Original Article

Congenital lobar emphysema: Thoracotomy versus minimally invasive surgery

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Abstract:

BACKGROUND: Congenital lobar emphysema (CLE) is a rare developmental malformation of the lung but can be associated with high morbidity and mortality. The objective of this study is to review our experience with 45 patients with CLE highlighting clinical features, aspects of diagnosis, and management.

METHODS: The medical records of all patients diagnosed with CLE in our center were reviewed. Patients age at the time of diagnosis, sex, clinical presentation, associated anomalies, the lobes affected, treatment modality, and outcome were described. A comparison was made between those who had lobectomy via open thoracotomy and those treated thoracoscopically.

RESULTS: From January 2000 to December 2018, a total of 45 infants with CLE were presented to our institution. There were 30 male and 15 female, and the mean age at presentation was 3.35 months. Twenty-five patients presented with respiratory distress. Nine patients presented immediately after birth, and two of them had surgery within the 1st week of life. Twenty patients were presented with repeated chest infections. Left upper lobe was affected in 27 patients, right middle lobe in 13, and right upper lobe in 5 patients. Forty-four patients had a lobectomy, and one was managed conservatively. One patient had a postoperative bronchopleural fistula. Nine patients had a thoracoscopic lobectomy, and two of them were converted to open thoracotomy because of persistent air leak. The operative time and hospital stay were nonsignificantly longer in thoracoscopic lobectomy (P = 0.5 and 0.4, respectively). There was no operative mortality in both groups.

CONCLUSIONS: CLE is a rare malformation with variable presentation. Infants presenting with respiratory distress or recurrent chest infection should be evaluated for the possibility of CLE. Lobectomy is the treatment of choice, and rarely, the patients may be managed conservatively. Thoracoscopic lobectomy is a safe procedure with the possibility of air leak and conversion to open lobectomy.

Keywords:

Acute respiratory distress, congenital lobar emphysema, lobectomy, minimally invasive surgery, open thoracotomy, recurrent chest infection, thoracoscopy

Congenital lobar emphysema (CLE) is a rare developmental lung anomaly, characterized by hyperinflation of one lobe most commonly the left upper lobe. CLE has a wide range of presentations, and it poses a diagnostic and therapeutic dilemma.[1,2] Among the congenital pulmonary disease, CLE has a low detection rate prenatally.[3] It has a progressive course, and infants presenting with respiratory distress, or repeated chest infection should be investigated for the possibility of CLE. Despite reports of successful conservative management,[4] surgical lobectomy of the affected lobe is the treatment of choice, which is usually curative. In the past, open lobectomy was the treatment of choice for CLE, but as a result of the advancement of minimally invasive surgery, thoracoscopic

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lobectomy has gained broad acceptance among pediatric surgeons and increasingly becoming the procedure of choice.[3,5] However, the thoracoscopic lobectomy in infants is associated with major complications and reports about the incidence of complications after thoracoscopic surgery in neonates, and infants are scarce.[6,7]

The objectives of this study are to analyze our experience in the management of CLE patients and describe patients' presenting symptoms and diagnostic modality. In addition, we compared patients who were treated with open thoracotomy versus those treated thoracoscopically.

Methods

Study design

The study is a retrospective cohort study. From January 2000 to December 2018, a total of 45 infants with CLE were presented to our center. Patients data, including the age at presentation, sex, the presenting symptoms, radiological investigations, management, and outcome, were retrieved from the medical charts. The study was approved by the Ethical Committee of our institution, and patients' consents were obtained because of the retrospective nature of the study.

Management of congenital lobar emphysema

Infants presenting with respiratory distress or recurrent chest infection are investigated for the possibility of CLE. The initial investigation is chest radiography with both posteroanterior and lateral views [Figure 1]. In some patients, the overdistended lobe resembles intraparenchymal pulmonary cyst or encysted pneumothorax, and the lung markings are not evident. In these patients, chest computed tomography (CT) is indicated for diagnosis [Figure 2]. Chest tube insertion is contraindicated until CT-scan confirms the diagnosis. In patients with suspected extrabronchial compression as a cause of lobar emphysema, a CT scan is also indicated.

After confirming the diagnosis of CLE, surgery is scheduled based on the severity of the symptoms. Patients who had severe respiratory distress had lobectomy scheduled on urgent bases, and patients with milder symptoms are scheduled for elective surgery. If the overdistention of the lobe improves during follow-up, the patients are managed conservatively with frequent follow-up visits.

The treatment of choice is lobectomy, which can be either performed through thoracotomy or thoracoscopy. Patients were assigned to either approach based on the preference and experience of the surgical team.

Prenatal diagnosis of CLE was suspected in two patients (4.4%) who were delivered in our institution, and most of the patients were referred to our tertiary center with no clear history of prenatal diagnosis.

Surgical technique

All interventions were done under general anesthesia using a single-lumen endotracheal tube (ET). We did not use lung isolation techniques, e.g., catheter or double-lumen ET. Patients were positioned in the lateral position.

In patients who had open thoracotomy, we used muscle-sparing lateral thoracotomy. Classically, there was protrusion of the distended lobe outside the thoracic cavity once the chest was opened [Figure 3]. We ligated the branches of the pulmonary artery first then the vein and finally the bronchus. We sutured the bronchus with polyproline suture and stapler was used in older children.

In patients who had thoracoscopic lobectomy, CO_2 was insufflated using 4 mmHg pressure to a maximum of 6 mmHg to deflate the lung. Three ports were inserted, the camera port was placed in the 7th or 8th intercostal space in the midaxillary line, and two working ports were placed in the 6th intercostal space on the midclavicular line and in the fourth or fifth intercostal space on the posterior axillary line. Positions of the ports were adjusted for each patient; occasionally, an extraport was used for retraction. For lower lobectomy, we tried to be as low as possible. We inserted the side port first at the 7th intercostal space and put the camera port just above the diaphragm. The affected lobe was dissected using LigaSure, and the vessels were dissected then the bronchus, which was dissected distally and clipped leaving a stump to avoid damage to the small main bronchus [Figure 4]. During thoracoscopy, the O_2 saturation and FiO_2 were monitored. If the patient became desaturated, the pressure was relieved and if no improvement, we converted the intervention to open approach. The affected lobe was resected and removed through mini-thoracotomy. At the end of the procedure, the intrathoracic CO_2 was removed and a small chest tube was inserted. The surgical technique was standardized in all patients, and a single consultant surgeon has performed all the operations.

For infants, the insufflation does not exceed 5 mmHg and 3 mm instruments were used. We clipped the bronchus, and no staplers were used.

Statistical analysis

Descriptive statistics were used to present patients data, diagnosis, and management. Continuous variables were presented as mean and range and categorical variables as numbers and percentages. A comparison was made between patients who had open lobectomy

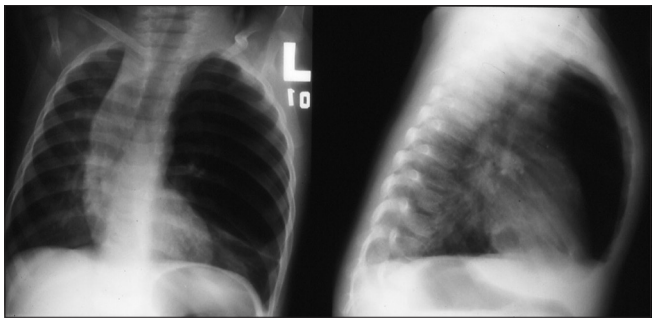

Figure 1: Anteroposterior and lateral chest X-ray showing congenital lobar emphysema of the left upper lobe with compression of the lower lobe and herniation to the other side

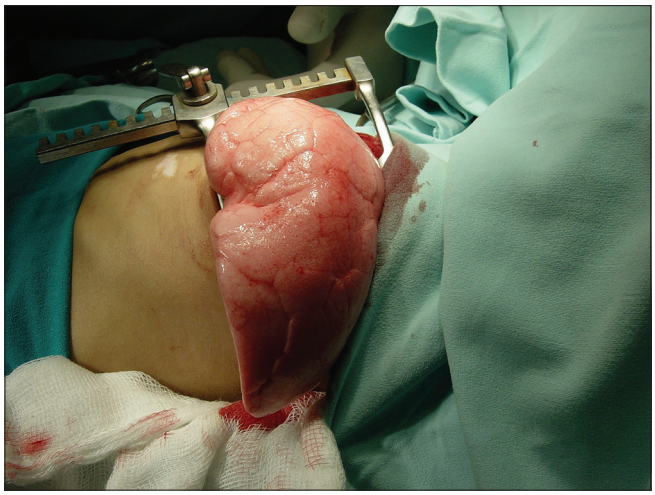

Figure 3: An operative photography showing the over distended left lobe herniating outside the chest cavity

versus thoracoscopic lobectomy. Numerical data were compared using *t*-test or Mann–Whitney test and qualitative data with Fisher exact test.

Results

Presentation

Forty-five infants with CLE were presented to our institution; 30 of them were male (66.67%). The mean age at presentation was 3.35 months (range: 1 day–8 months). Twenty-five patients (55.56%) presented with respiratory distress, nine of them (36%) presented immediately after birth, and two required surgery within the 1st week after delivery. Seven out of the nine patients were discharged and readmitted at the age between 2 and 5 months based on the severity of their respiratory symptoms. Twenty patients (44.4%) presented with repeated attacks of chest infection. One patient had pneumonia and mild respiratory distress and found to have CLE diagnosed when she was 3 months old. The patient was treated conservatively, and the emphysema regressed. One patient presented at 1 month with a chest infection and was diagnosed with CLE. This patient suffered recurrent

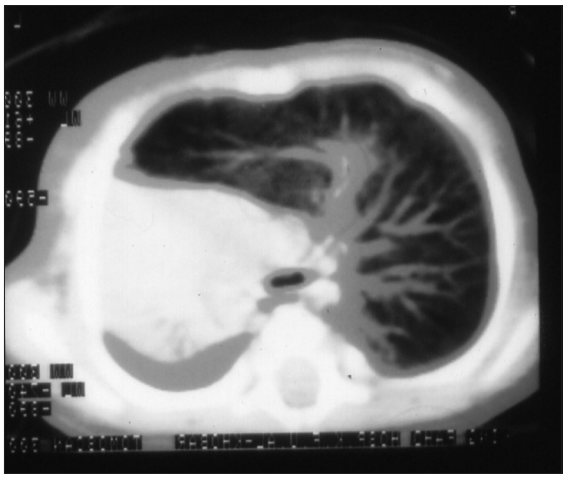

Figure 2: Chest computed tomography showing congenital lobar emphysema with herniation of the distended left upper lobe to the other side

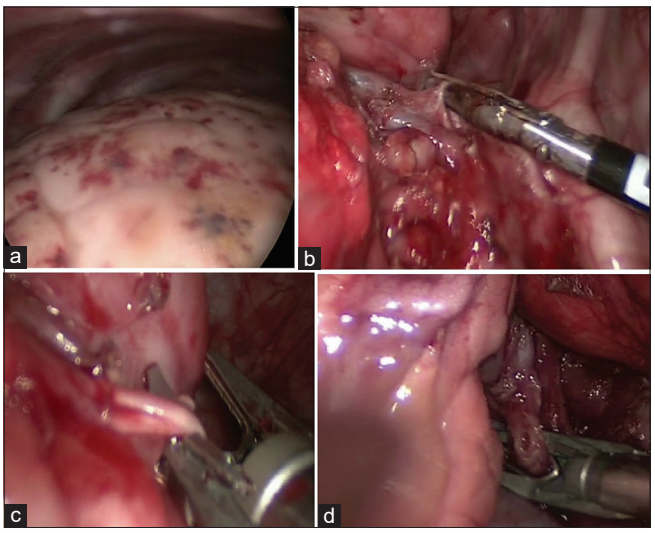

Figure 4: Thoracoscopic lobectomy for congenital lobar emphysema. (a) Inflation of the affected lobe with limited workspace. (b) Dissection and seal of the small branches using ligature (c) Clipping of the artery and (d) the bronchial stump

attacks of chest infection, and the parents refused surgery initially, then surgery was performed at the age of 6 months because of the high frequency of repeated chest infection.

Disease distribution and diagnosis

The left upper lobe was affected in 27 patients (60%), the right middle lobe in 13 patients (28.9%), and the right upper lobe in 5 patients (11.11%). The best initial test was chest radiograph, which was performed in all patients. Fifteen patients (33.3%) required CT-scan of the chest to confirm the diagnosis.

Management

Surgery was performed in 44 patients and 1 patient was managed conservatively. The surgical procedure was lobectomy of the affected lobe. Nine patients had

a thoracoscopic lobectomy, five had a right middle lobectomy, three had left upper lobectomy, and one had a right upper lobectomy. Two were converted to open thoracotomy (right middle lobectomy and left upper lobectomy). The reason for conversion was persistent air leak from the bronchial stump. The age of these two patients was 3 and 4 months old, and a 10-mm stapler was used to close the bronchial stump. The operative time and hospital stay were longer in patients who had thoracoscopy but did not reach a statistically significant level (P = 0.5 and 0.4, respectively).

Postoperatively, one patient developed a bronchopleural fistula and was treated conservatively then required operative closure. Histological examination was done for the all resected lobes, and the diagnosis of CLE was confirmed histologically. There was no operative mortality. The follow-up ranged from 18 months to 9 years, and all patients were asymptomatic with normal growth and development [Table 1].

Discussion

Congenital cystic diseases of the lung may be associated with high morbidity and mortality, especially in patients with delayed diagnosis.[1] Patients with CLE can present early in the neonatal period with respiratory distress, and in mild disease forms, it can present later with a recurrent chest infection and malignant transformation were rarely reported in CLE.[8,9] Surgical resection is essential even in asymptomatic and incidentally discovered cystic lesions. However, patients with CLE can be managed conservatively in stable and asymptomatic patients, and close follow-up is recommended.[1,8,9]

CLE commonly affects the left upper lobe, and in our series, 60% had left upper lobe affection and 29% had right middle lobe emphysema. Respiratory distress occurs because of overdistension of the lobe with compression of the healthy portions, and in some cases, compression of the contralateral lung. Therefore, early detection and management are vital to prevent mortality in those patients. Patients with CLE should be evaluated with echocardiography preoperatively because of the associated cardiac lesions in up to 10% of patients;[10,11] however, none of our patients had associated cardiac lesions.

Table 1: Comparison of open and thoracoscopic lobectomy

	Open lobectomy (*n*=36)	Thoracoscopic lobectomy (*n*=9)	*P*
Age at operation (months)	3.9 (1-7)	3.4 (1-6)	0.37
Sex (male:female)	24:12	6:3	>0.9
Operative time (min)	71 (51-112)	110 (80-145)	0.5
Complications, *n* (%)	1 (2.8)	2 (22)	0.097
Hospital stay (days)	10.4 (6-20)	12 (7-20)	0.4

Continuous variables are presented as mean, and range and categorical variables are *n* (%)

The pathophysiology of CLE is not fully understood, and it could occur as a result of a cartilaginous anomaly of the involved bronchus.[10,12,13] Another theory is the polyalveolar lobe, where there is an increase in the number of alveoli up to five-folds.[14] CLE may rarely occur because of extraluminal obstruction and compression of the bronchus either by a cyst or a blood vessel. The ultimate result of these factors is overinflation of the affected lobe through a ball-valve mechanism.

Most CLE patients present before 6 months of age, and it affects males more than females. This statement agrees with our finding, in which 44 patients presented before minimally months and 1 patient presented at the age of 8 months, and males were affected twice as the females. Patients in our series either presented early with respiratory distress or later with a recurrent chest infection. Chest radiograph was enough for the diagnosis in most of our patients. However, in patients with overdistended lobe, chest X-ray could not differentiate CLE from congenital cyst or pneumothorax, and a CT scan was mandatory. CT-scan can additionally localize any extrinsic lesion causing CLE.

The accepted treatment modality of CLE is resection of the affected lobe. Early surgery is recommended to prevent the complications of lobar overdistension. On the other hand, a conservative approach can be used in stable and asymptomatic patients, which can lead to improvement and avoid surgery.[12,15] One of our patients was managed with this approach, and CLE has resolved. CLE patients cannot tolerate positive pressure ventilation, which can lead to the rapid expansion of the affected lobe, mediastinal shift, and cardiac arrest.[16,17] Lobectomy is well tolerated in young age as the remaining lobes expand and fill the thoracic cavity, and currently, the thoracoscopic approach has gained popularity for lobectomy in children.[9,18]

We treated nine patients with CLE thoracoscopically. This study is a small series to conclude from, but thoracoscopic resection of CLE was shown to be safe, effective, and beneficial for these patients.[9,18] Despite the increased utilization of thoracoscopic surgery in pediatric congenital lung lesions, comparative studies with the open approach are lacking. This could be attributed to the rarity of the condition and small patients' number. In comparing the open thoracotomy group to the thoracoscopic group in our series, the operative time and hospital stay were longer in patients who were

treated thoracoscopically. The study presents our initial experience, and a longer operative time is expected initially. Polites and associates found no difference in postoperative length of stay or complications between thoracoscopic and open lobectomy.[19] Rothenberg *et al*., on the other hand, concluded that thoracoscopic lobectomy is safe for infants <10 kg and avoids the morbidity associated with thoracotomy and also better in term of less operative time and shorter hospital stay.[18] In addition to this, the superior cosmetic appearance. In two of our patients, thoracoscopic resection was converted to open thoracotomy because of a persistent leak from the bronchial stump. We think this resulted from the use of the 10-mm stapler, which is large for infants <4 months old. The availability of the 5-mm stapler is beneficial in these patients.

Study limitations

The major limitations of the study are the retrospective nature and nonrandom assignment either to open or thoracoscopic lobectomy. Several factors could have affected the outcomes, including the surgeons' experience. Another limitation is the small patients' number, which could be attributed to the rarity of the disease.

Conclusions

The presentation of CLE is variable, but the majority of patients present with acute respiratory distress or repeated attacks of chest infection. Pediatricians should be aware of the possibility of CLE, and a high index of suspicion is vital to avoid associated morbidity and mortality. Anteroposterior and lateral chest X-rays are essential to diagnose CLE, and CT scan may be required to confirming the diagnosis in some patients. Lobectomy is the treatment of choice, and rarely, the patients can be managed conservatively. Thoracoscopic lobectomy is a safe procedure with the possibility of air leak and conversion to an open approach.

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Conflicts of interest

There are no conflicts of interest.

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