CONGENITAL CYSTIC DISEASE OF THE LUNG IN INFANTS AND CHILDREN (EXPERIENCE WITH 57 CASES)

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SUMMARY:

A retrospective analysis of 57 consecutive cases with congenital cystic disease of the lung admitted to King Faisal Specialist Hospital and Research Center and King Khalid University Hospital, Riyadh between 1985-1995 is presented.

There were 37 CLE, 7 CAM, 8 BC, and 5 PS. There were 39 males and 18 females with ages ranging from 1 day to 5 years. All patients were symptomatic except three. Respiratory distress, repeated chest infections, and cystic changes noted in chest x-ray were the commonest presentation. Five of eight patients with BC presented with symptoms related to pressure effect of the cyst on the surrounding structures, these included bronchiectasis in two patients, bronchopleural fistula in one, pulmonary artery stenosis and bronchomalacia in one and airway obstruction mimicking bronchial asthma in one.

Seven patients (12.2%) were treated conservatively, the remaining underwent surgery. Surgery included excision of the bronchogenic cyst and lobectomy for CLE, CAM, and intralobar sequestration.

The post operative course in most cases was uneventful. There were no deaths in this series, and the majority of patients have a satisfactory outcome with follow-up ranging from 1-72 months (mean 24 months).

It appears that lobectomy for symptomatic CLE, CAM, and intralobar sequestration and excision for bronchogenic cyst offer the best treatment modality and is well tolerated by paediatric patients. Careful search for associated anomalies is important to obtain better outcome.

KEY WORDS: Lung cyst, congenital lobar emphysema, cystic adenomatoid malformation, bronchogenic cyst, pulmonary sequestration.

INTRODUCTION:

Congenital cystic disease of the lung in infants and children is rare. It consists of congenital lobar emphysema (CLE), cystic adenomatoid malformation (CAM), Bronchogenic cyst, and pulmonary sequestration (PS).[2,13] These lesions show close relationship in terms of embryology and clinical presentation.[5] Some cause lifethreatening respiratory distress at birth while others appear late in life as an unexpected findings on an accidental roentgenogram.

The majority of patients are symptomatic and present with progressive respiratory distress or repeated chest infections with cystic changes noted in chest x-ray. Surgical treatment has been shown to be safe and effective in treating these lesions. We report our clinical experience in managing 57 patients with these lesions as seen in King Faisal Specialist Hospital and Research Centre and King Khalid University Hospital, Riyadh, over 10 year period.

MATERIALS AND METHODS:

Over a 10 year period (1985 - 1995), all paediatric patients with congenital cystic disease of the lung managed at King Faisal Specialist Hospital and Research Centre and King Khalid University Hospital, Riyadh, were reviewed. The medical records of 57 patients were studied in respect of age, sex, clinical presentation, location of anomalies, imaging studies, associated anomalies, treatment and outcome. Criteria for inclusion include histologically confirmed diagnosis or highly suspected on clinical and histological findings.

RESULTS:

Between 1985 - 1995, 57 patients were seen at King Faisal Specialist Hospital and Research Centre and King Khalid University Hospital with congenital cystic disease of the lung. Their age range was 1 day to 5 years with a mean of age of 24 months. There were 39 males and 18 female patients. There were 37 CLE, 7 CAM, 8 BC, and 5 PS. Review of their clinical presentation showed all patients were symptomatic except three. Two of the asymptomatic patients presented with congenital lobar emphysema and one with bronchogenic cyst. Respiratory distress was the most common symptom being present in 46 patients (80.7%) (Table 1). On further evaluation, 5 patients with bronchogenic cyst presented with complication related to the pressure effect of the cyst on the surrounding structures. These included bronchiectasis in two patients, bronchopleural fistula in one, bronchial asthma in one, pulmonary artery stenosis and bronchomalacia in the final patient.

In patients with congenital lobar emphysema the left upper lobe was involved in 23 (63.1%), the right middle lobe in 10 (27%) and right upper lobe in 8 (21.6%). Four patients had bilobar lesions. In patients with CAM 3 involved the right lower lobe, 3 the left lower lobe and one the right upper lobe. Bronchogenic cysts were mediastinally located in 5 patients and intralobar in 3 patients. Pulmonary sequestrations were intralobar in all patients, 3 in the right lower lobe, one in the left lower lobe and one involving the entire left lung.

Twenty-four patients (42%) with congenital cystic disease of the lung presented with associated anomalies or abnormalities (Table 2). Congenital heart disease was the commonest associated anomaly and was found mainly in patients with CLE and PS.

Radiological studies included chest x-ray in all patients and computed tomography in 30. Ventilation perfusion scan was done in 29, angiogram in 5, esophagogram in 4, and magnetic resonance imaging in 2 patients. Chest x-ray film was sufficient to confirm the diagnosis in more than half of the patients (Fig. 1) . CT scan, VQ scan and MRI were used to distinguish between different types of congenital cystic lesions, when the diagnosis was uncertain and to accurately locate the cystic lesion. CT scan missed the diagnosis of mediastinal bronchogenic cyst in one patient which was detected by MRI (Fig. 2A, 2B). Treatment consisted of medical (conservative) management in 7 patients (12.2%), arterial embolization of sequestered lobe in 2 patients and surgical intervention in the remaining 48 patients (84.2%).

Conservative treatment was offered to 7 patients with CLE. Two were asymptomatic and the other 5 were experiencing mild symptoms.

Surgery consisted of excision of mediastinal bronchogenic cysts and total lobectomy in CAM, CLE, intralobar sequestration, and intralobar bronchogenic cysts.

Fifty-three (53) surgical procedures were done through 48 thoracotomies. The procedures included 44 lobectomies, 2 pneumonectomies, (one for the whole left lung sequestration and the other for right lower lobe cystic adenoid malformation and hypoplastic remaining lobes), 5 excisions of bronchogenic cysts, one wedge excision of left lower lobe

and one resection of oesophageal and gastric duplication. Two patients with CLE who were

conservatively treated missed follow-up, the remaining five patients were followed up for 3 years and showed clinical and radiological improvement. The remaining patients were followed up between 1-72 months and all of them alive and well. Two patients with PS treated with arterial embolization had a good result.

Two patients developed preoperative complications due to chest tube insertion at referring hospital in assumption of a tension pneumothorax as the primary diagnosis. These were pneumothorax in one and bronchopleural fistula in another.

Seven patients (12.2%) developed post-operative complications. These included pneumothorax in 4, pleural effusion in 1, surgical emphysema in 1 and pneumonia in one.

Of those who were treated surgically 38 (79%) were asymptomatic, 6 patients (12.5%) had minimal respiratory symptoms. Three patients had recurrent pneumonia and one patient had a moderate bronchial asthma. All resected specimens were submitted for histological studies. Resected lobes from the patients with CAM revealed Type II in 6 and Type I in one patient. The histological diagnosis of BC confirmed in 6 patients and was difficult to make in 2 patients because of infection.

DISCUSSION:

Congenital cystic disease of the lung is a group of lesions that share similar clinical and embryological features. The exact incidence of these lesions is not known. In recent years, a number of series have reported these anomalies in groups or as individual lesion.[2, 5,11,13,14,16]

Normal lung development in the embryo depends on the normal interaction between the developing bronchial tree and the peribronchial mesenchyme. Abnormalities in the inductive interaction appear to be responsible for these lesions.[13] The sex incidence of these lesions have been shown to be more common in males.[2,17] In our series, the incidence showed a ratio of 2M:1F. The majority of our patients presented below the age of one year especially patients with CLE and CAM. Patients with PS and BC tend to present later. These observations were also made by others.[1,2,13]

On reviewing the clinical presentations of these lesions, respiratory distress, cyanosis and repeated chest infections were the commonest presenting symptoms. This observation was also found by many others.[2,5,17] The majority of our patients are symptomatic. Only three were asymptomatic and discovered incidentally during reviewing chest radiograph done for other purposes.

Twenty-four patients (42%) had associated anomalies. The presence of associated anomalies can make diagnosis difficult and modify the surgical approach.

In this series, the left upper lobe was the commonest site for CLE followed by right middle lobe, in CAM the lower lobes were equally affected by CAM. A bronchogenic cyst was centrally located in the majority of patients. These findings were more or less the same as previously reported.[2,14,16] All our patients with PS were intralobar variety. Savic et al[9] analyzed 400 intralobar and 133 extralobar sequestration reported between 1862 - 1975 and found most of intralobar sequestration in the lower lobes. Radiological studies such as CT scan, VQ scan, MRI, oesophagogram and angiogram had been used in uncertain cases or to evaluate the associated anomalies. Sonography and/or CT scan with contrast has been shown to have high diagnostic efficacy in patients with congenital cystic disease of the lung.[2,6,7,15,17]

The indication for surgery in most of these lesions is significant respiratory distress. An asymptomatic bronchogenic cystic or extralobar PS needs excision because of risk of infection later in life and the rare association of BC and CAM with malignancies such as rhabdomyosarcoma.[8,12,18] Nine of our patients were treated non-surgically. Seven of these were asymptomatic or mildly symptomatic CLE, and 2 patients with intralobar PS underwent arterial embolization because they were having major cardiac anomalies and unfit for general anaesthesia.

Out of 7 patients with CLE treated conservatively, 5 were followed up for 3 years. All of them showed clinical and radiological improvement and require no surgery. Conservative treatment for asymptomatic CLE has been recommended some cases.[3]

Because the benefit from lobectomy in asymptomatic CLE has not been shown and pulmonary function tests results in patients with asymptomatic CLE and symptomatic CLE patients who underwent lobectomy are similar, it is reasonable to treat these patients conservatively.[4,10]

The surgical treatment includes resection of bronchogenic cysts and lobectomy for CLE, CAM, and intralobar PS, and intralobar bronchogenic cysts. This surgical approach has been shown to be safe and effective.[2,13,17] Segmental resection for CAM and intralobar PS carries high morbidity rate with prolonged air leak and recurrent infection.[17] In another study, limited pulmonary resection for CAM was1 feasible and can preserve lung ti1ssue.[11] Our experience showed that complete lobectomy is safe and effective. There were no deaths in this series. Seven patients (14.5%) had postoperative complications which were relatively minor and responded to conservative treatment.

On follow-up ranging from 1-72 months the majority of patients (79%) were asymptomatic and has no physical limitation. Six patients (12.5%) had minimal respiratory symptoms.

In conclusion, congenital cystic disease of the lung is rare anomaly and the majority of cases are symptomatic and can be diagnosed radiologically. Careful search for associated anomalies is important to help obtain better outcome. Lobectomy for symptomatic CLE, CAM and intralobar PS and excision of bronchogenic cyst offer the best modality of treatment and is well tolerated by paediatric patients. Although our experience in the conservative treatment of asymptomatic and mildy symptomatic CLE patients is limited, this approach is recommended whenever patient can be closely followed up.

TABLE I

CLINICAL PRESENTATION

SYMPTOMS/SIGNS	NO. OF PATIENTS
Respiratory Distress	46 (80.7%)
Cyanosis	20 (35%)
Repeated Chest Infection	19 (33.3%)
Cough	10 (17.5%)
Failure to Thrive	7 (12.2%)
Asymptomatic	3 (5.2%)
Empyema and Bronchopleural Fistula	1 (1.7%)

TABLE 2

ASSOCIATED ANOMALIES/ABNORMALITIES

ANOMALIES	NO. OF PATIENTS	TYPE OF CYSTIC LUNG DISEASE
Congenital heart disease	10 (17.5%)	CLE, PS
Pectus Excavatum	2	CLE
Lt. diaphragmatic hernia	1	CLE
Rt. upper lobe tracheal bronchus	1	CLE
Hiatus hernia/gastroesophageal reflux	1	CLE
Prematurity	3	CLE
Esophageal and gastric duplication	1	CAM
Bronchiectatic changes	2	BC
Pulmonary artery stenosis, and bronchomalacia	1	BC
Hypoplastic lung	2	PS
T O T A L	24 (42%)	

REFERENCES:

1.Becker MR, Schindera F, Maier WA: 1987, Congenital cystic adenomatoid malformation of the lung. Prog Pediatr Surg. 21;112-117.

2.Coran AG, Drongowski R: Congenital cystic disease of the tracheo-bronchial tree in infants and children - Experience with 44 consecutive cases. Arch Surg. 129; 1994:521-527.

3.Eigen H, Lemen RJ, Waring WW: Congenital lobar emphysema - Long-term evaluation of surgically and conservatively treated children. Am. Rev. Respir. Dis. 113; 1976:823-831.

4.Frenckner B, Freyschuss U: 1982, Pulmonary function after lobectomy for congenital lobar emphysema and congenital cystic adenomatoid malformation. A follow-up study. Scand J Thorac Cardiovasc Surg. 16;293-298.

5.Haller JA, Golladay ES, Pickard LR et al: Surgical management of lung bud anomalies: Lobar emphysema, bronchogenic cyst, cystic adenomatoid malformation, and intralobar pulmonary sequestration. Ann Thorac Surg. 28; 1979:33-43.

6.Hartenberg MA, Brewer WH: Cystic adenomatoid malformation of the lung: Identification by sonography. AJR. 140; 1983:693-694.

7.Kaude JV, Laurin S: Ultrasonographic demonstration of systemic artery feeding extrapulmonary sequestration. Pediatr Radiol. 14; 1984:226-227.

8.Krous HF, Sexauer CL: Embryonal rhabdomyosarcoma arising within a congenital bronchogenic cyst in a child. J Pediatr Surg. 16; 1981:506-508.

9.Savic B. Birtel FJ, Tholin W, et al: Lung sequestration: Report of seven cases and review of 540 published cases. Thorax 34; 1979:96-101.

10.McBride JT, Whol MEB, Strieder DJ, et al: Lung growth and airway function after lobectomy in infancy for congenital lobar emphysema. J Clin Invest. 66; 1980:962-970.

11.Mentzer SJ, Filler RM, Phillips J: Limited pulmonary resections for congenital cystic adenomatoid malformation of the lung. J Pediatr Surg. 27; 1992:1410-1413.

12.Murphy JJ, Blair GK, Fraser GC, et al: Rhabdomyosarcoma of the lung arising within congenital pulmonary cysts: Report of three cases. J Pediatr Surg. 27; 1992:3164-3167.

13.Nuchtern JG, Harberg FJ: Congenital lung cysts. Seminar in Pediatric Surgery 3; 1994;233-243.

14.Ribet M, Pruvot FR, Dubos JP et al: Congenital cystic adenomatoid malformation of the lung. Eur J Cardiothorac. 4; 1990:403-406.

15.Shackelford GD, Siegel MJ: CT appearance of cystic adenomatoid malformations. J Comput assist Tomogr. 13; 1989:612-616.

16.Suen HC, Mathisen DJ, Grillo HC, et al: Surgical management and radiological characteristics of bronchogenic cysts. Ann Thorac Surg. 55; 1993:476-481.

17.Wesley JR, Heidelberger KP, Dipietro MA, Cho KJ, Coran AG: Diagnosis and management of congenital cystic disease of the lung in children. J Pediatr Surg. 21; 1986:202-207.

18. Ueda K, Gruppo R, Unger F, et al: Rhabdomyosarcoma of the lung arising in congenital cystic adenomatoid malformation. Cancer. 40; 1977:383-388.

FIGURES/LEGENDS:

FIGURE 1: Chest radiograph showing bronchogenic cyst of the right lower lobe with air-fluid level indicating communication to bronchial tree.

FIGURE 2A:T1 weighted coronal image of the chest showing a subcarinal lesion of low to mid signal intensity suggesting a cystic nature of this lesion.

Also it demonstrates the presence of bronchiectatic changes with consolidation at the left base.

FIGURE 2B:This is a T2 weighted axial images of the chest at the level of the previously mentioned lesion which demonstrate high signal intensity confirming the cystic nature of this lesion.

Also we can see the bronchiectatic changes at the left base in the more caudal image.

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Re:Manuscript: Congenital cystic disease of the lung in infants and children (Experience with 57 cases)

Dear Dr. Spiro,

Enclosed with this letter you will find the above mentioned manuscript for consideration for publication in your journal and the references for our paper.

This paper is not under consideration by any other journal at the same time and it has not been accepted for publication elsewhere.

I hope you will find the manuscript suitable for publication.

Yours sincerely,

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REFERENCES:

1.Nuchtern JG, Harberg FJ: Congenital lung cysts. Seminar in Pediatric Surgery. 1994;233-243.

2.Coran AG, Drongowski R: Congenital cystic disease of the tracheo-bronchial tree in infants and children - Experience with 44 consecutive cases. Arch Surg. 1994;129:521-527.

3.Haller JA, Golladay ES, Pickard LR et al: Surgical management of lung bud anomalies: Lobar emphysema, bronchogenic cyst, cystic adenomatoid malformation, and intralobar pulmonary sequestration. Ann Thorac Surg. 1979;28:33-43.

4.Ribet M, Pruvot FR, Dubos JP et al: Congenital cystic adenomatoid malformation of the lung. Eur J Cardiothorac. 1990;4:403-406.

5.Mentzer SJ, Filler RM, Phillips J: Limited pulmonary resections for congenital cystic adenomatoid malformation of the lung. J Pediatr Surg. 1992;27:1410-1413.

6.Suen HC, Mathisen DJ, Grillo HC, et al: Surgical management and radiological characteristics of bronchogenic cysts. Ann Thorac Surg. 1993;55:476-481.

7.Wesley JR, Heidelberger KP, Dipietro MA, Cho KJ, Coran AG: Diagnosis and management of congenital cystic disease of the lung in children. J Pediatr Surg. 1986;21:202-207.

8.Becker MR, Schindera F, Maier WA: Congenital cystic adenomatoid malformation of the lung. Prog Pediatr Surg. 1987;21:112-117.

9.Luck SR, Reynolds M, Raffensperger JG: Congenital bronchopulmonary malformations. Curr Prob Surg. 1986;23:245-314.

10.Shackelford GD, Siegel MJ: CT appearance of cystic adenomatoid malformations. J Comput assist Tomogr. 1989;13:612-616.

11.Hartenberg MA, Brewer WH: Cystic adenomatoid malformation of the lung: Identification by sonography. AJR. 1983;140:693-694.

12.Kaude JV, Laurin S: Ultrasonographic demonstration of systemic artery feeding extrapulmonary sequestration. Pediatr Radiol. 1984;14:226-227.

13.Krous HF, Sexauer CL: Embryonal rhabdomyosarcoma arising within a congenital bronchogenic cyst in a child. J Pediatr Surg. 1981;16:506-508.

14. Ueda K, Gruppo R, Unger F, et al: Rhabdomyosarcoma of the lung arising in congenital cystic adenomatoid malformation. Cancer. 1977;40:383-388.

15.Murphy JJ, Blair GK, Fraser GC, et al: Rhabdomyosarcoma of the lung arising within congenital pulmonary cysts: Report of three cases. J Pediatr Surg. 1992;27:3164-3167.

16.Eigen H, Lemen RJ, Waring WW: Congenital lobar emphysema - Long-term evaluation of surgically and conservatively treated children. Am. Rev. Respir. Dis. Am. Rev. Respir. Dis. 1976;113:823-831.

17.Frenckner B, Freyschuss U: Pulmonary function after lobectomy for congenital lobar emphysema and congenital cystic adenomatoid malformation. A follow-up study. Scand J Thorac Cardiovasc Surg. 1982;16:293-298.
18.McBride JT, Whol MEB, Strieder DJ, et al: Lung growth and airway function after lobectomy in infancy for congenital lobar emphysema. J Clin Invest. 1980;66:962-970.