Original Article

Lobectomy for Congenital Lobar Emphysema, Experience of Two Centers in Pakistan

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Objective: To assess outcome of lobectomy in congenital lobar emphysema in pediatrics group of patients.

Methods: This retrospective study was conducted in Department of Thoracic Surgery Nishtar Hospital Multan and Services Hospital Lahore from October 2003 to December 2015.All the patients of pediatric group who underwent lobectomy for congenital lobar emphysema were included in study. Demographic data, operative findings outcome of the procedure in terms of post operative complications and 30 days morbidity and mortality were recorded .All the patients presented with respiratory distress. Standard lobectomy was performed and bronchial stump was buttressed with surrounding lymph nodes, pericardial or pleural flap.

Results: Of these 41 study cases, 30 (73.2%) were boys while 11 (26.8%) were girls and male to female ratio was 2.72:1. Mean age of our study cases was 3.32 ± 1.59 months (with age range; 15 days to 7 months). Mean age of the boys was noted to be 3.38 ± 1.52 months while that of girls was 3.18 ± 1.84 months (p = 0.405). Of these 41 study cases, left upper lobectomy was performed in 27 (65.9%) patients, right middle lobectomy in 11 (26.8%) patients, left lower lobectomy in 2 (4.9 %) patients and right upper lobectomy in 1 (2.4%) patients. Out of 41 patients 8 (19.5%) were preoperatively on ventilator. Out of 8 ventilated patients one had left sided chest intubation which was malpositioned and was in the lung parenchyma. Morbidity was superficial wound infection in 2 (4.9%) patients and post lobectomy empyema in 2 (4.9%) patients who required prolonged tube thoracostomy, antibiotics and chest physiotherapy. Mortality was recorded in 1 (2.4%) patient who was preoperatively on ventilator and had misplaced chest tube. That patient died on the 3rd post-operative day due to respiratory failure secondary tolung parenchymal injury.

Conclusion: Our study results support surgical management of congenital lobar emphysema as Lobectomy was found to be safe, reliable and effective in these children presenting with respiratory distress due to lobar emphysema. There were no significant morbidities in these patients and clinical outcomes were satisfactory and surgical management is reported to be treatment of choice in pediatric patients with CLE.

Keywords: congenital lobar emphysema, lobectomy, surgical management.

Introduction

Nelson described congenital lobar emphysema (CLE) for the first time in 1932 which was later designated by Robertson and James in 1951.[1] congenital lobar emphysema is a rare anomaly with its proportion has been reported to be 1 per 20-30000 births all over the world.[2] However, the incidence of CLE during pregnancy is yet uncertain.[3] In approximately 50 % of cases, the etiology of CLE remains idiopathic while remaining 50% have been shown to be associated with various mechanisms such as air-trapping, which can further be sub - divided into intrinsic and extrinsic subtypes.[4]

Early diagnosis is crucial and plays an important role in the management while in most of cases it is challenging to distinguish between CLE versus hyperinflation which may result from extrinsic bronchial obstructions such as lymph nodes, vessels, masses and/or cysts which may compress the bronchus and can produce obstructions in the valve. Moreover, it has been documented that more than 50 % of patients with CLE are not associated with that of airway anomalies. Surgical excision is the safe and most commonly used more of treatment having operative mortality rates ranging from 3 to 7%.[5] Different studies have documented left upper lobe to be most common involvement which is followed by right upper and middle lobes respectively.[6] It generally exhibits in the first month of life after birth but its diagnosis may get delayed up to 5-6 months in more than 5% patients.[3] It may also be diagnosed during course of pregnancy on antenatal ultrasound.[6]

The classical signs of presentation may range from mild tachypnoea, wheeze to severe dyspnoea and/or

Mild tachypnoea, wheeze to severe dyspnoea and/or with cyanosis in these patients. Congenital heart disease has also been shown to be associated with congenital lobar emphysema.[7]

Some of these cases are autosomal dominant in nature while most of them may be sporadicand bronchial obstruction may also be a result of external compressions by any aberrant vessel or internal narrowing by musosal flaps or the wall may be linked by herniation in the mediastinum.[9,10]

Male gender preponderance has been shown to be more common with a male to female ratio reported to be 3:1. However exact reason for this male gender preponderance is still unknown.[6,10-11] The treatment of choice in children with congenital lobar emphysema is surgical excision of the affected lobe[12,13] which has been effective and safe mode of treatment.

This study was conducted to assess the outcome of lobectomy in congenital lobar emphysema in pediatrics group of patients at Department of Thoracic Surgery of Nishtar Hospital Multan and Services Hospital, Lahore. There was scarcity of local data on this topic and few local case reports have only been available on this topic. The results of this study have reported baseline data of our local population to ascertain the role lobectomy in children with CLE.

Methods

This retrospective study was done at department of Thoracic Surgery, Nishtar Hospital Multan Multan and Services Hospital Lahore from October 2003 to December 2015 in which we reviewed the medical records of these 41 children with CLE undergoing surgical treatment between. All the patients of pediatric group who underwent lobectomy for congenital lobar emphysema were included in study. Demographic data, operative findings outcome of the procedure in terms of post operative complications and 30 days morbidity and mortality were recorded. All the patients presented with respiratory distress.

The diagnosis of CLE was done showing the presence of different respiratory symptoms, having typical radiological findings (such as routine chest X-rays in all cases and on chest CT scans where applicable, figure 1 demonstrate the HRCT scan chest of a baby with the diagnosis of CLE which shows a hyperinflated left upper lobe with shifting and compression of mediastinum), bronchoscopic findings and

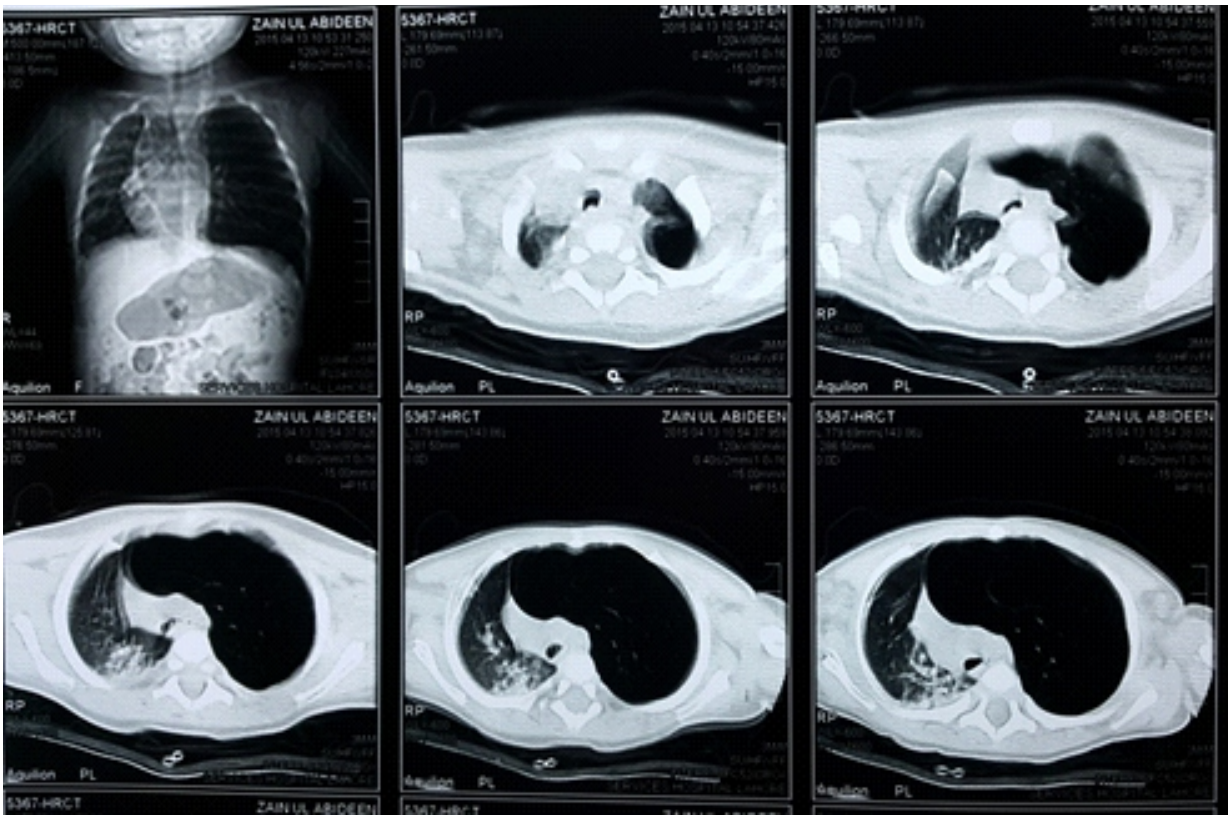

Fig-1: HRCT scan chest of patient with congenital lobar emphysema.

Pathological findings (figure 2 is the preoperative picture of CLE which clearly demonstrate the abnormal upper lobe with compressed lower lobe). Standard lobectomy was performed and bronchial stump was buttressed with surrounding lymph nodes, pericardial or pleural flap. SPSS version 20 was used for analysis purpose and frequencies/percentages have been determined for categorical variables and cross-tabulation was also done by applying chi square test at level of significance of 0.05.

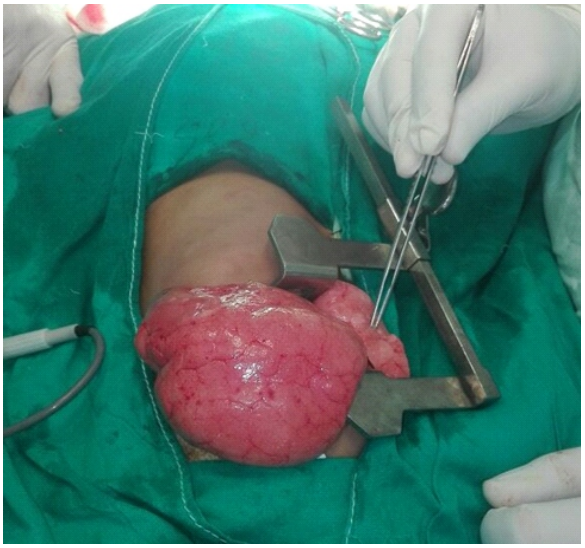

Fig-2: Operative Findings of Lobar Empysema.

Results

Our studied a total of 41 children with CLE in this time period of 12 years retrospectively. Of these 41 study cases, 30 (73.2%) were boys while 11 (26.8%) were girls and male to female ratio was 2.72:1.

Mean age of our study cases was 3.32 ± 1.59 months (with age range; 15 days to 7 months) while age wise distribution of study cases has been presented in Figure 3. Mean age of the boys was noted to be 3.38 ± 1.52 months while that of girls was 3.18 ± 1.84 months (p = 0.405). Of these 41 study cases, left upper lobectomy was performed in 27 (65.9%) patients, right middle lobectomy in 11 (26.8%) patients, left lower lobectomy in 2 (4.9 %) patients and right upper lobectomy in 1 (2.4%) patients.

Out of 41 patients 8 (19.5%) were preoperatively on ventilator. Out of 8 ventilated patients one had left sided chest intubation which was malpositioned and was in the lung parenchyma. Morbidity was superficial wound infection in 2 (4.9%) patients and post lobectomy empyema in 2 (4.9%) patients who required prolonged tube thoracostomy, antibiotics and chest physiotherapy.

Mortality was recorded in 1 (2.4%) patient who was preoperatively on ventilator and had misplaced chest tube. That patient died on the 3rd post-operative day due to respiratory failure secondary tolung parenchymal injury. These outcomes have also been presented in **table no. 3**.

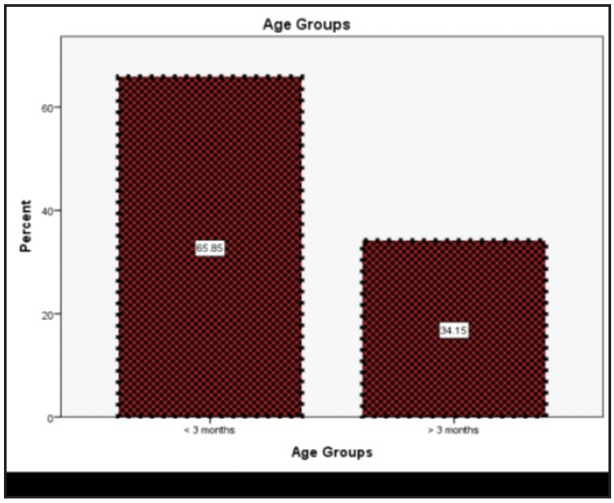

Fig-3: Age wise distribution of study cases. (N = 41)

Table-1: Stratification of lobectomy with regards to gender. (n=41)

Lobectomy	Gender Male (n=30)	Female (n=11)	P-value
Left Lower (n=01)	01	01	
Left upper (n=27	22	05	0.192
Right middle (m=11)	07	04	
Right Upper (n=01	00	01	
Total	41		

Table-2: Stratification of lobectomy with regards to age groups. (N = 41)

Lobectomy	Age Groups =3 Months (n=27)	>3 Months (n=14)	P-value
Left Lower (n=02)	00	02	
Left upper (n=27	17	10	0.126
Right middle (m=11)	09	02	
Right Upper (n=01	01	00	
Total	41		

Table-3: Distribution of outcome of the procedure among study cases. (N = 41)

Outcome	Frequency	Percentage
Empyema	02	4.9
Superficial wound infections	02	4.9
Expired	01	2.4
No complication	36	87.8

Discussion

Congenital lobar emphysema (CLE) is characterized by over distension and air-trapping in the affected lobe, and is one of the causes of infantile respiratory distress.[14] Regarding the diagnosis and treatment of congenital lobar emphysema (CLE) controversial findings have been reported, yet surgical excision of the affected lobe still remains widely employed mode of treatment while diagnosis of milder cases or even asymptomatic cases is generally followed by a more conservative form of management in these patients, i. e. non-surgical treatment and follow-up.[15]

In patients with CLE, male gender preponderance has been shown to be more common with a male to female ratio reported to be 3:1. However exact reason for this male gender preponderance is still unknown.[10,11] Similar findings have been observed in our study as well as out of these 41 study cases, 30 (73.2%) were boys while 11 (26.8%) were girls and male to female ratio was 2.72:1. Ozeik et al[14] from Turkey has also reported male gender predominance with 60 % boys which is close to our findings. However a study conducted in Brazil by Cataneo et al[16] reported equal distribution of male to female gender (50% each) which is different from these findings.

Mean age of our study cases was 3.32 ± 1.59 months (with age range; 15 days to 7 months) at the time of surgery. Mean age of the boys was noted to be 3.38 ± 1.52 months while that of girls was 3.18 ± 1.84 months (p = 0.405). Ozeik et al[14] from Turkey has also reported 4.9 ± 6.7 months mean age of the children with CLE undergoing lobectomy which is similar to that of our study results. A study conducted in Brazil by Cataneo et al[16] reported 6.9 months mean age at the time of surgery which is similar to our study results.

In majority of cases, most common involvement of left upper lobe has been reported [6]and similarly in our study, left upper lobectomy was performed in 27 (65.9%) patients, right middle lobectomy in 11 (26.8%) patients, left lower lobectomy in 2 (4.9 %) patients and right upper lobectomy in 1 (2.4%) patients. Ozeik et al[14]from Turkey has also reported left upper lobe involvement in 57 %, and right upper in 30 % which is in compliance with our study results. Morbidity was superficial wound infection in 2 (4.9%) patients and post lobectomy empyema in 2 (4.9%) patients who required prolonged tube thoracostomy, antibiotics and chest physiotherapy. Mortality was recorded in 1 (2.4%) patient who was preoperatively on ventilator and died on the 3rd post-operative day due to respiratory failure. A study conducted in Turkey by Dogan et al[13] and Cataneo et al[16] from Brazil have also reported similar results which are consistent with our findings.

Conclusion

Our study results support surgical management of congenital lobar emphysema as Lobectomy was found to be safe, reliable and effective in these children presenting with respiratory distress due to lobar emphysema. There were no significant morbidities in these patients and clinical outcomes were satisfactory and surgical management is reported to be treatment of choice in pediatric patients with CLE.

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Medical Guidelines

Lung Cancer CT Screening Guidelines Revised

Older, current and former heavy smokers should receive annual, low-dose CT screening, according to revised guidelines published in the Journal of the American Medical Association on Sunday. The revised guidelines follow, and in the JAMA paper are accompanied by, a systematic review of evidence on the role of CT screening for individuals at higher risk of lung cancer.

CT (computerised tomography) or CAT scans are a type of x-ray that can detect early signs of lung cancer, but they can give false-positive results. They use a computer to create detailed images of the inside of the body.

Regular chest x-rays produce less detailed images than CT scans and can also give false-positives. They are not recommended as a lung cancer screening test because there is no evidence they save lives.

Several groups collaborated in the systematic review, namely the American College of Chest Physicians (ACCP), the American Society of Clinical Oncology (ASCO), the American Cancer Society, and the National Comprehensive Cancer Network, with input from the American Thoracic Society (ATS).

The review concludes that:

"Low-dose computed tomography screening may benefit individuals at an increased risk for lung cancer, but uncertainty exists about the potential harms of screening and the generalizability of results."

The review forms the basis of clinical practice guidelines developed by the ACCP and ASCO and was endorsed by ATS.

The revised guidelines recommend that current and former smokers aged 55 to 74 who have smoked for 30 pack years or more, and either are still smoking or have quit in the past 15 years, should be offered low-dose CT screening over both annual screening with chest x-ray or no screening.

And, this offer should only be made in settings that can deliver the comprehensive care provided to National Lung Screening Trial participants, which essentially means only centers with specialist radiologists and surgeons.

The guidelines say CT screening should not be performed on current or former smokers who have not accumulated 30 pack years of smoking, or who are outside the 55 to 74 age range.

Sick people with limited life expectancy or whose illness is not likely to be cured should also be excluded.

The JAMA report has a full account of guideline remarks and explanations.

30 pack years is the equivalent of smoking one pack of 20 cigarettes a day for 30 years. For instance, two packs per day for 15 years is equivalent to 30 pack years.

The review included a large National Cancer Institute study involving more than 53,000 people with a 30 pack year history.

The study showed that CT screening prevented about 80 deaths from lung cancer over 6 years, but 16 participants died after CT screening, including 6 who did not have lung cancer.

Lung cancer is the leading cause of cancer death both globally and in the United States, where it causes as many deaths as the next 4 most deadly cancers combined: breast, prostate, colon, and pancreatic. Although lung cancer deaths in the US have fallen slightly since 1990, the disease is likely to kill more than 160,000 Americans in 2012.

The survival rate for lung cancer is poor, the current 5-year survival rate stands at 16%, and most people diagnosed with the disease are at an advanced stage (40% at stage IV, 30% at stage III).