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CONGENITAL LOBAR EMPHYSEMA*

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Congenital lobar emphysema is among the most important causes of respiratory distress in newborns and infants, and sometimes the appearance of the symptoms may be delayed to a later age. The pathology of lobar emphysema is generally characterized by hyperinflation or overdistention of a lobe[1]. Early diagnosis and treatment are required since an extremely distended lobe causes pressure on the neighboring lobes and mediastinum resulting in mediastinal shift and respiratory distress[1,2].

Clinical Presentation

For this study 15 cases of congenital lobar emphysema which had undergone surgical resection in our clinic between May 1965 and September 1982 were analyzed. Nine cases were boys and 6 were girls. The youngest patient was 3 days old when operated on and the oldest 18 months of age (average: 4.5 months). Ten subjects were referred to us as cases of severe respiratory distress since birth and 5 on account of repeated pulmonary infections, retarded growth and mild respiratory distress. Symptoms of the patients and localizations of the pathology are shown in Table I. Lobectomy was performed in 14 cases and a right pneumonectomy in

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1 case on an emergency basis. In this last case, all the lobes on the right side were involved by the pathology. Four patients (26.6%) expired in the early postoperative period (gastroenteritis and sepsis: 1, persistent pulmonary infection: 2, severe bronchospasm and pulmonary insufficiency: 1). Ten patients were discharged in a satisfactory condition, and long term follow-ups (average 2 years) of these showed normal growing children without any respiratory distress. One patient was referred back with repeated pulmonary infection.

In pathological studies none of the resected specimens was easily collapsible by pressing. The pathologic specimens showed sponge-like structure of the lung. Emphysematous alveoli were detected in all cases. In 4 patients (26.6%) hypoplasia of the bronchial cartilage was noted by serial microscopic slices. Uniform subpleural (visceral) bullae of 0.1-0.7 cm in size were found in 3 cases (20%).

Some interesting chest x-rays of our patients are shown in Figures 1a, b and 2a, b.

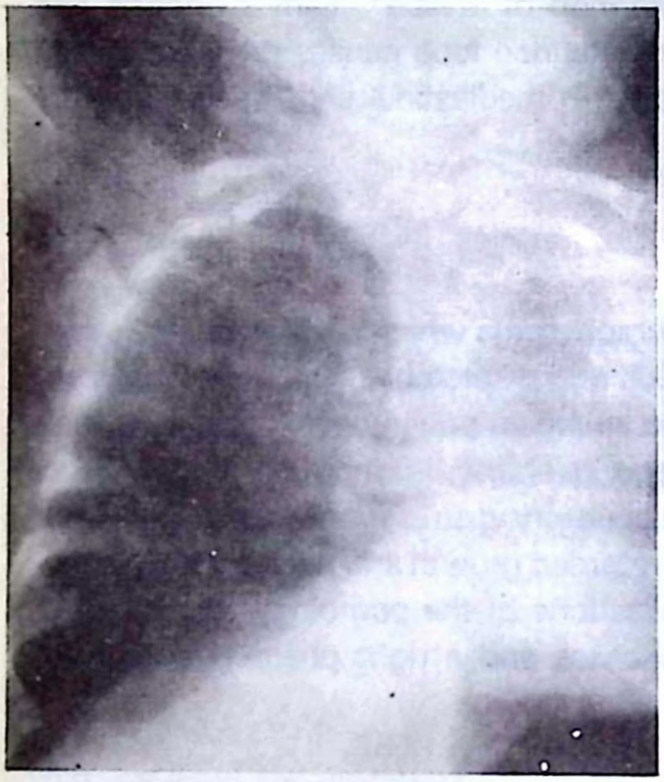

Figure 1a

X-rays taken preoperatively.

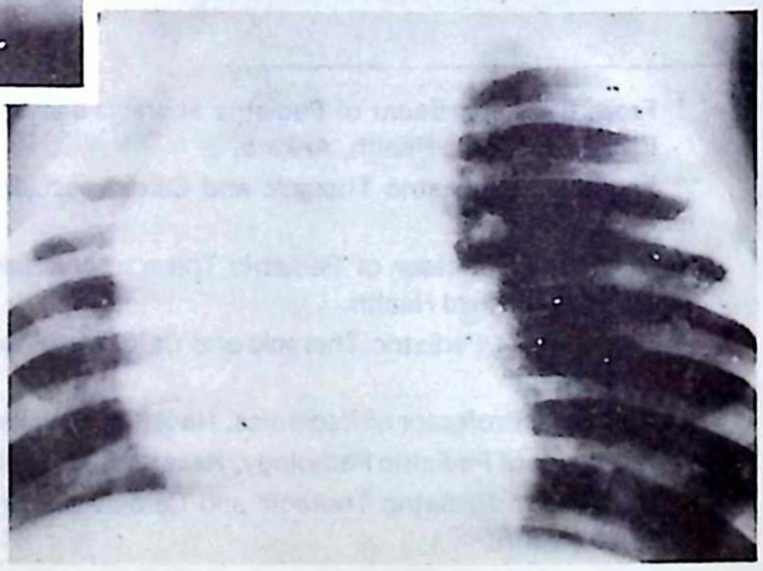

Figure 1b

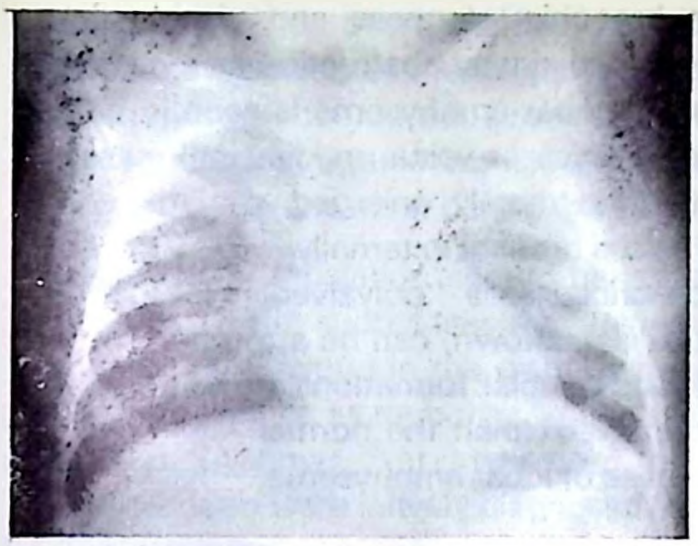

Figure 2a

X-rays taken postoperatively.

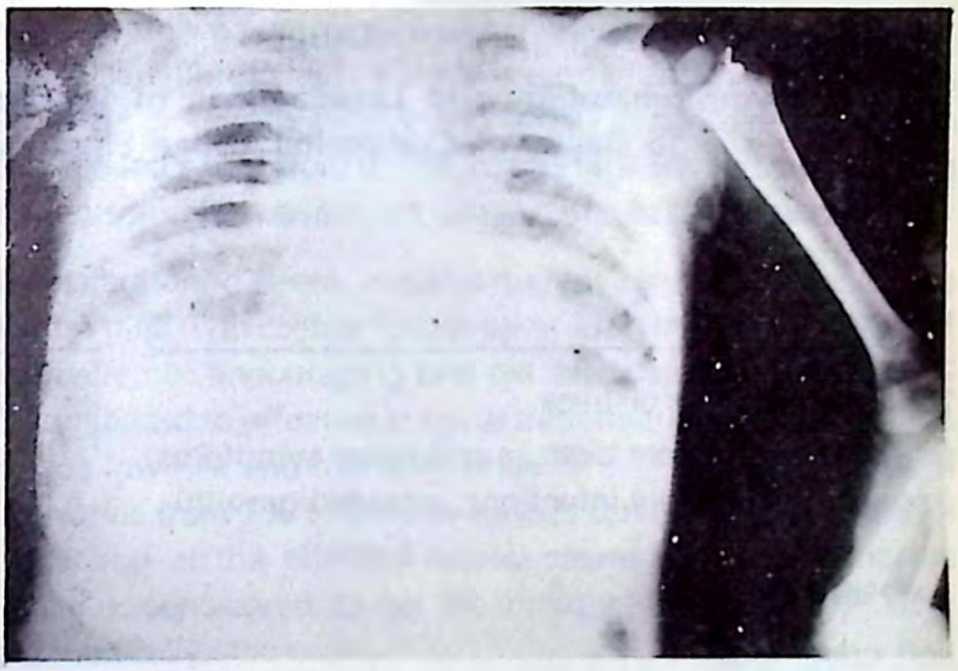

Figure 2b

Discussion

Congenital lobar emphysema usually develops in one lobe of a lung, and it is most commonly encountered in the left upper lobe (43%), followed by the right middle lobe and then the right upper lobe. The localizations in our cases are shown in Table I. On occasion more than one lobe is involved in the pathology[2,3] as seen in our case who underwent right pneumonectomy. The exact pathogenesis is not known in 50% of the cases despite careful pathological studies[2]. It is believed that whatever the cause may be, there is partial airway obstruction giving rise to a check-valve mechanism. Some publications describe a segmental or diffuse dysplasia or hypoplasia in the bronchial cartilage formation[2-7]. We have established hypoplasia in the bronchial cartilage formation in four of the cases. Murray[7] reported bronchial hypoplasia in 79 out of 166 patients (53.6%). Cartilage deficiency leads to the collapse of the bronchi leading to an extreme degree of emphysema during expiration. Endo-

bronchial mucosal webs, post-infectious bronchial stenoses and viscid sticky mucous plaques also cause emphysema by partial airway obstruction as a result of the flap-valve mechanism[8,9]. Less commonly, lobar emphysema is seen in cases with abnormal branching of the pulmonary arteries or veins, mediastinal masses, hypertrophied lymph nodes; bronchial sling, extremely enlarged left atria, and patent ductus arteriosus exerting pressure on the bronchi externally[2,3,5,10]. Recently, a new pathological entity has been described as a "polyalveolar lobe"[11,12]. Cases of lobar emphysema, where the cause is unknown, can be attributed to this new entity. In these subjects, the bronchial and alveolar formations are normal, but the emphysematous lobe contains far more alveoli than the normal[11,12]. Loss of recoil elasticity has also been reported as a cause of lobar emphysema[13,14].

TABLE I

Symptomatology and Localizations of the Pathology in 15 Cases of Congenital Lobar Emphysema

Symptoms	Number of Patients	%
Severe respiratory distress	10	66.6
Moderate respiratory distress and other symptoms (repeated pulmonary infections, retarded growth)	5	33.3
Localizations		
Left upper lobe	8	53.3
Right upper lobe	3	20.0
Right middle lobe	2	13.3
Right lower lobe	1	6.7
Right lung (all 3 lobes)	1	6.7

Symptoms of congenital lobar emphysema become apparent early in life in the majority of cases, mostly soon after birth and in infancy[15]. Signs and symptoms generally consist of tachypnea, dyspnea, intercostal and supraclavicular retractions, progressive cyanosis, growling breathing sounds and expiratory wheezing on the affected side. In 66.6% of our cases the patient was referred with severe respiratory distress. In 15% of cases there is a related congenital heart disease such as patent ductus arteriosus, a ventricular septal defect or tetralogy of Fallot. None of our cases had an associated congenital heart disease, but we did encounter a chest-wall deformity (pectus excavatum) in one of our patients, as an associated malformation.

Radiologically, an overdistended, radioluscent emphysematous lobe is seen along with the atelectasis of the neighboring lobes. Mediastinal shift to the opposite side is frequently encountered. Bronchography is not advised in such cases, owing to the danger of iatrogenic bronchial obstruction by the contrast material which might prove fatal to the patient during the study[2].

Foreign body inhalation should be considered in differential diagnosis. Unilateral hyperinflation in such cases may mimic congenital lobar emphysema[16]. Bronchoscopy is very helpful in the diagnosis and removal of foreign bodies or endobronchial lesions such as mucous plaques simulating the radiologic picture of congenital lobar emphysema[2,16] and was used to remove sunflower seeds in two patients in our clinic who were initially diagnosed as lobar emphysema.

The specimens removed by surgery do not generally show any specific changes in pathological studies. Dysplasia of bronchial cartilage is seen in 25% of the cases[5,6]. Macroscopically, the lobe has a smooth surface and is stiff in consistency, pink in color, and air cannot be expelled easily from it. The cut surface shows a sponge-like appearance. Emphysematous and torn alveoli are usually seen microscopically[2].

In the presence of slight respiratory distress, medical therapy consisting of selective bronchial intubation may be tried with clinical follow-up at short intervals[13]. Trials of medical treatment are usually not encouraging and are associated with high mortality rates such as 50% compared to effective surgical treatment by resection of the involved lobe(s), which has low risk and mortality rates[6-8,17-19]. Reported surgical mortality in the literature varies from 7% to 21% in various series[7,8]. In our series it was 26.6%. Surgical resection of the affected lobe(s) seems currently to be the method of treatment that is considered to be the most effective[8,15,17,19]. Our patients were not treated medically.

Summary

A series of 15 patients with congenital lobar emphysema is presented. Lobectomy was performed in 14 cases and pneumonectomy in 1. Four patients died due to various causes (26.6%). An average follow-up of 2 years in the surviving individuals showed satisfactory results. We believe that surgical resection of the involved lobe(s) is the only effective method of treatment for this congenital malformation.

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