Evaluation of early and late complications in patients with congenital lobar emphysema: A 12 year experience

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ABSTRACT

Background: Congenital lobar emphysema (CLE) is characterised by over distension of one lobe and pressure on the adjacent lobe and mediastinum. In this study, we review the pathological results of our paediatric patients with CLE, highlighting the early and late complications that occurred in these patients. **Patients and Methods:** In a prospective study from 1996 to 2008, we evaluated 30 patients with CLE diagnosis. Variables collected included sex, age at the time of diagnosis, radiological diagnostic method, type of treatment, pathological analysis, surgical findings and early postoperative complications. Parents were asked to refer to our clinic for follow-up and evaluation of late complications. **Results:** Thirty patients and males accounted for majority of the study population (n = 20, 67%). The mean age of male and female patients (at admission) was 7.2 ± 2.3 and 4.7 ± 1.2 months respectively (*P* = not significant). The main diagnostic method was chest x-ray (CXR) in all patients. Abnormal bronchial cartilage was found in 71% of patients. The most affected lobe was left upper lobe (50%). Associated anomalies were seen in four patients. Early postoperative periodhadtwo cases of pneumothoraces. At six month follow up, five (25%) males and four females (40%) had delayed weight gain. Permanent oxygen dependency was seen in two patients. Twenty- six patients underwent thoracotomy. Mortality rate was 13%. Base deficit at the time of admission was greater in those patients who eventually died, (-8.6 ± 1.2 versus –3.1 ± 0.4) (*P* = 0.0003). There were two deaths in the bilobar involvement group and two in the unilobar involvement group (*P* = 0.07, near significant). **Conclusion:** This study confirms that the number of affected lobes and base deficit at the time of admission were associated with significantly increased mortality.

Key words: Congenital lobar emphysema, lung malformation, mortality

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INTRODUCTION

Congenital lobar emphysema (CLE) is a rare anomaly of the lung that usually presents as acute respiratory distress. This malformation is characterised by overinflation of one or more pulmonary lobes leading to compression of the surrounding lung tissue and mediastinal shift.[1-3] CLE is classically the result of developmental deficiency of the cartilage that supports the bronchus to the involved lobe, but this specific defect is demonstrable in only one- third to two- thirds of surgically resected emphysematous lobes.[4] This study aims to evaluate the pathological results of our paediatric patients with CLE. We also assessed the early and late complications in these patients.

PATIENTS AND METHODS

In a prospective study from 1996 to 2008, 30 patients with CLE diagnosis were evaluated. Variables collected included sex, age at the time of diagnosis, radiological diagnostic method, type of treatment, surgical findings and early postoperative complications (cyanosis, oxygen supplement dependency, pneumothorax). Post-operative oxygen-dependency was determined based on the results of arterial blood gas measurement. An experienced pathologist reviewed the pathohistological findings of specimens. Patients were followed up for one year. Parents were asked to refer to our clinic for follow-up and evaluation of late complications such as growth retardation (based on growth charts), respiratory states (cyanosis, oxygen supplement dependency, wheezing, asthma and recurrent infection) and early fatigue (inability to sucking or playing perfectly based on history). We did not perform pulmonary function test routinely in our patients. Data were compiled into a single database and analyzed using non parametric tests (Chi square, Fisher's exact, Mann-Whitney) to compare the above parameters over the two groups (alive vs. deceased patients). Data were analysed using SPSS 13.0 (SPSS, INC, Chicago, IC; and *P* less than 0.05 was considered significant.

RESULTS

A total of 30 patients with CLE diagnosis were admitted or transferred to paediatric surgery service at Isfahan university medical centre (Al-Zahra hospital), in the period from July 1,1996 to July 30 2008.

Males accounted for majority of the study population (n = 20, 67%), as compared with 33% (n = 10) females. All diagnosis was postnatal, we have noprenatal diagnosis. The mean age of male and female patients (at admission) was 7.2 ± 2.3 and 4.7 ± 1.2 months respectively (P = NS). The main diagnostic method in all patients was CXR. Chest CT scan was used in two patients for documentation. Table 1 demonstrates the study variables stratified by sex of patients. Table 2 demonstrates the pathological results of patients. Abnormal bronchial cartilage was found in 71% of patients. Sixteen patients required preoperative ventilation. Postoperative ventilation wasused in 12 patients.

The most affected lobe was left upper lobe (50%). Unilobar involvement was seen in 26 patients (16 boys [80%] and nine girls [90%]). Bilobar involvement was seen in one girl and four boys. Associated anomalies were seen in four patients (congenital heart disease as opening up of PDA, VSD). In one patient, neonate had respiratory tachypnea , dispnoea and wheezing at birth and managed as respiratory distress syndrome with surfactant, but a few days later, typical radiological findings of CLE appeared.

In early postoperative period, there were two cases (girls) of pneumothoraces. These patients were on postoperative ventilator support. Although, all our patients had term delivery and normal birth weight (weight >2.5 kg) but at six-months follow-up, five (25%) males and four females (40%) had delayed weight gain based on growth chart. The height of three males (15%) and two females (20%) was shorter than normal range. Permanent oxygen dependency was seen in one boy and one girl. Twenty six patients underwent thoracotomy because of progressive respiratory dyspnoea associated with abnormal radiological findings. Lobectomy was the main operative treatment in these patients. For multi-lobar involvement, affected lobes were removed. Bilobar disease was operated on the same day. The clinical course of patients treated conservatively was good. They were followed up with serial x-rays.

Mortality rate was 13.3% (four patients), occurring in three boys and one girl (P = NS). There were two deaths in the bilobar involvement group and two in unilobar involvement group (P = 0.07, near significant). There was no difference in clinical presentation while comparing the survivors with the the dead. Congenital heart disease was higher in those who died (3 vs. 1, P = NS). Initial postoperative oxygen dependency, was different between survivors and death patients (P = 0.07, near significant).

The mean age of in those patients who died was 3.2 ± 2.5 months. Of note, base deficit at the time of admission was greater in those patients that eventually died, (-8.6 ± 1.2 vs. –3.1 ± 0.4) (P = 0.0003). CO2 retention was similar in groups. There was no significant difference for PO2 and PCO2 between groups.

Table 1: Study variables stratified by sex of patients

Variable	Male (n=20)	Female (n=10)	*P*
Number of affected lobe (S) $\left(\frac{\text{one}}{\text{more}}\right)$	$\frac{17}{3}$	$\frac{9}{1}$	NS
Postoperative wound infection	1 (5)	2 (20)	NS
Postoperative oxygen dependency	1 (5)	0	NS
Pneumothorax	0	2 (20)	NS
Low weight	5 (2)	4 (40)	NS
Short height	5 (25)	4 (40)	NS
Early fatigue	0	2 (20)	NS
Death	3 (15)	1 (10)	NS

Figures in parentheses are in percentage.

Table 2: Pathological results stratified by sex study

	Abnormal bronchial cartilage (%)	Alveolar hyperplasia (%)	Bronchial Artesia (%)
Male	14 (77)	2 (11)	1 (12)
Female	6 (66)	2 (22)	1 (12)
Total	20 (71)	4 (16)	2 (12)

DISCUSSION

CLE is a rare anomaly of the lung and is characterised by over inflation of pulmonary lobe, air getting trapped in the effected lobe, compression of the surrounding lung tissue, mediastinal shift and emphysematous lobe herniation.[5] In this study, we report 30 patients with CLE during 12 years.

In our patients, abnormal bronchial cartilage was the most common pathological finding, as noted by others.[5,6] We documented an association between number of the affected lobes and increasing in mortality despite controlling for multiple factors that effect outcome in CLE patients. The mortality in this series was high, and well above most reports in the contemporary literature.[7,8] It seems that in multiple

lobar involvements, pulmonary function is not the best after lobar resection and this can cause an increase in mortality. This hypothesis can be documented by comparison of pulmonary function test in CLE patients based on number of affected lobar. Although several authors [7-9] have reported a good clinical course after lobectomy with CLE,none of these results are in patients with unilobar involvement. In four patients, we used conservative management with good results. Conservative treatment can be a preferable choice in mildly symptomatic patients. This is similar to findings of Ulku *et al.*[10] who reported successful conservative management in 20% of their patients. Eigen *et al.*[11] show that lung growth and function in children treated conservatively was no different from children with lobectomy. This finding is also consistent with Man *et al* study.[12]

We also demonstrate that base deficit at the time of admission is an independent factor predicting mortality. Abnormal base deficit reflects tissue hypo perfusion. Admission base deficit has also been documented as a predictor of outcome in trauma; including mortality, intensive care unit (ICU) and hospital length of stay.[13,14] However, its ability to predict the outcome of CLE patients is a new hypothesis. Respiratory dyspnoea appears to have the potential to increase respirator muscles metabolism and lactate in these patients and hyper lactemia could increase base deficit. Dyspnoea may be associated with hypoxia and respiratory acidosis. Lactic acidosis and increased base deficit may be because of shock associated with hypoxia and acidosis. Although prolonged hypoxia is not common situation for CLE patients after lobectomy, but in some patients, residual volume and lung capacity changed post operatively, on the other hand, concomitant cardiac anomalies can be an important factor for post operative oxygen dependency.

In summary, this study confirms that the number of affected lobes and base deficit at the time of admission were associated with significantly increased mortality.

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