediastinum to the right. CT scan showed Emphysema of left upper lobe of lung with compression of left lower lobe and also right upper lobe.

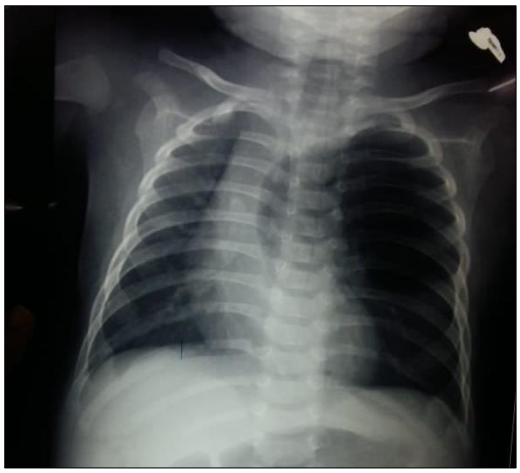

Figure 1: Hyperlucency of left upper zone with herniation of left upper lobe and tracheal shift and mild shift of the mediastinum to the right.

Management

With left postero-lateral thoracotomy approach, left upper lobectomy was done. Surgery went uneventful. Appropriate post-operative care was given. Infant recovered well with good left lung expansion.

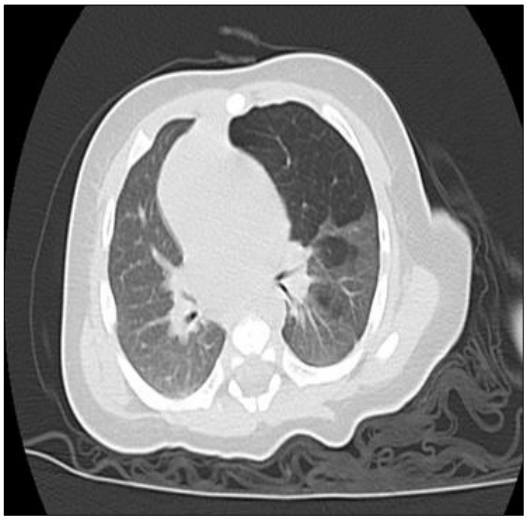

Figure 2: Emphysema of left upper lobe of lung with compression of left lower lobe and also right upper lobe.

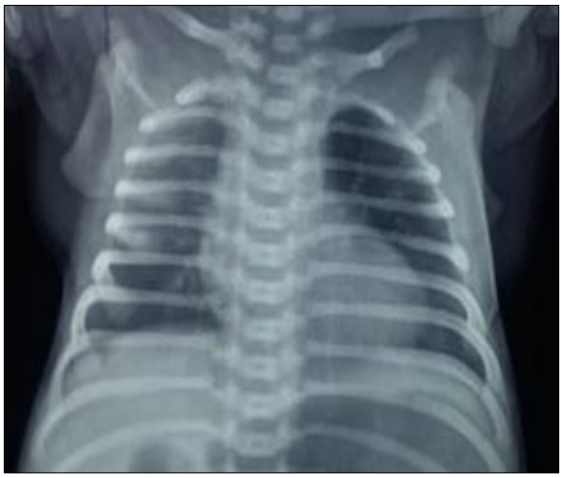

Figure 3: Post-operative radiographic imaging of chest showing good expansion of the left lung after lobectomy.

DISCUSSION

The patient had all the features suggestive of CLE. The baby presented in the 4th month of infancy which is very common. With adequate expertise and investigative facilities a baby with respiratory distress like our can be diagnosed easily. Antenatal diagnosis can be done with ultrasonography which was missing in our case.[8] Controversy exists regarding surgical and conservative management of this malformation and there is no contentious opinion. In our case surgical management was undertaken and baby recovered well post operatively with good lung expansion and follow up showed normal growth with no morbidities.

CONCLUSION

The high index of suspicion is required to diagnose congenital lobar emphysema and it should always be considered as a differential diagnosis in neonates or infants presenting with respiratory distress. As both surgical and conservative management are possible, protocol should be individualised according to the clinical presentation and status of the child along with status of the family.

ACKNOWLEDGEMENTS

Authors would like to thank the Department of Radiology, Kamineni Medical College for radiological investigations.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not Required

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Cite this article as: Aditya K, Parvathi K, Prabhakar V. A case report of congenital lobar emphysema. Int J Contemp Pediatr 2020;7:945-7.