Original Article

CONGENITAL CYSTIC LESIONS OF THE LUNGS IN CHILDREN

Nasir Iqbal, Ahmad Shahbaz,Aasim Malik, M Nawaz Anjum.

ABSTRACT

Background: *Congenital cystic lesions of the lung are uncommon but share similar embryologic and clinical characteristics. The purpose of this study is to review experience of congenital cystic lung disease at a tertiary children's hospital.*

Patients: *From 1999-2000, 10 patients (6 males and 4 females) under 5 years underwent evaluation and surgical treatment for congenital cystic lung disease. Four patients were under 1 year of age, and six were over 1 year of age. There were 6 bronchogenic pulmonary cysts, 2 congenital cystic adenomatoid malformations (CCAM) and 2 congenital lobar emphysemas.*

Results: *All patients under 1 year of age showed respiratory distress with mediastinal shift but no episodes of infection. In contrast, 3 of the 6 patients over 1 year of age had symptoms of recurrent chest infection. Lobectomy was performed in 4 patients and excision of the cyst in 6 patients. There was no postoperative mortality or morbidity. 8 patients at short-term follow-up from 2 months to 10 months after surgery are doing well with no subsequent limitation of physical activities due to lung resection.*

Conclusions: *In patients under 1 year of age, cystic lesions were noticed by respiratory distress; and in patients over 1 year of age signs of infection were predominant. Early diagnosis of these rare congenital cystic lung lesions would lead to the immediate, proper surgical treatment.*

Key Words: *Bronchopulmonary malformation, Congenital cystic lung lesions.*

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INTRODUCTION

Congenital lesions of the lung are relatively rare among pulmonary diseases that require operation. Nevertheless, precise diagnosis and therapy are necessary for optimal management and may be life saving for the neonate or infant[5].These diseases may result from compromised interaction between embryologic mesoderm and ectodermal lung components during development[4,9]. Pulmonary sequestration, Congenital lobar emphysema, Congenital cystic adenomatoid malformation (CCAM), and broncho-pulmonary cysts are four major congenital cystic lesions in the lung, that share similar embryologic and clinical characteristics[7]. This study retrospectively reviews our experience at the Children's Hospital Lahore of congenital cystic lung disease.

Address for correspondence
Nasir Iqbal
Surgical Unit 1
Services Hospital, Lahore. Pakisatan.
Tel No.+92-42-7588806.
E-mail: drnasiriqbal@hotmail.com

MATERIALS AND METHODS

From April 1999 to November 2000, 56 patients under 15 years of age with thoracic surgical conditions underwent evaluation and surgical treatment in the Department of Thoracic Surgery of the Children's Hospital & Institute of Child Health Ferozepur Road Lahore. Congenital cystic disease of the lung was seen in 10 patients. Of these 10 patients 6 were males and 4 were females, ranging in age from 43 days to 5 years of age.4 patients were under 1 year old, and 6 were over 1 year old. There were 6 broncho-pulmonary cysts,2 Congenital cystic adenomatoid malformations (CCAM) and 2 Congenital lobar emphysemas. Comparison regarding the clinical manifestations between the patients under 1 year of age and those over 1 year of age was done. Diagnostic studies included standard chest X-Rays and CT scans. Surgical therapy was mainly indicated which was based on the symptoms of respiratory distress and episodes of infection in addition to the clinical diagnosis of congenital cystic disease of the lung.

RESULTS

A 2 month old male child presented in the Emergency of Children's Hospital with severe dyspnea. Symptomatic treatment (e.g. oxygen therapy, bronchodilators and steroids) was immediately instituted. His chest X-Ray showed opacity in the left hemi thorax occupying the upper and

mid zones. His condition deteriorated after few hours. Prompt surgery was planned and done. Left postero-lateral thoracotomy revealed upper emphysematous lobe herniating towards the mediastinum and right hemithorax. Left upper lobectomy was done. Histo-pathology confirmed the diagnosis of infantile lobar emphysema. Post-operative recovery was uneventful.

Another patient, 43 days old, came in the Emergency with the complaints of shortness of breath for 7days

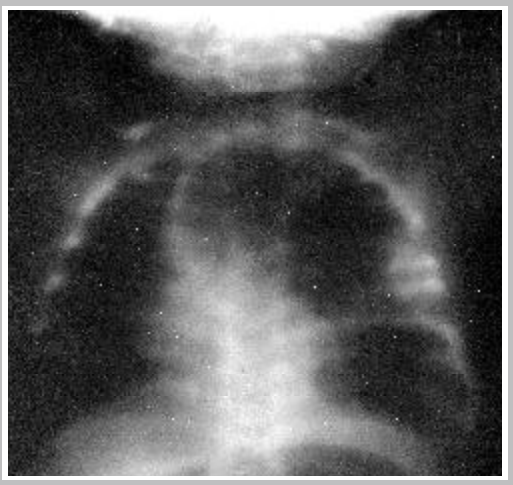


Fig:1 (X-Ray Chest PA View)

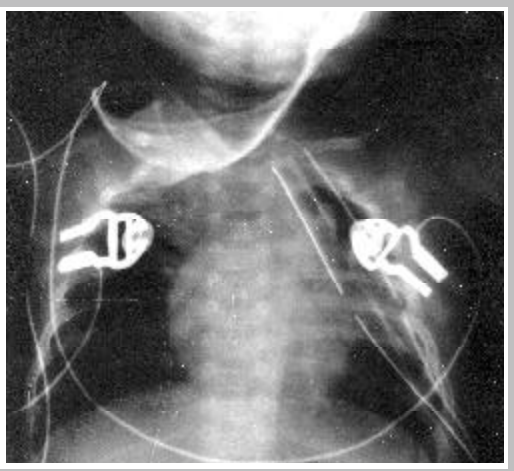


Fig:2 (X-Ray Chest PA View)

and fever for 5 days. Shortness of breath was gradually increasing in intensity with the passage of time as observed by the parents. His chest X-Ray showed an opacity in the right hemi thorax, occupying 3/4th of the hemi thorax.

Right postero-lateral thoracotomy for middle lobectomy was done. Histo-pathology report gave the diagnosis of congenital cystic adenomatoid malformation (CCAM).

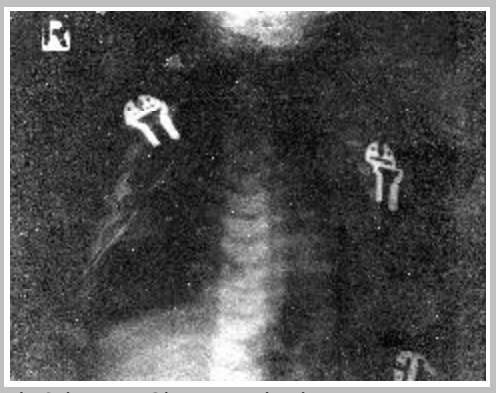


Fig:3 (X-Ray Chest PA View)

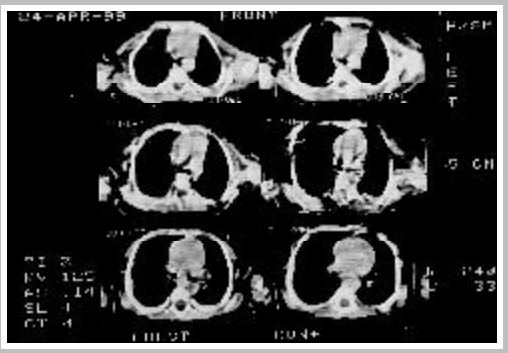


Fig: 4 (CT Scan Thorax)

A 4 months old, female child referred from the Medical ward of Children's Hospital Lahore with the complaints of shortness of breath (gradual) for the last 1 month, cough and vomiting for 1 month and fever for 15 days. There was difficulty in taking oral feed as well. On chest x-ray there was hyper lucency in the right hemi thorax, mimicking right pneumothorax. CT scan of chest was done, which showed a large cyst compressing the healthy lung.

Right postero-lateral thoracotomy was done for the

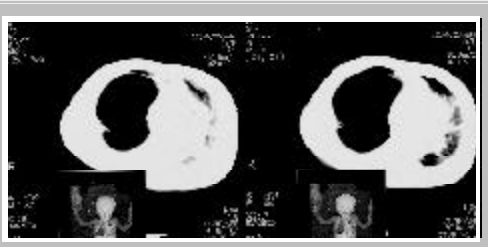
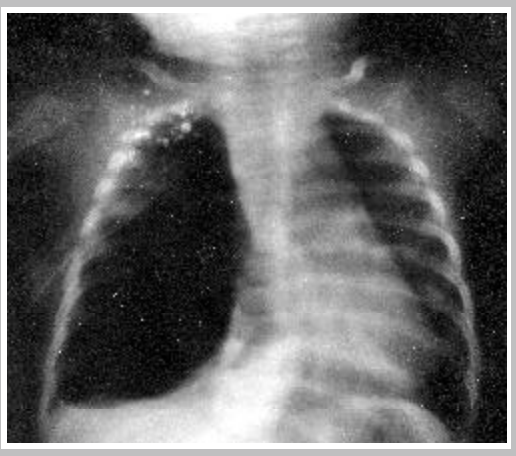


Fig:5 (CT Scan Thorax)



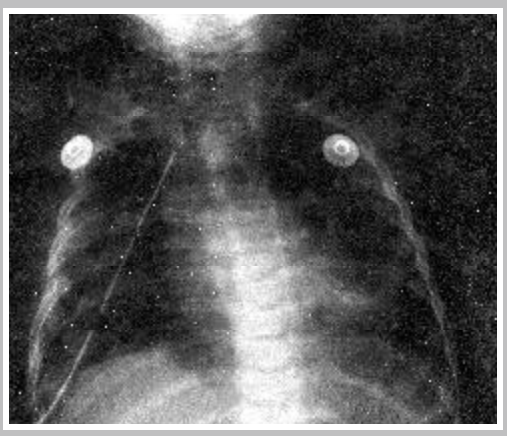


Fig:6-7 (X-Ray Chest PA View)

excision of the lung cyst. The right lung blossomed after the removal of cyst. Histo-pathology report confirmed the pre-operative diagnosis of a lung cyst.

A 6 months old, male child referred from general medical ward with the complaints of shortness of breath for the last 1 month, cough which was productive for the last 1month, and fever for the last 25 days. Though the general course of the disease was down hill but he was stable when examined by the thoracic team. His chest X-Ray showed hyperlucency with absent bronchovascular markings in the right hemi thorax. CT scan chest confirmed the diagnosis of a cyst in the right hemi thorax.

Right Thoracotomy was done. There was a large cyst which was compressing the whole lung completely. The cyst was excised totally. It's communication with the upper lobe was suture ligated.The post operative recovery was smooth and uneventful. Histo Pathology report: simple lung cyst.

Of these 10 patients, 6 had bronchogenic pulmonary cysts, 2 had congenital cystic adenomatoid malformation (CCAM), and 2 were congenital lobar emphysemas. Surgery was done in all the 10 patients. Excision of the cyst was done in 6 patients. Lobectomy was done in 4 patients with congenital cystic adenomatoid malformation and congenital lobar emphysema. There was no incidence of post operative mortality or morbidity. Emergency thoracotomy had to be performed in 1 case of congenital lobar emphysema and in 1 case of broncho pulmonary lung cyst.

To summarize the cases we experienced, all the patients under 1 year of age were symptomatic, and the major cause of respiratory distress was mediastinal shift due to the cystic lesions. In contrast the patients over 1 year old presented with recurrent chest infections predominantly. Chest x-ray and CT scan chest were the major investigations performed.8 patients at short-term follow-up ranging from 2 months to 10 months after surgery are doing well with no subsequent limitation of physical activities due to lung resection/ thoracic surgery.

DISCUSSION

During the development of the embryo, separation of the trachea and esophagus occurs, and migration of the early lung bud takes place. The lung tissue shows further differentiation thereafter into airway epithelium and alveolar cells. In this developmental stage, numerous abnormalities can take place[1] .Bronchopulmonary foregut malformations are combinations of these forms of disordered lung growth such as dysplasia, hypoplasia,or hyperplasia involving one or more structural components of the lung.

Bronchogenic pulmonary cysts were the most common lesion in our series, e.g. 6 cases, and are examples of anomalies occurring early in lung development. They may be lined with squamous epithelium rather than respiratory epithelium, presumably due to recurrent infection[6].

Congenital cystic adenomatoid malformation probably results from a cessation of bronchial maturation and concomitant overgrowth of mesenchymal elements, which produce the adenomatoid appearance of the anomaly in the early stage of development[2, 10]. Histologically,cartilage is absent, reflecting the bronchial maldevelopment.There often exists a small bronchial communication which leads to infection and over inflation of the cystic disease. In our study there were 2

cases of CCAM. Lobectomy was done in both cases with excellent results.

Congenital lobar emphysema is a marked pulmonary hyperinflation state that resembles all of the clinical features of obstructive emphysema. One possible etiology is that cartilaginous defect weakens the bronchus, which may collapse on expiration followed by air-block hyperinflation. It is necessary to evaluate associated anomalies e.g. congenital heart disease[3,11]. In this study, 2 cases of congenital lobar emphysema had lobectomy done in each patient.

Pulmonary sequestration probably occurs very early in the embryonic development before the pulmonary and aortic circulation becomes separate. Sequestration could represent as a separate mini-lung bud, which keeps a systemic artery and drains into either venous system depending on its intimacy with normal lung bud to which it is adjacent[8].

Reviewing the patients it was seen that in patients under 1 year of age, cystic lesions were diagnosed by respiratory distress; and sign of infection was a core clinical feature in patients over 1 year of age[14]. Since newborns and infants have a weak chest wall and mediastinum stucture,the intrapulmonary expanding mass may cause tracheal compression resulting in respiratory distress. This observation in this study is supplemented by different papers e.g. Takeda S[14]. All the 4 patients under 1 year of age had severe respiratory distress at presentation.

Regarding the diagnostic modality, chest X-Ray was the most cost effective and time efficient method of diagnosing surgical lesion. Nowadays congenital cystic lung lesions in children can be readily diagnosed with current imaging modalities, such as CT, MRI, Ultrasongraphy and digital subtraction angiography.

Infants and children tolerate lobectomy extremely well with compensatory lung growth[12,15]. There was no incidence of post operative mortality or morbidity. In most of the cases with cystic lung disease, prognosis is good after lung resection.

Early recognition of these relatively rare congenital cystic lung lesions should lead to the immediate, proper surgical treatment. This approach should salvage almost all affected infants, who would otherwise face a dismal future.

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