Original Article

Congenital lobar emphysema in children: Institutional experience and errors to be avoided in the management

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ABSTRACT

Background: Congenital Lobar emphysema (CLE) is one of the rarest cystic lesions in the thoracic cavity among children. It is often misdiagnosed and managed incorrectly. We are reporting our institutional experience and recommendations regarding the prevention of common errors in the management of this condition.

Methods: This study was conducted at the Paediatric Surgery Department, The Children’s Hospital & the University of Child Health Sciences Lahore from January 2015 to December 2018. All the patients who underwent thoracotomy for congenital lobar emphysema at our center were included in the study and their charts were reviewed.

Results: A total of 19 patients had been treated at our center in this duration. The mean age of patients was 2.85 ± 2.11 months. Thirteen of them (68%) were male and the most common presenting complaint (94.7%) was respiratory distress. Chest X-ray was done in all of the patients while a CT scan was needed in 11 patients (57.9%). The most involved lobe was left upper (n=15). Lobectomy was done in all patients. Post-operative ventilation was required in 2 patients (10.5%) and pneumonia developed in 2 patients (10.5%). The most significant complication was wound infection (9/19, p=<0.01).

Conclusion: Our results about perioperative management are consistent, however, we received cases with misdiagnosis. We have proposed recommendations to address this issue.

Keywords: Congenital Lobar Emphysema, Child, Pneumonia, Intubation.

INTRODUCTION

Congenital Lobar Emphysema (CLE) is one of the rare anomalies of the lungs, having a prevalence of 1 in 20,000 to 1 in 30,000.[1] It was first reported by Nelson in 1932 and was named by Robertson and James in 1951.[2] Its antenatal prevalence is not known yet. CLE is defined as hyperinflation of lobes of the lungs which allows airflow on inspiration only. The most suggested theory regarding its etiology is a congenital deficiency of bronchial cartilages and compression by aberrant vessels.[3] However, there has been an alternate theory of hyperalveolosis, according to which the increased number of alveoli leads to hyperinflation of the affected lobe.[4] The affected lobe is hyperinflated, which ultimately becomes non-functional because of excessive air trapping. Due to this, the mediastinum is shifted to the contralateral side, which leads to bilateral functional compromise and eventually leading to respiratory distress, cyanosis, and repeated pneumonia, which may be the typical feature in this disease. Due to hyperinflation of the affected lobe, it gives a typical appearance on chest X-ray (CXR) including the presence of lung markings and ipsilateral atelectasis. The most common differential diagnosis of CLE is pneumothorax; however, certain features can be found which are distinct in CLE like...
the presence of bronchogram markings, and atelectasis of ipsilateral lower lobes.[5-7] These features may not be found in pneumothorax, which makes it possible to distinguish CLE on CXR. It is very common to see these patients presenting to us having mistaken chest intubation.[8-10] We will present here our institutional experience in managing these cases.

METHODS

This retrospective study was conducted at The Children’s Hospital & the University of Child Health Sciences, Lahore. After approval from the ethical review board of the hospital, we extracted the record of CLE patients, who underwent thoracotomy in our hospital in the last 4 years, from January 2015 to December 2018. The diagnosis of CLE was labeled as per the perioperative findings mentioned by the operating surgeon. We extracted all the information from the records including age, gender, weight, presenting complaint, investigations done including Computed Tomography (CT) scan, lobe and side involved, surgery done, and postoperative events.

RESULTS

A total of 19 patients underwent thoracotomy for CLE. All of them were included in this study. The mean age of patients was found as 2.85 ± 2.11 months (Range: 27 days – 9 months). Four patients were neonates and 15 were infants. Thirteen of them (68%) were male and their mean weight was 4.49 ± 2.61 kg. The most common presenting complaint (94.7%) was respiratory distress and the mean duration of symptoms was 42.94 ± 40.38 days (Range: 27-180 days). CXR was done in all patients while a CT scan was needed in 11 patients (57.9%).

CLE was misdiagnosed in seven patients as pneumothorax and these patients presented to us having chest tubes (36%). All of these were referred to us from the periphery by primary care physicians. The antenatal diagnosis was not made in any patient.

In 18 cases (94.7%), it was left-sided while one patient had it right-sided. Similarly, in 17 patients (89.4%) it was involving the upper lobe while one patient had involved the middle and one had the lower lobe. Most commonly involved lobe was left upper (n=15) (78.94%), followed by left middle (n=1), left lower (n=1) and right upper lobe (n=1). Lobectomy was done in all patients. Most common postoperative complications were wound infection (n=9), pneumonia (n=3) and persistent non-expansion (n=2). Post-operative ventilation was required in 3 patients (16%) and postoperative hospital stay was 9.22 ± 5.21 days (Range: 0.2 – 27 days). Of our series, 14 patients survived and were discharged with a mean follow-up of 2.65 ± 0.84 years. All the postoperative outcomes are summarized in table 1. Three patients died because of hospital-acquired pulmonary infection while 2 patients left the hospital against medical advice.

| Complications | Yes | No |
|---------------|-----|----|
| Wound infection | 9 (47.3%) | 3 (15.7%) |
| Pneumonia | 3 (15.7%) | |
| Persistent non-expansion | 2 (10.52%) | |
| Postoperative Chest drain | 5.53 ± 4.46 days (Range: 0.2 – 23 days) | |
| Postoperative hospital stay | 9.22 ± 5.21 days (Range: 0.2 – 27 days) | |
| Total hospital stay | 21.68 ± 8.49 days (Range: 7 – 40 days) | |
| Post-operative Ventilator | | |
| Alive | 14 (73.6%) | |
| Dead | 3 (15.7%) | |
| LAMA | 2 (10.52%) | |

DISCUSSION

CLE is a rare disease, which is mostly diagnosed in neonatal life, however, 5% are reported after 6 months of age.[8] In our series, all patients were below 9 months of age and only 2 patients (10.5%) were above the age of 6 months with the mean age being 2.85 ± 2.11 months. It is more common in males than females (3:1). We had 13 male patients, with the male preponderance being 2.1:1.

The most common presenting complaint was respiratory distress in our patients. All patients had CXR and 11 patients needed CT scan in our series. Other investigations which may be helpful and may be needed at times may include Magnetic Resonance Imaging, bronchoscopy, bronchography, angiography, and ventilation-perfusion scan.[11]

Regarding prenatal diagnosis, in a recent study, it has been reported in 24.5% of patients with CLE as compared to 73.1% of patients with other lung malformations.[7] In our series, none of the patients was antenatally diagnosed. It is because of limited resources and that most patients in our country do not get proper antenatal care. In a recent study, the authors found that 53.7% of patients receive four or more visits to any healthcare worker during the antenatal period, and this narrates the condition of antenatal care in our setup.[1]

In previous studies, the left upper lobe is the most commonly involved lobe, followed by the right middle lobe and rare bilateral involvement. Left upper lobe involvement has been reported as 33% [2], 50% [3], and even 66% [12] with a varied range. Similarly, in our series, the
left upper lobe was most commonly involved (89%) but no bilateral case was observed. It may be due to our inclusion criteria as we only included the patients who had undergone surgery in a resource-limited country and those with severe symptoms may not have made it to our center.

The final treatment for CLE is lobectomy and it was first done by Gross and Lewis in 1946.[2] However, now lobectomy is reserved for those presenting at an early age or those having severe symptoms. In those with age, more than 6 months and mild to moderate symptoms, conservative management with closed observation is narrated.[13,14] We had lobectomy of all patients, as was indicated.

In our series, 3 patients (15.7%) had postoperative ventilator requirements and a similar number of patients had postoperative pneumonia. The wound infection was the most commonly found complication. All infections were superficial and recovered very quickly. The outcomes in our study showed that 16 patients (84.2%) survived. Fourteen patients are in long-term follow-up with us. Two patients got away from the department without any further treatment against the medical advice after surgery had been done.

The diagnosis of CLE is not easy and it can be easily missed and mostly misinterpreted as pneumothorax if the physician is not vigilant enough.[15,16] In a series of data over a 10 year period, authors found the misdiagnosis of CLE in 30% of cases.[17] However, in another large series, which included 20 patients over a period of 30 years, authors found misdiagnosis of CLE in only one patient (5%).[2] In our series, it was misdiagnosed in 7 patients (38.5%).

Based on our experience with these cases we present a summary of common mistakes in the management of CLE and our recommendations regarding how to avoid them:

1. Chest Tube Placement based on x-ray findings:

As we wrote above, 38.5% of patients were referred to us from outside with chest tubes already inserted with a misdiagnosis of pneumothorax. If the following points on assessing the chest x-ray are kept in mind, then the diagnosis becomes noticeably clear and chest tube insertion can be avoided:

A. In CLE, pulmonary vessels are visible. In other words, vascular markings on lung markings are visible. All these words mean the same thing, despite hyperlucency, clear opaque marks are seen within the hyperlucency. These opaque marks are from pulmonary vessels in the hyperinflated lung.

B. In pneumothorax, a pleural line is visible. This line is present between parietal and visceral pleura and contains air. No lung tissue is present; thus, no vascular markings are visible.

2. Positive pressure ventilation:

Conventional ventilation leads to air trapping via a one-way valve; thus, the condition will deteriorate. The advancements like high-frequency low-pressure ventilation and selective bronchial intubation will improve the patient’s condition. However, the mainstay of treatment in deteriorating cases remains urgent thoracotomy and resection of the affected lobe.

3. Computed Tomography of Chest:

Computed tomography will confirm the diagnosis including side and lobe involved and should always be considered in stable patients with suspicion of CLE.

The results in our case series are consistent with the international literature. Since this study we have raised standards of sterility to reduce its effect on postoperative wound infection. We received cases with chest tubes placed due to misdiagnosis. We strongly reiterate that pediatricians, intensivists, pediatric surgery residents, and emergency department physicians should be aware of CLE and follow the recommendations outlined here.

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