ORIGINAL RESEARCH PAPER

Cardiothoracis

CONGENITAL LOBAR EMPHYSEMA (CLE)

KEY WORDS: Congenital lobar emphysema, Thoracotomy, Lobectomy.

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ABSTRACT

Aims: Diagnostic modalities and role of surgery for Congenital Lobar Emphysema (CLE) were evaluated in this study. Congenital Lobar Emphysema is a rare and life threatening clinical condition presenting mostly in neonatal period and early childhood with respiratory distress. Due to rarity as well as ignorance, this clinical entity may remain unrecognized and present later in life with recurrent respiratory distress. Chest radiography is the most valuable diagnostic tool. CT Chest confirms the diagnosis of CLE and provides all other related information. Lobectomy is the treatment of choice .

Materials and methods: We studied a series of 12 cases of congenital lobar emphysema (CLE) in infants and children who had surgical resection of their emphysematous lobes.

Results: Lobectomy was done in all the cases of CLE. 2 cases had PDA and 1 case had Bronchogenic cyst, associated with CLE .Ligation of PDA and resection of the Bronchogenic cyst were done in the same sitting along with lobectomy for these 3 cases. There was no mortality in our series and morbidity was insignificant. 7 patients were available for follow up for 3 years. All of them were asymptomatic with bilateral expanded lungs.

Conclusions: Congenital Lobar Emphysema (CLE) is a serious clinical condition of neonates and infants. Resection of the emphysematous lobe is the treatment of choice and can be safely performed even in very sick patients.

INTRODUCTION :

Congenital lobar emphysema (CLE) is a rare clinical condition presenting mostly within first six months of life with respiratory distress of varying severity due to progressive hyperinflation of the diseased lobe of a lung.

The hyper inflated lobe causes compression atelectasis of the ipsilateral adjacent lobes and herniates through the anterior mediastinum into the other chest.

As a result, the healthy contra lateral lung gradually gets more and more compressed. Further hyperinflation of the diseased lobe leads to gross mediastinal shift and severe compression atelectasis of the opposite healthy lung in addition to the compression atelectasis of the same sided lung. Thereafter, impairment of venous return, hypoxia,hypotension and further hemodynamic deterioration follows.

Congenital lobar emphysema (CLE) was first described in 1932, by Nelson[1],and was so designated in 1951, by Robertson and James. It is a rare disease, its incidence being 1 per 20-30 thousand births[2].

Congenital alveolar over distension, congenital large hyperlucent lobe, and Congenital lobar over inflation are other terms synonymous with CLE.

The commonest clinical presentation of CLE is neonatal acute respiratory distress.

Progressively worsening respiratory distress usually starts in the first few days of life in some cases but before the age of 6 months in the majority.

However, the onset of signs and symptoms of respiratory impairment can occur as late as the age of 5 years[3].

Etiology of CLE is still not clear and seems to be Idiopathic in 50% of cases[4].

Absence, hypoplasia or dysplasia of bronchial cartilage may be responsible for CLE in another 25% of cases. In the remaining cases several causes have been postulated as the etiology of CLE. Pulmonary Parenchymal diseases like Polyalveolar lobe or Pulmonary alveolar glycogenosis, Bronchial stenosis, Bronchomalacia, Meconium aspiration, Retained bronchial foreign body, Bronchial polyp,External bronchial obstruction by Pulmonary artery sling anomaly, Bronchogenic cyst, Lymph adenopathy or Mediastinal mass are few of them .

Histopathological examination showed that the alveolar number in the affected lobes was three to five times higher than in the other normal parts. It was not known why the air was trapped in this polyalveolar lobe[5].

CLE has been seen in twin babies, in a mother and her daughter, and in a father and his son. The disease is more common in white people than in black people. Although CLE is sporadic, data suggests that hereditary transition may be responsible for the disease[6-7].

Infants with CLE present a diagnostic challenge.

The most common clinical presentation is neonatal acute respiratory distress.

Diagnosis of CLE is usually made by clinical examination and chest X-ray. Ipsilateral hyperlucent lung field as well as mediastinal shift to the other side with compression atelectasis of lungs are usual radiological observations (Fig 1).

Role of CT scan of chest is invaluable for confirmation of diagnosis of CLE as well as for detailed information about both the lungs including extent of atelectasis, mediastinal shift, other associated congenital anomalies etc.

Bronchoscopy has a limited role in the diagnosis of CLE and indicated for excluding acquired bronchial obstruction in children whose symptoms may appear later in life.

Routine Echocardiography is very helpful to detect associated congenital cardiac anomalies.

Surgery is the treatment of choice for CLE and has been recommended since 1945, when Gross and Lewis reported the first case of CLE successfully treated by lobectomy[8-9].

The prognosis is good even when more than one lobe is resected and probably reflects the potential for compensatory alveolar growth in children.

MATERIALS AND METHODS:

- In this retrospective study, records of all the patients admitted for surgery with Congenital Lobar Emphysema (CLE) during the period between 2006 and 2017, were reviewed.
- Detailed history specially about respiratory distress as well as other clinical parameters were studied.
- Chest X ray and CT scan of thorax were also reviewed.
- Any other relevant investigations were taken into account.
- Diagnosis was confirmed in all the cases with clinical data, Chest X ray and CT Chest images.
- Intercostal chest drainage (ICD) was done in 2 cases based on the mistaken diagnosis of tension pneumothorax.

All the patients diagnosed as Congenital Lobar Emphysema (CLE) were planned for lobectomy.

Avoiding over-distention of the emphysematous lobe before thoracotomy remains the major anesthetic concern in such cases. Anesthesia was induced with 2 to 4 % halothane in 100% oxygen.

Endotracheal intubation was done carefully in spontaneously breathing patients.

Surgical team was ready and had to do emergency thoracotomy due to haemodynamic instability in 3 patients of the series.

Muscle sparing lateral Thoracotomy was done in all the cases. The hyper inflated emphysematous lobe of the lung herniated out of the chest instantaneously (Fig 3). Unstable haemodynamic parameters settled immediately with herniation of the emphysematous lobe after thoracotomy.

The diseased hyperinflated lobe of the lung was identified and lobectomy was done. Resection of one lobe was done in 10 cases while bilobectomy was required in 2 cases of our series.

Compressed and atelectatic lobes of the lung re expanded completely with the efforts of the anaesthesiologist. Chest was closed with ICD.

Post-operativenebulisation,spirometry as well as chest physiotherapy were essential exercises and were practiced meticulously in all the cases during post-operative period.

The patients were followed up every 3 months in OPD. All the patients were asymptomatic with expanded lungs in follow up after 3 years (Fig 4).

RESULTS:

Out of 12 patients admitted with CLE, 8 were males and 4 were females.

7 patients of our series were below the age of 6 months and the remaining 5 were below the age of 3 years.

All the patients of our series had history of recurrent respiratory distress.

6 of them presented with acute respiratory distress. Chest X ray was done in all the cases and was suggestive of CLEin 10 of them.

In the other 2, mistaken diagnosis of tension pneumothorax was made and ICD was introduced. Both the patients had marginal and non-sustained improvement. Repeat X ray and CT Chest confirmed the diagnosis of CLE.

CT scan of chest was done in 6 patients including 2 of, mistaken diagnosis of tension pneumothorax having ICD already introduced. Images confirmed the diagnosis of CLE in all the cases.

Hyperinflation of the affected lobe and its herniation into contralateral thoracic cavity through the anterior mediastinum, mediastinal shift, as well as collapse of the healthy lobes due to compression were the CT findings (Fig 2).

Associated Bronchogenic cyst in 1 case was also diagnosed in CTscan of chest.

Routine Echo screening detected PDA in 2 children.

Rigid bronchoscopy was performed in 4(33.33%) cases only just before thoracotomy to exclude suspected endo bronchial foreign bodies.

Only haemodynamically stable patients were selected for bronchoscopy.

Surgery was done in all the cases.
Treatment of choice was 'Lobectomy' of the hyper inflated lobe. Single lobectomy was done in 10 cases and bilobectomy was done in 2 cases of our series.

In the group of single lobectomy, 7 had Left upper lobectomy, 1 had right middle lobectomy and 2 had right lower lobectomy.

Right upper lobectomyalong with right middle lobectomy was done in 2 cases of bilobectomy.

Retained endo bronchial foreign bodies were not found in any case.

Associated Bronchogenic cyst in 1 case and PDA in 2 cases were operated in the same sitting along with lobectomy.

Intercostal chest drainage (ICD) was done in 2 cases based on the mistaken diagnosis of tension pneumothorax.

- Both of these 2 patients had single lobectomy .
- Complete re expansion of the atelectatic lobes of the lungs was confirmed on operation table.
- There was no mortality in this series.
- The patients were followed up in OPD every 3 months.

4 patients were lost from follow up. Remaining 8 patients were asymptomatic with well expanded lungs in follow up after 3years.

DISCUSSION:

Congenital lobar emphysema (CLE) is a rare clinical condition

usually presenting in the neonatal life or early infancywith acute respiratory distress.

Wheezing, chronic cough, and recurrent respiratory tract infections can be seen in infants suffering from CLE[5].

All the patients of our series had history of recurrent respiratory distress and 6 (50%) of them presented with acute respiratory distress.

It has been reported that the disease is more common in male children, usually unilateral, affecting more often the left upper lobe (43%) followed by right middle lobe (32%), although bilateral involvement is not unknown[10,11].

In our series,we had 66.66% male children.
7 (58.33%) of our patients were below the age of 6 months and the remaining 5 (41.67%) were below the age of 3 years.

We observed that,CLE affected Left upper lobe in 7 (58.33%), isolated right middle lobe in 1 (8.33%), isolated right lower lobe in 2 (16.67%)and right upper with right middle lobe together in2 (16.67%) cases.

Out of 9 patients having upper lobe lesions, 6 (12%) had severe symptoms.

In the group of single lobectomy, 7 had Left upper lobectomy, 1had right middle lobectomy and 2 had right lower lobectomy.

Right upper lobectomy along with right middle lobectomy was done in 2 cases of bi-lobectomy.

There was no case of bilateral involvement in our series.
Thakral CL et al in a study of 21 cases of CLE observed that, 13 had left-upper-lobe, 7 had right-middle-lobe, and 1had right-upper-lobe involvement. Of the 14 upper-lobe cases, 10 had severe symptoms whereas only 2 of the 7 middle-lobe cases had severe symptoms[12].

High level of suspicion remains the key for correct diagnosis. Chest radiography is helpful, but not definitive[13,14].

Some patients can be mistakenly diagnosed with pneumothorax and pneumonia instead of CLE in later life[15].

ICD insertion does not help to improve the condition and increases the respiratory distress further[16].

Misinterpretation of Chest X ray of CLE as "Tension Pneumothorax" was done in 2 (16.67%) of our patients and they were treated with Intercostal Chest Drains (ICD). Worsening of respiratory distress after ICD and reevaluation with CT Chest confirmed the diagnosis of CLE. Both the patients recovered uneventfully after lobectomy.

CT scan of Chest is confirmatory in the majority.
Bronchoscopy is considered as a high risk diagnostic procedure for CLE and is reserved for a group of mild or moderately symptomatic children in whom retained endo-bronchial foreign body needs to be excluded.

In the opinion of Karnak et al, the objective of Bronchoscopy was to exclude obstruction by a foreign body or thick secretion plugs and avoid inappropriate surgery, in accordance with their algorithm[8].

Echocardiography has been accepted as a routine diagnostic tool to exclude associated congenital cardiac anomalies.

Congenital Cardiac Defects may be associated with CLE infrequently.

Patent DuctusArteriosus (PDA), Atrial septal defect (ASD), Ventricular septal defect (VSD), Tetralogy of Fallot (TOF) etc are usually associated with CLE.

In a study of "Surgical management of infants with congenital lobar emphysema and concomitant congenital heart disease"Dogan R et al observed that Concomitant congenital heart disease (CHD) and CLE was not uncommon. In the literature a 12% to 20% concomitance rate was given[17].

We had associated PDA in 2 (16.67%) cases and associated Bronchogenic in 1 (8.33%) case of our series. Ligation of PDA and resection of the Bronchogenic cyst were done along with the surgery of CLE in the same sitting in all the 3 cases. All of them had uneventful recovery.

CLE may also be associated with other congenital anomalies rarely .

Lobectomy was done in all the cases of CLE of our series with no mortality.

We had surgical site infection (SSI) in 1 case. In another patients having associated PDA with CLE, drainage of chylous fluid through the ICD continued for 2 weeks . SSI healed in a week with dressing and the chylous drainage stopped with conservative management only .

literature also shows that the surgical treatment of CLE is safe, the morbidity and mortality from the procedure being low in most studies[18,19].

Images :

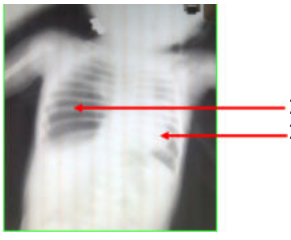

Fig 1:Chest X Ray PA view (Pre-operative)

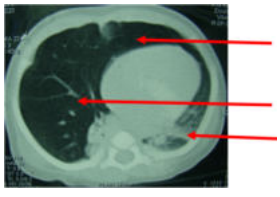

Fig 2:CT Scan of Chest (lung window)

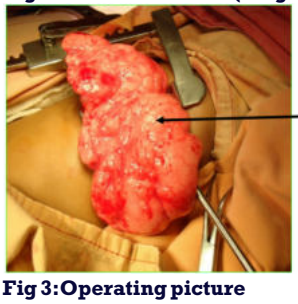

Fig 3:Operating picture

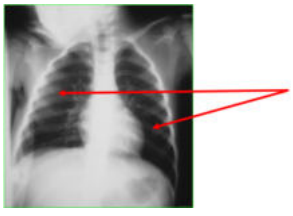

Fig: 4 - Chest X Ray PA view (Post-operative)

CONCLUSIONS :

Congenital lobar emphysema (CLE) is a rare clinical entity of neonates and infants. Etiology of this disease is uncertain in the majority.

Diagnosis of this life threatening disease remains a major challenge .

High degree of suspicion in neonates and infants with respiratory distress, is the most important tool for the diagnosis of CLE .

Chest X ray is an invaluable imaging modality for confirmation of diagnosis as well as for planning therapy .

Lobectomy is the treatment of choice and has been established as an effective as well as safe therapeutic option for CLE .

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