ORIGINAL ARTICLE

CONGENITAL LOBAR EMPHYSEMA

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Abstract

Background: Congenital lobar emphysema (CLE) is relatively an uncommon congenital lung malformation presenting with respiratory distress of varying severity mostly during the neonatal or the infantile period due to air trapping in the affected lobe or lobes.

Aim: The authors review surgical experience in this congenital malformation; the simplest way of diagnosis and the standard treatment of choice in addition to the comparison of this study with other international studies.

Patients and Methods: Eighteen patients with congenital lobar emphysema were successfully managed during a ten years period (1999-2008) at the Thoracic Department of the Medical City Teaching Hospital in Baghdad, Iraq.

Results: These patients were studied according to their age, sex distribution, presenting symptoms, physical findings, and methods of diagnosis, surgical treatment, thoracotomy findings and the postoperative outcome.

Conclusion: Though the diagnosis can be accomplished by chest X-Ray in the upright position, the use of a more advance technology has resulted in the early identification and serial evaluation of this congenital anomaly and the treatment of choice is surgery, as lobectomy of the involved lobe offers the best chance of cure.

Keywords: Congenital lobar emphysema CLE, lobectomy, bilobectomy.

INTRODUCTION

Congenital lobar emphysema is an important cause of respiratory distress in the neonatal and infantile period characterized by over inflation, over distension and air trapping of the affected lung lobe or occasionally, more than one lobe, leading to compression and collapse of the adjacent healthy lobe associated with herniation of the emphysematous lobe through the mediastinum into the contra lateral side.

Many factors have been incriminated as causing congenital lobar emphysema. These include bronchomalacia, bronchial stenosis, redundant bronchial

mucosa, aberrant blood vessels, and other forms of bronchial obstruction, as well as intrinsic alveolar defects, either alone or in combination.[1]

This manuscript describes the authors experience in treating eighteen patients with congenital lobar emphysema during ten years (1999-2008) at the thoracic department of the Medical City Teaching Hospital in Baghdad, Iraq.

PATIENTS AND METHODS

This is a retrospective and comparative study of eighteen patients, with varying degrees of respiratory difficulty and the query diagnosis of CLE, who were admitted to Thoracic Department of the Medical City Teaching Hospital for evaluation.

A careful history was taken from their mothers, a detailed physical examination was done and a plain Chest X-Ray was taken both in the postero-anterior (PA) and lateral views in the upright position while in two of the patients bronchoscopic examination was needed to confirm the diagnosis.

A special formula was used to describe the patients with regards to their age, sex distribution, presenting symptoms, radiographic findings and bronchoscopic findings.

Preoperative preparation regarding the administration of oxygen to the patients to decrease the dyspnea as well as antibiotics to combat chest infection were also done.

Surgery was conducted successfully to remove the emphysematous lobe or lobes leading to an excellent outcome.

RESULTS

Of the eighteen patients studied with the diagnosis of CLE, ten were males (55.5%), and eight were females (44.4%).

The youngest patient was a twenty – five days old boy who presented with severe respiratory difficulty and attack of cyanosis, while the oldest was a two years old girl who presented with recurrent chest infection and progressive shortness of breath. The age distribution of the patients are illustrated in (Fig. 1) which shows that eleven patients presented within the first two months of life (61.1%) and sixteen patients were seen within the first six months of life (88.8%).

These patients presented with cough, varying degrees of dyspnea, recurrent chest infection and cyanosis and the distribution of these symptoms were elucidated in Table 1.

The earlier the baby presented, the more severe the dyspnea, and the later the patient presented, the higher the recurrent chest infection.

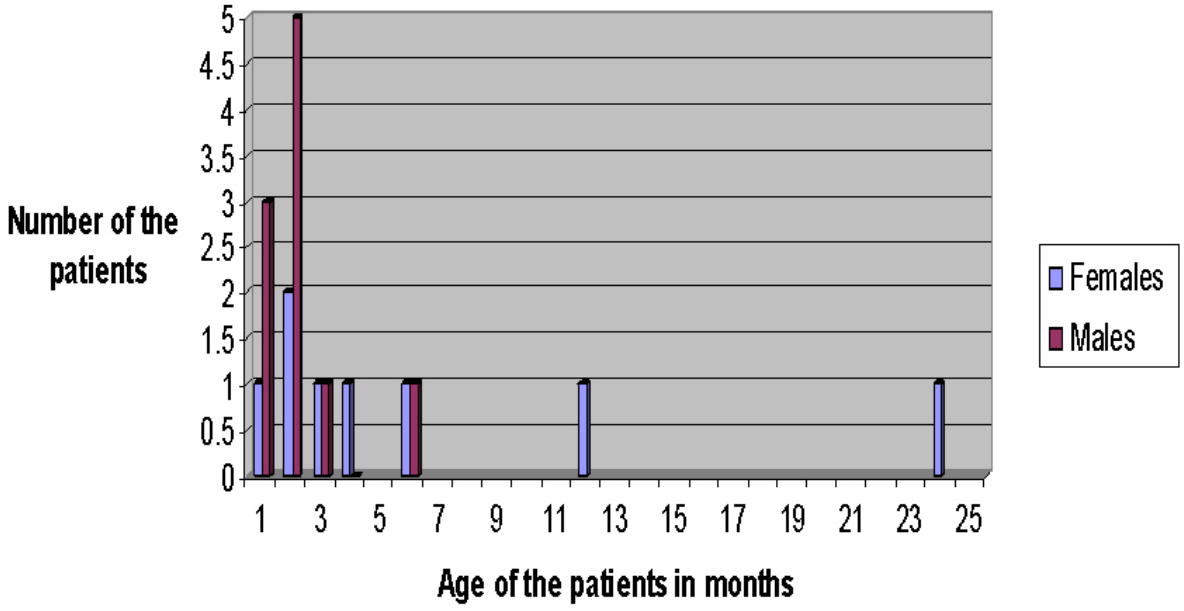

Fig 1. Age distribution of the patients.

Table 1. Clinical presentation of the patients.

Presentation	Number	%
Respiratory Distress	8	44.44%
Dyspnea	7	38.88%
Repeated chest infection	2	11.11%
Cyanosis	1	5.55%

In all the patients studied, a chest x-ray in upright posture in both the posterior-anterior and lateral views was the main diagnostic tool for the diagnosis of CLE; which clearly shows the emphysematous lobe, compression of the adjacent lobe and herniation of the emphysematous lobe into the contrlateral side. (Fig. 2) shows a CLE of the left upper lobe while (Fig. 3) a CLE of the right middle lobe .Bronchoscopy was used only in two patients with a doubtful diagnosis and this confirmed the presence of the left upper lobe bronchial stenosis.

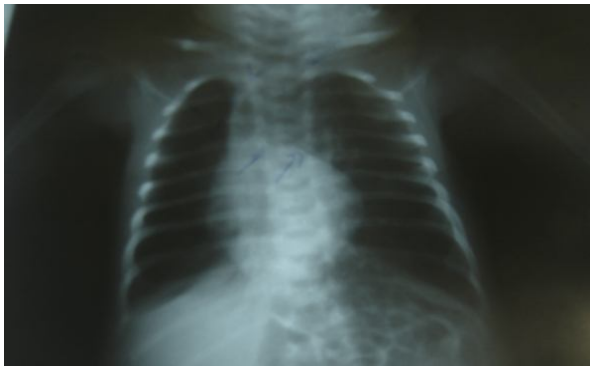

Fig 2. Congenital Lobar Emphysema left upper lobe.

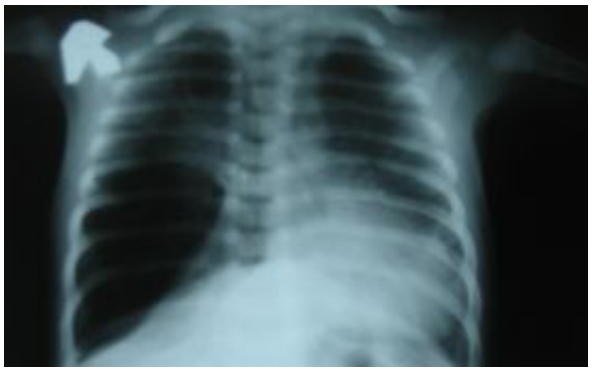

Fig 3. Congenital lobar emphysema right middle lobe.

In all patients there was no associated congenital anomaly detected.

A careful preoperative preparation which included administration of antibiotics to control the already present chest infection and oxygen administration to combat dyspnea.

Written high risk consent was taken from the parents after explaining all the possible operative and postoperative risks.

Surgery was performed as early as possible under general anesthesia with the baby in the lateral decubitus position while the anesthetist was instructed not to over ventilate prior to opening the thoracic cavity to avoid the danger of increasing the emphysematous changes and herniation through the mediastinum which may lead to a more serious respiratory Insuffiency.

Classical thoracotomy was done through the 4th intercostals space. Left thoracotomy was done in ten patients and right thoracotomy in the other eight. The affected lobe or lobes were resected. The left upper lobe was resected in ten patients while the right middle lobe was resected in five other patients and both the right middle and upper lobes were resected in the three other patients, resulting in a total lobe resection of twenty-one and as shown in Table 2.

Table 2. Distribution of the resected lobes.

Lobe affected	No. of the patients	Lobes resected
Left Upper Lobe	10	10
Right Middle Lobe	5	5
Right middle lobe & Right upper lobe	3	6
Total Number	18	21

The obtained histopathological reports seven to ten days after, confirmed the diagnosis of CLE of the resected lobes.

All the patients had a smooth postoperative course, with the remaining lobe or lobes expanded to fill the thoracic cavity. Two patients developed postoperative wound infection which was treated conservatively and a third patient developed post-operative severe chest infection and respiratory difficulty that necessitated admission to the respiratory care unit to maintain assisted ventilation for few days, after which the patient improved gradually and was discharged on the 18th Postoperative day. The rest of the patients were discharged from the hospital on the 7th Post-operative day.

There was no mortality. The patients were doing well during the follow up period.

Table 3. Comparison between different studies.

No.	Study	No. of Patients	Duration of the study
1	Ibrahim Kanara	14	8 Years
2	Ugur Ozcelik	27	30 Years
3	Dogan R. Demicrin	9	10 Years
4	AH al-salem	3	3 Years
5	J.C.R.Lincoln	28	15 Years
6	Waleed Mustafa Hussen	18	10 Years

DISCUSSION

CLE is uncommon but not rare in Iraq. The Thoracic Department of the Medical City Teaching Hospital is concerned with the management of the Pediatric Thoracic emergency cases.

Eighteen patients with this congenital malformation were admitted under our care during a ten years period (1999-2008); and this relatively small number of patients is comparable to other studies as shown in Table 2.

Male patients were more commonly affected constituting (55.5%) in our study and this is comparable to other studies,[2-4,6,7] in contradistinction to the study done by Stiger KB et al[5] reporting females having been affected more than males.

Most of our patients were presented during the neonatal and infancy period& specially during the first six months of life, and this is also seen in other studies.[2-4,6,7]

Respiratory distress and dyspnea were the major presenting symptoms seen in fifteen patients, recurrent chest infection in patients who presented late during the second year of life; and cyanosis was seen in only one patient who presented early within the first month of life, and this in agreement with other studies.[2-4,6]

Plain chest X-Ray in both the posterior anterior and lateral views taken in the upright posture was the main diagnostic tool and this is in agreement with the other studies;[R6-8] while radio- isotope study of the lung, Computerized Tomography (CT) and Magnetic Resonance Imaging (MRI) which had been used in other studies[2,3,5] were not used in this study of ours .Recently the prenatal diagnosis of CLE which was achieved by prenatal ultrasonagraphy and ultra-fast fetal MRI has resulted in early identification of the lesion with ready access to postnatal surgical intervention.[9]

Pre-operative bronchoscopy (rigid or flexible) is rarely used for diagnosis in suspicious cases. It shows stenosis of the orifice of the affected lobe.[3,5]

Conservative treatment in CLE has no place in this study, apart from the pre-operative antibiotics and oxygen administration to reduce the dyspnea and chest infection, in contradistinction to other studies in which it was used as a main step in the treatment of some cases of CLE which were usually older children with milder symptoms.[2,5]

The left upper lobe was the most common lobe affected, with the next, being the right middle lobe and this is in agreement with other studies[2,4,7, 8]

Right upper and middle lobe involvement which was seen in three of our patients was also reported by Dogan et al[4] and by Ugur et al[3], but cases of bilateral CLE which is reported by Okazaki[10] and Kumart[11] was not seen in this study.

There is no mortality reported in this study which compares with others;[2,6] while the lower mortality rate seen in other studies[3,7] was due to the associated congenital cardiac disease or multi lobar disease.

In conclusion we conclude that diagnosis although can be accomplished by simple chest X-ray in the upright position, but the use of CT , lung perfusion study , MRI and recently prenatal ultrasonography has resulted in the early identification and serial evaluation of this congenital lung lesion.

Surgery of the affected lobe / lobes through resection was the treatment of choice and offered the best chance of cure.

REFERENCES

1. Lucian L. Leape, Luther A. longino. Infantile Lobar Emphysema, Pediatrics. 1964;34:246-55.

2. Ibrahim Karnak, Mehmet Emin, Senocak, et al. Congenital Lobar Emphysema. Diagnostic and therapeutic considerations, Journal of Ped. Surgery. 1999;34:1347-51.

3. Ugur Ozcelik, Ayhan Gocmen, Nural Kiper, et al. Congenital Lobar Emphysema: Evaluation and long term follow up of a thirty cases in single center, Pediatric Pulmonlogy. 2003;35:384-91.

4. Dogan R, Demircin M, Sarigul A, et al. Surgical management of Congenital Lobar Emphysema, Turk J Pediatr. 1997;39:35-44.

5. Stigers KB, Woodring JH, Kanga JF. The clinical and imaging spectrum of findings in patients with congenital lobar emphysema , Pediatr Pulmonol. 1992;14:160-70.

6. AH al-salem,Y Adu –Gyamfi and CS Grant. Congenital Lobar emphysema, Canadian Journal of anesthesia. 1990;37:377-379.

7. JC R. Lincon , J Stark , S Subramanian et al ,Congenital Lobar Emphysema , Ann Surg. 1971;173:55-62.

8. Osman Faruk Senyuz, Nur Danismend ,Ergun Erdogan et all , Congenital Lobar Emphysema –A report of 5 cases, J, Surgery Today. 2006;2813:64-767.

9. N. Scott Adzick, Prenatal diagnosis and management of congenital lobar emphysema, Journal of Pediatric Surgery. 2000:792-5.

10. Okazaki Tadaharu, Kono Sumio, Urushihara Naoto et al, Management of Bilateral Congenital Lobar Emphyseam , Japanese Journal of Pediatric Surgery. 2002;34:1380-5.

11. Kumar T Sathish, Simon Anna, Sen Sudipta, Bilateral congenital emphysema. Unusual cause for recurrent respiratory tract infection, Jiaps. 2006; 11:151-3.