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Congenital Lobar Overinflation (CLO) presenting as Severe Pneumonia

Authors
Dr Sunil Kumar Agarwalla[1], Dr Arpita Jalan[2]
[1]Associate Professor, [2]Junior Resident
Department of Paediatrics, M.K.C.G. Medical College & Hospital, Berhampur, Odisha

Abstract
Congenital lobar inflation (CLO) is a rare congenital pulmonary condition with prevalence of 4.5/1,00,000. It accounts for 10% of all congenital lung malformations. This commonly presents as respiratory distress mostly in neonatal period. Congenital alveolar over distension, congenital large hyperlucent lobe, and congenital lobar emphysema are other terms previously used.(1)
Here we present the case of a 4 month old male baby who presented to the Pediatric Emergency Department with severe respiratory distress. Clinically a diagnosis of Pneumonia with Pneumothorax was made but after seeing the X ray which was suggestive of a hyper Lucent, well marginated and delineated cystic shadow occupying left lower lobe, a clinical entity of Pulmonary cyst was made. This was finally diagnosed as a case of Congenital Lobar Inflation of left lower lobe. We are reporting this case because of its rarity where strong clinical suspicion and imaging tools helped in diagnosis and timely intervention.
Keywords: *congenital lobar over inflation, congenital lobar emphysema, respiratory distress, pulmonary cyst.*

INTRODUCTION

CLO is defined as the hyperinflation of one or more pulmonary lobes due to the partial obstruction of the bronchus.(2) It was first described by Nelson in 1932 and later by Robertson and James in 1951.(3,4)

It is more common in males, and the male to female ratio is 3:1.According to literature left upper lobe involvement is most common (43%), followed by right middle lobe (32%) and right upper lobe (21%) . Lower lobe involvement (2%) is the rarest form.(5,6)

In 50% of cases, a cause can be known.(8) Familial occurrence is known.(8)

Congenital deficiency of bronchial cartilage, bronchial stenosis, bronchomalacia, bronchiectasis, and abnormal bronchi are congenital causes of bronchial obstruction leading to CLO.(8) Bronchogenic cysts, mediastinal tumors, Vascular abnormalities such as pulmonary arterial sling, compression of bronchus by abnormal vessels, collaterals or by dilated pulmonary artery, foreign body aspirations ,redundant mucosal flaps plaques are other causes of CLO.

CLO is accompanied by cardiac anomalies in 12 - 14% of the patients as the development of heart and cartilage structure of bronchial system occurs at same time. Therefore cardiac diseases should be ruled out before surgery.

Prenatal USG can help in diagnosis of congenital lung disease where CLO can be seen as a cyst or

with increased echogenicity. Prenatal magnetic resonance imaging (MRI) finding scan also be helpful.

One-third of cases are symptomatic at birth while the other half mostly develop symptoms in the first 6 months of life.[5,6] It mostly present as respiratory distress in neonate or during infancy but can also remain asymptomatic for as long as 5-6 yr in 5% of patients.[8] Clinical signs vary from mild tachypnea and wheeze to signs of severe respiratory distress.[8]

Chest radiographs reveals a radiolucent lobe with a mediastinal shift.[8] MRI can demonatrate vascular lesions causing extraluminal compression. The gold standard diagnosis is HRCT scan.

The differential diagnosis includes pneumonia with or without an effusion, pneumothorax, pulmonary sequestration and congenital cystic adenomatoid malformation.[8]

Conservative management is done in mild cases and surgical intervention in severe respiratory distress cases.Positive pressure ventilation in a patient with CLO can cause hyperinflation so low pressure high frequency or pressure regulated volume controlled ventilation should be done.

OUR CASE DETAILS

Here we present a 4month old male child, product of nonconsanguineous marriage who presented to the Pediatric Emergency with chief complaints of fever, cough, breathlessness and difficulty in feeding for 4 days.

On physical examination the childhad tachycardia, tachypnea, low SpO2 (80%) on room airwith normal blood pressure. He had signs of severe respiratory distress in the form of nasal flaring, intercostal and subcostal retractions, chest in drawing, wheezing, cyanosis and grunting.On percussion hyperresonance was found in left lower lobe.

On auscultation decrease air entry was present in left posterior lung field with bilateral wheeze more on left side.Other system examination revealed no abnormality. The child was diagnosed as a case of severe pneumonia complicated with left pneumothorax and managed with intravenous fluids, antibiotics & supportive oxygen therapy.Emergency pleural tap was done in 2nd left Intercostal space and a good amount of air was drained. Inspite of air drain, child did not improve, respiratory distress increased and SpO2 kept falling. Childwas shifted to pediatric ICU,intubated and ventilated by synchronized intermittent mechanical ventilator (SIMV). The child was kept on mechanical ventilator support (figure 1a) for one week and managed with higher antibiotics and steroids. Gradually the child improved and was extubated. Chest radiographs (figure 1b & c) and HRCT thorax (figure 1d) was done which was suggestive of congenital lobar hyperinflation in left lower lobe with mediastinal shifting to the right.

Pneumothorax, event ration of diaphragm, encysted pneumothorax, lung hypoplasia and opposite lung hyperinflation due to atelectasis ruled out in HRCT Thorax. Child improved and referred to pediatric surgery center for lobectomy (figure 1f). He was discharged in a stable condition.

The learning lesson is how a surgical pulmonary condition like CLO which presented as severe pneumonia with pneumothorax where clinical suspicion and imaging tools guided towards proper diagnosis and ultimately surgery saved the child's life.

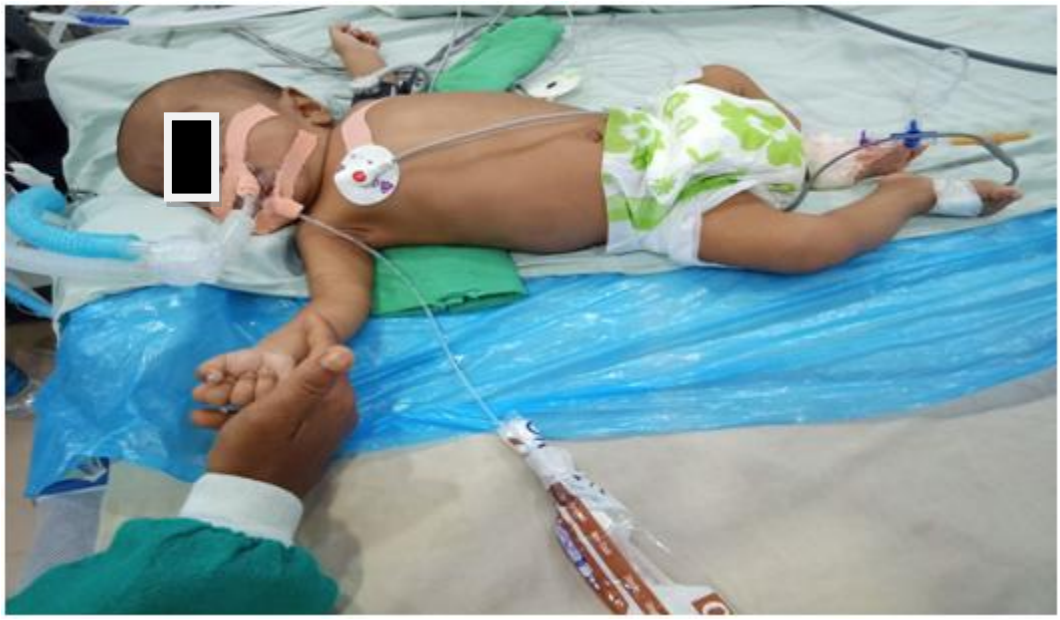

Figure 1a: Childon mechanical ventilator support

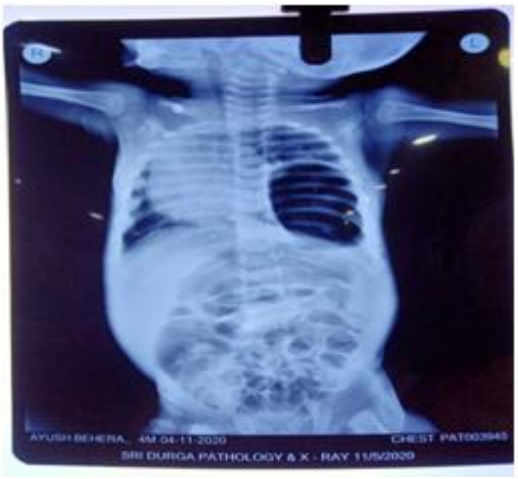

Figure 1b: Overinflation and hyperlucency of lung can be seenin leftlower lobe with mediastinal shifting to right side

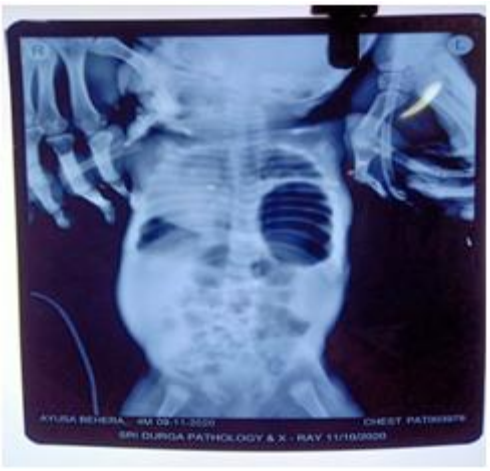

Figure 1c: Repeat X-ray showing CLO in left lower lobe

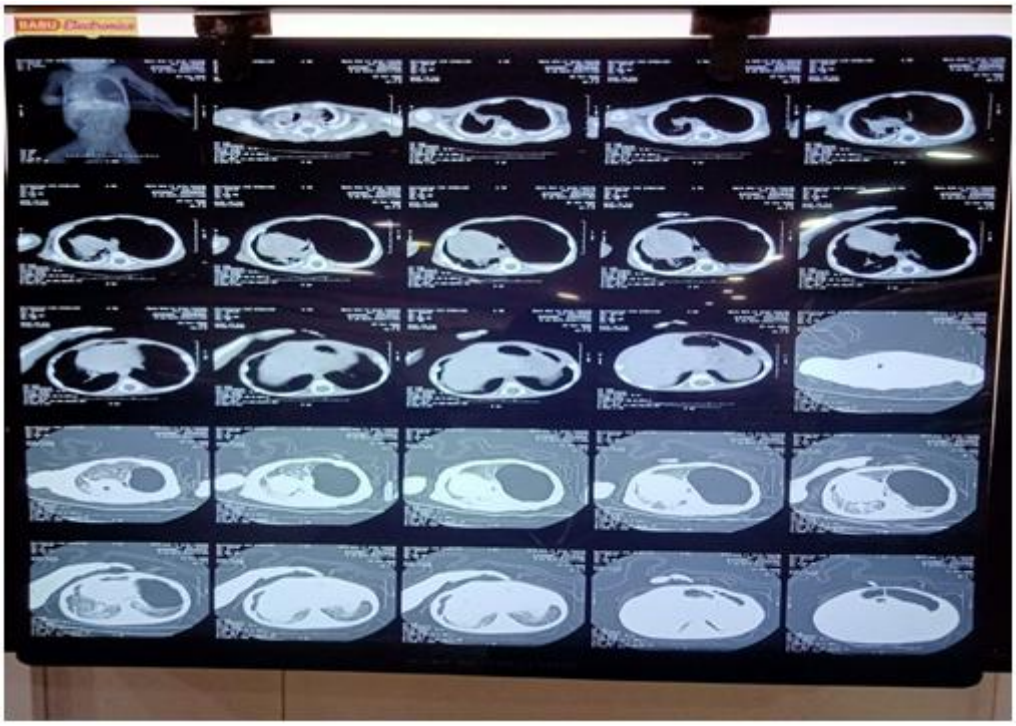

Figure 1d: HRCT shows L lower lobe CLO with mediastinal shift to R side

Figure 1f: Post surgery at the time of discharge

DISCUSSION

Congenital lobar overinflation (CLO) is a rare developmental lung malformation which commonly affects the upper and middle lobes with left upper lobe being the commonest .The affected lobe becomes nonfunctional due to over inflation. There is ventilation profusion mismatch and compression atelectasis of the ipsilateral normal lung in due course of time.[7] Mediastinum is shifted to the contralateral side, with impaired function.[8] All this leads to hypoxia and progressive respiratory failure.

This case of CLO was not picked up on Day1. Initially it was diagnosed as a case of severePneumonia and pneumothorax.Only after chest X ray and HRCT thorax, a diagnosis of CLO was made. This case israre as the presentation was in the left lower lobewhich is the least site for CLO.

Conservative treatment of CLO is done in mild and moderate disease, and lobectomy in severe disease cases. Kunisaki et al described 51 patients with diagnosis of CLE, who were treatment with lobectomy.In our case selective lower lobectomy has given the best outcome.Selective intubation of the unaffected lung is another option for treatment although difficult.[8] Patients with CLO can develop pneumothorax if the hyperinflated lobe ruptures.

CONCLUSION

CLO is one of the rarest causes of respiratory distress in newborn. Antenatal ultrasonography helps in picking up these cases early. Lung computed tomography (CT) is the gold standard in the diagnosis of CLO and lobectomy is the definitive management.

So any child who comes with respiratory distres, chest Xray is mandatory.This will give us a clue towards many congenital surgical conditions of the lung like CLO which needs to be confirmed further by HRCT.

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