Comparing Late-onset and Neonatally-diagnosed Congenital Cystic Adenomatoid Malformation of the Lung

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Background: Most congenital cystic adenomatoid malformations (CCAM) are found in utero or during the immediate neonatal period. Some malformations regress in utero, while others persist and remain unnoticed until later in childhood. The optimal clinical management of patients with CCAM is controversial. The aim of this study is to suggest a safe strategy for treatment of CCAM based upon the age of the patient at diagnosis, by analyzing the clinical features of CCAM and considering the possibility of regression in early infancy.

Method: This is an observational retrospective study of 19 patients with CCAM. The clinical features, histopathological classification, status of lesion regression, diagnostic method, treatment, and outcome were collected. Patient data were analyzed highlighting age at disease presentation.

Results: Five out of the seven neonates with neonatally-diagnosed CCAM presented with respiratory distress. Eight of the twelve patients in the late-onset group had respiratory tract infections. Regression of the lesion during the early postnatal period was documented in 4 neonatally-diagnosed CCAMs, while none of the patients in the late-onset group showed signs of radiographic changes after a mean follow up of 4 years. Skeletal malformation was the most common associated anomaly in our series.

Conclusion: Conservative treatment is suggested for neonatally-diagnosed CCAM because of possible postnatal remission. Surgery may be required in older patients because of possible recurrent infections, infrequent mass regression, radiation exposure, and inconveniencies during follow-up visits. A thorough survey of possible associated skeletal anomalies in patients with CCAM is also recommended as early correction can improve life quality.

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Key words: congenital cystic adenomatoid malformation, congenital pulmonary airway malformation, cystic lung lesion, late-onset, neonates

Congenital cystic adenomatoid malformation (CCAM) is an uncommon anomaly characterized by multicystic lesions due to proliferation of the respiratory bronchioles. This adenomatoid appearance of the lung tissue was first described as a distinct disease entity by Chi'in and Tang in 1949.[1] In 1977, Stocker et al. classified CCAM into 3 histopathological groups.[2] An expanded classification of 5 types was further proposed in 2002. A new term, congenital pulmonary malformation (CPAM),

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was suggested, and is currently used by several authors.[3-7] The etiology remains unknown, but a disturbed interaction between mesodermal and ectodermal components of the lung during embryonic development has been suggested.

CCAM represents about 1/4 of all congenital malformations of the lung.[8] About eighty percent of patients have respiratory distress in the neonatal period, The other patients have no symptoms or have onset of respiratory infections after the first years of life.[9] The clinical manifestations can vary from hydrops or fetal death to complete absence of symptoms, with CCAM discovered only later in life. We performed a retrospective study of 19 patients with CCAM, emphasizing age at disease diagnosis. We present here our clinical experience during a 20-year period.

METHODS

We conducted a computer search for CCAM from 1985 to 2005 at Chang Gung Children's Hospital, a 500-bed teaching hospital. Twenty-two patients with a diagnosis of CCAM were identified, and their patient and clinical characteristics, imaging studies of the chest, and histological features were documented. The diagnosis of CCAM was confirmed by histopathological results in 12 patients. The tissue sections were reviewed by an experienced pediatric pathologist (CH), and reclassified according to Stocker's new classification system. For those patients without tissue sampling, imaging studies were also carefully reviewed and only those with typical cystic changes were included. Three patients were excluded from our study, two because of equivocal imaging results, and one because of a pathological diagnosis of pulmonary sequestration. Data collected included age at assessment, presenting symptoms, histological types, associate malformations, and outcome.

In the new histological classification by Stocker, lesions are classified as follows: in type 0, the lung mass contains cysts measuring < 0.5 cm, lined by tall ciliated, columnar, pseudostratified cells with the presence of cartilage; type 1 lesions have large cysts measuring 4-10 cm, lined by tall ciliated, columnar, pseudostratified cells with the occasional presence of cartilage; type 2 lesions are comprised of many cysts < 2.5 cm that are lined by ciliated, cuboidal or columnar cells; type 3 lesions have cysts < 1.5 cm, lined by ciliated cuboidal cells; and type 4 lesions have cysts measuring around 2-4 cm, lined by flattened alveolar cells. No cartilage is present in types 2, 3, and 4.[3]

For patients diagnosed solely by computed tomography or magnetic resonance, images showed multicystic air-filled lung lesions of various sizes with visualization of thin internal septa. Three types of CCAM can be identified through imaging studies. Type 1 malformation consists of variable-sized cysts, with at least one cyst more than 2 cm in diameter. Type 2 is comprised of multiple small thin-walled 1-10 mm cysts. Type 3 CCAM appears as a solid mass on visual inspection, but actually contains multiple tiny cysts smaller than 2 mm.[9,10] The systemic arterial supply was carefully scrutinized to exclude pulmonary sequestration. In differentiating from pneumatocele, patients were excluded if they had active infection with rapid subsequent radiographical disappearance of the lesion. Three neonates had prenatal ultrasonographic confirmation. Lesions were considered to have spontaneous regression if pre-existing lesions disappeared radiographically and the patients remained asymptomatic.

To compare the clinical spectrum of CCAM in older children and adults with neonates, we divided patients into two groups, the neonatally-diagnosed group, in which patients had disease found prenatally or during the immediate neonatal period, and the late-onset group, in which the malformation went unnoticed and persisted until later in life.

RESULTS

Patient data are shown in Table 1. There were 11 females and 8 males. The age at diagnosis ranged from gestational age 20 weeks to 21 years old. Seven patients were diagnosed prenatally or during the immediate neonatal period. Five out of these seven neonates (71%) had respiratory distress shortly after birth. In contrast, most patients in the late-onset group were either asymptomatic or presented with respiratory infections (Table 2). Two patients had multiple lobular involvement. There were no sites of predilection for tumor location in either group. Type 1 CCAM predominated in the late-onset group. The incidence of skeletal anomaly was similar between groups, but major cardiopulmonary malformation

Table 1. Clinical Profiles of Patients with Congenital Cystic Adenomatoid Malformation

Case No.	Age at diagnosis	Clinical manifestation	Age at surgery	Initial radiographic finding (CT or MR)	Outcome
1	3 d/o	Respiratory distress	1y 6 m/o	Soft tissue mass with cystic like lesion	Well by 3 y/o*
2	3 d/o	Respiratory distress	8 d/o	Probable congenital diaphragmatic defect	Well by 3 m/o
3	5 d/o	Respiratory distress	21 d/o	Multiple cystic lesion suggest CCAM	Well by 5 y/o
4	GA 20 wk	Coincidental	none	Multiple radioluscent cysts suggest CCAM	Regressed but persistent lesion by 3 y/o
5	GA 20 wk	Respiratory distress	none	Multiple cystic lesions suggest CCAM	No lesion by 1y10 m/o
6	GA 26 wk	Coincidental	none	Multiple cystic lesions (< 10 mm) suggest CCAM	No lesion by 9 y/o
7	1 d/o	Respiratory distress	none	Air-containing structure suggest CCAM	No lesion by 7 m/o†
8	1 y/o	Coincidental	1y/o	Multiple cysts suggest CCAM	Well by 1y2 m/o
9	1 y 6 m/o	Failure to thrive	2 y/o	Large lobulated cyst suggest lobar emphysema	Well by 3y6 m/o
10	2 y/o	Coincidental	2 y/o	Large lobulated cyst suggest CCAM or pneumatocele	Well by 4 y/o
11	5 y/o	Recurrent lung infection	5 y/o	Multiple cystic lesions suggest CCAM	Well by 8 y/o
12	11 y/o	Recurrent lung infection	15 y/o	Multiple cystic lesions suggest CCAM	Well by 16 y/o‡
13	21 y/o	Recurrent lung infection	21 y/o & 25 y/o	CXR: Left upper lung cyst	Residual air space at LUL§
14	5 y/o	Lung infection (once)	none	Multiple cystic lesions suggest CCAM	Lesion persisted by 6 y/o
15	5 y/o	Lung infection (once)	none	Cystic lesions suggest CCAM	Lesion persisted by 8 y/o
16	4 y/o	Lung infection (once)	4y3 m/o	Large cystic lesion suggest CCAM or pneumatocele	Well by 7 y/o
17	16 y/o	Lung infection (once)	none	Muliple cystic lesions suggest CCAM	Lesion persisted by 18 y/o
18	16 y/o	Coincidental	16 y/o	Multiple cystic lung lesions suggest CCAM	Well by 17 y/o
19	4 y/o	Lung infection (once)	4 y/o	Multiple cystic lung lesions suggest CCAM	Well by 5y10 m/o

Abbreviations: LUL: left upper lobe; CT: Computed tomography; MR: Magnetic resonance; *: In Patient 1, the lesion in the right lower lobe resolved spontaneously a year after diagnosis, and therefore, only a left lower lobectomy was performed. No lesion was found intraoperatively; †: Patient 7 was discharged after 5 days without signs of infection or respiratory symptoms. Minimal O2 support was given during hospitalization; ‡: Patient 12 had lesions followed radiographically for 4 years, Surgery was eventually done because of persistence of the lesion; §: Patient 13 had a chest radiograph showing a persistent oval air collection in the left upper lobe after video- assisted resection. Because of frequent subsequent respiratory tract infections, a lobectomy was performed 4 years later with pathology revealing a bronchogenic cyst.

predominated in the neonatally-diagnosed group. Regression of the malformation was detected in 4 neonates during the first few years of life. An example is shown with patient 7 in Fig. 1. No patients from the late-onset group showed radiographic changes in lesion size during a mean follow-up of 4 years. Fig. 2 shows the image of patient 12 having persistence of the mass lesion.

Five of the 12 patients who underwent surgical excision had type 1 (42%), 6 had type 2 (50%), among which 2 patients were diagonsed by radiographic findings; and 1 had radiographic diagnosis of type 3 CCAM (8%).

The parents of 7 patients opted for conservative therapy. Radiographic regression of the mass lesion was documented in patients 1, 5, 6, 7 of the neonatally- diagnosed group. No respiratory compromise or airway infection occurred during the follow-up period. In contrast, serial radiological survey did not show signs of lesion change in patients 14, 15, and 17.

Scoliosis and funnel chest were the most commonly associated anomalies. The number of skeletal

Table 2. Comparison between Neonatal-assessed and Late-onset Congenital Cystic Adenomatoid Malformation of the Lung

	Neonatally-assessed (N = 7) N (%)	Late -onset (N = 12) N (%)	*p*
Stocker's classification*			
Type 0	0	0	
Type 1	0	5 (42)	0.204
Type 2	2 (29)	4 (33)	1.000
Type 3	1 (14)	0	0.250
Type 4	0	0	
Sex			
Male	4 (57)	4 (33)	0.377
Clinical manifestation			
Shortness of breath	5 (71)	0	0.018*
Lung infection	0	8 (67)	0.013*
Free of symptoms	2 (29)	4 (33)	1.000
Location of lesion			
RUL	0	1 (8)	1.000
RLL	3 (38)	4 (31)	1.000
RML	1 (12)	1 (8)	1.000
LUL	0	3 (23)	0.263
LLL	4 (50)	4 (31)	0.377
Number of lesions†			
Single	6 (86)	11 (92)	1.000
Multiple	1 (14)	1 (8)	
Associate anomaly			
Cardiac	3 (43)	0	0.036
Renal	0	1 (8)	1.000
Lung	1 (14)	0	0.368
Skeletal (scoliosis, funnel chest, genu varum)	3 (43)	3 (23)	0.617
Operation			
Lobectomy	3 (43)	9 (69)	0.326
Video-assisted resection	0	1 (8)	
None	4 (57)	3 (23)	
Regression of mass lesion	4 (50)	0	0.009*

Fisher's exact test

Abbreviations: RUL: right upper lobe; RLL: right lower lobe; RML: right middle lobe; LUL: left upper lobe; LLL: left lower lobe; *: Results only available for 3 neonates and 9 patients of the late-onset group; †: percentage based on 8 lesions in the neonatal group and 13 lesions in the late-onset group.

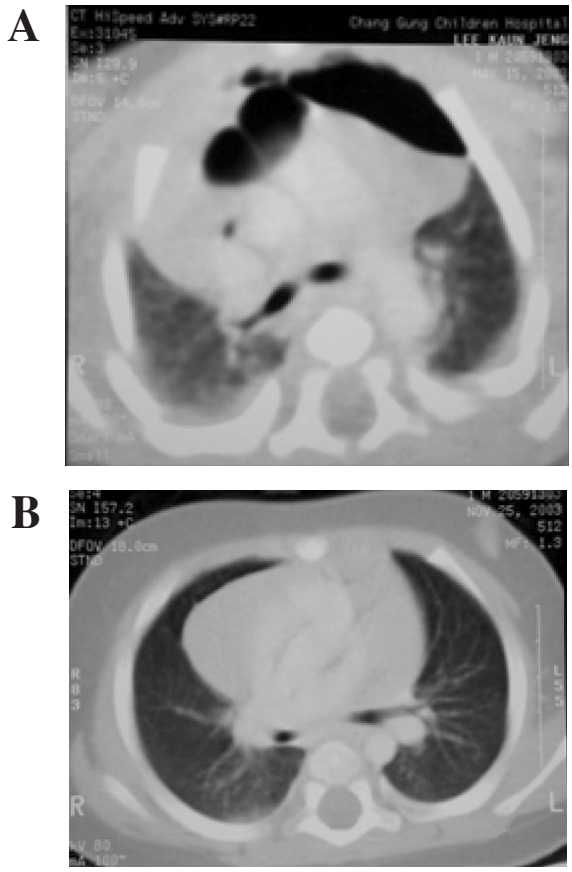

Fig. 1 (A) Computed tomography (CT) of patient 7 at 3 days old shows abnormal air-containing structure with septum in the anterior aspect of the thymus. (B) Disappearance of pre-existing lung lesion 7 months later.

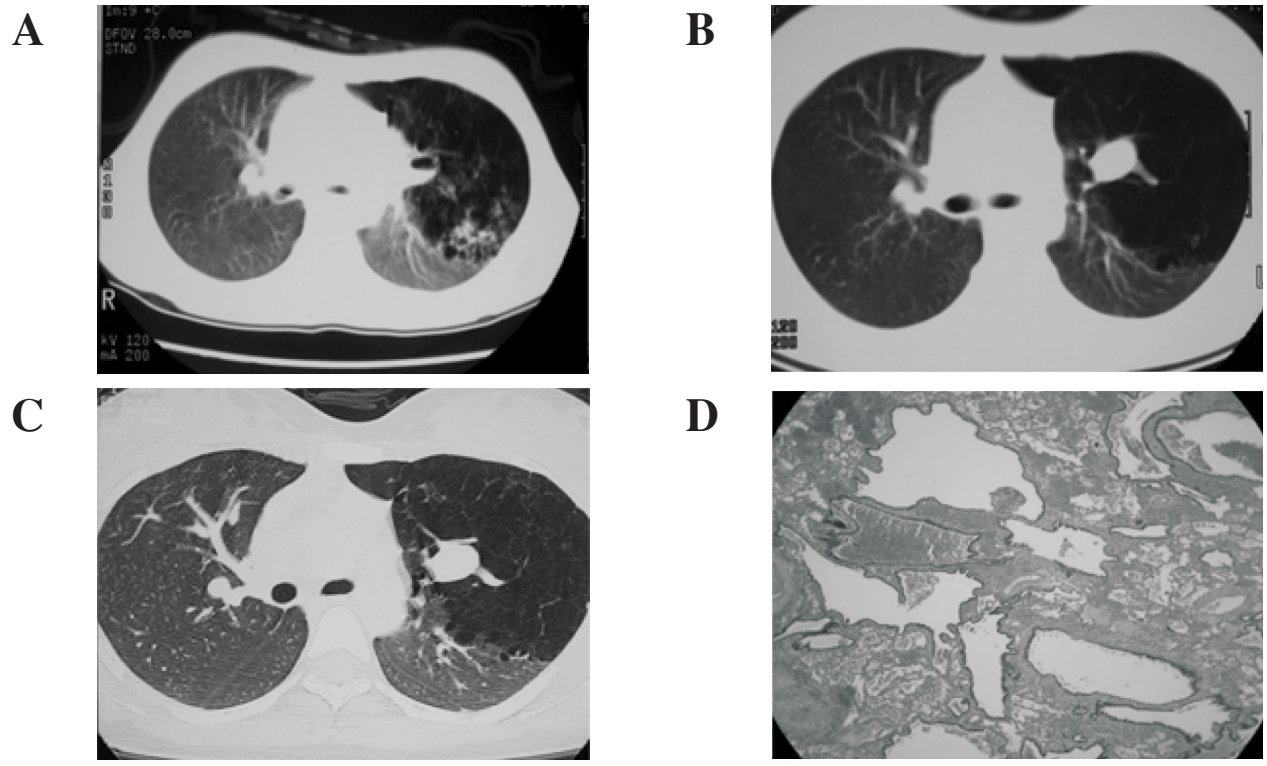

Fig. 2 (A) CT of patient 12 at 11 years old shows emphysematous left upper lobe with irregular honeycombing and air-fluid level in the parenchyma. (B) CT shows a persistent lesion one year later. (C) High resolution CT at 15 years old shows no obvious changes compared to previous images. (D) Histopathology shows CCAM type 2 with multiple- sized cysts lined by respiratory epithelium.

anomalies was similar between groups, but major cardiopulmonary malformations appeared only in the neonatally-diagnosed group. Five of ten patients with associated anomalies had type 2 CCAM.

Most patients who underwent surgery tolerated daily activity well after lobectomy of the lung despite their age. No signs of recurrence were detected.

DISCUSSION

A new classification system was suggested for patients with CCAM to indicate the site of origin with progression down the airway. The exact typing of the malformation might be helpful in excluding associated anomalies and highlighting the risk of developing subsequent malignancies. Type 1 CCAM predominated in the late-onset group in our series, and these patients were more likely to remain asymptomatic. In contrast, type 2 CCAM was often associated with other major anomalies. Neither type 0 nor Type 4 CCAM was found in our study. Since type 0 is incompatible with life, it was reasonable that we identified no cases. Our case number may have been too small to find any cases of type 4 CCAM, but it is commonly misdiagnosed as lobar emphysema or grade 1 pleuropulmonary blastoma.[4,11]

The most common clinical manifestation in neonates with CCAM is respiratory distress. On the contrary, older children present more frequently with recurrent lung infections. The majority of CCAM cases reported in the literature are unilobular. Only a few cases have multilobular involvement and bilateral disease is even rarer. Two patients in this series had multilobular lesions, of which 1 patient had bilateral lung involvement. Spontaneous resolution of the right lower lobe lesion in patient 1 probably explained his symptom-free periods through infancy, and thus a bilobectomy was not needed.

In utero regression or even complete resolution

of CCAM has been documented by several authors,[12-17] but continuous postnatal regression has rarely been reported, and the possibility has even been questioned by some. We were able to follow 7 patients whose parents refused immediate surgery. We found spontaneous postnatal regression of the malformation in 4 patients. This finding was similar to that of Butterworth et al. who reviewed 56 cases of antenatally diagnosed CCAM, and identified 2 patients with postnatal regression of the lesions at 5 months and 37 months of age.[18] Theoretically, malformations don't simply disappear, but the possibility of continuing regression beyond the postnatal period can not be excluded, since lung development is not completed until 3 years of age.[19] The number of patients in this study may have been too small to make this conclusion, however, this is an important issue that deserves further investigation. For this reason, we suggest that for asymptomatic neonates with CCAM, postponing surgery for 3 years might be beneficial because of the possibility of shrinking of the lesion, thus, facilitating surgery. In addition, the probability of complete remission cannot be overlooked, as occurred in two of our cases. Several authors have advocated an observational approach before surgical intervention if patients remain symptom free.[20-23] However, for neonates with CCAM who have respiratory distress, the physician needs to remember that other disorders such as transient tachypnea of the newborn, respiratory distress syndrome, congenital pneumonia, and congenital diaphragmatic hernia should be excluded as the cause. We believe that for those patients who have prompt resolution of respiratory distress shortly after birth without any associated anomalies, observation alone may spare the patient unnecessary surgery. However, we suggest immediate surgical treatment for patients in the late-onset group, because spontaneous regression of the lesion is less likely to occur at older ages. Repeated infections, further radiographic exposure during follow-up, and frequent medical visits are important concerns that may affect the decisions on more aggressive intervention. Furthermore, the possibility of malignant change of the malformation should also be taken into consideration.[4,7,24]

Simple cyst excision may leave residual anomalous tissue resulting in recurrence. One of our patients who had video-assisted resection of the cyst presented with recurrent infections and subsequent development of a bronchogenic cyst, eventually requiring a lobectomy of the lung. Similar problems have been reported when a lobectomy is not performed.[22,25] Thus, lobectomy remains the standard surgical approach for patients with CCAM. The operation was well tolerated in all of our patients with minimal complications.

Congenital heart disease and pulmonary malformations are the most commonly reported associated anomalies in patients with CCAM.[26-28] In our series, skeletal anomaly (pectus excavatum, genu varum, and scoliosis) was a prominent feature (6 out of 19 patients). Similar findings have been reported but not emphasized.[26,29,30] Although funnel chest and scoliosis might be secondary to respiratory compromise or major thoracic procedures, there were a number of patients who have had preexisting concomitant skeletal abnormalities without any respiratory symptoms or surgery. Consistent with other reports, we suggest a thorough investigation for coexisting cardiac, renal, or skeletal malformations in patients with type 2 CCAM, which has the highest incidence of major anomalies.

Conclusion

Clinical manifestations of CCAM vary depending on the age at presentation. We recommend conservative treatment for neonates with CCAM because of the possibility of postnatal regression of the lesion. For CCAM found at later ages, prompt surgical intervention should remain the gold standard, because of infrequent mass remission, possible repeated infections, and radiation exposure during frequent hospitalizations. We also suggest a thorough survey for possible associated skeletal anomalies in patients with CCAM, as early correction can improve their life quality.

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晚期及新生兒時期先天性肺泡腺瘤畸形之比較

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背 景： 近年來由於產前超音波的普遍及進步，先天性肺泡腺瘤畸形常可於產前時即被診斷，但仍有些案例遲至孩童甚至成年才被確診。藉由此回溯性的研究，我們將探討先天性肺泡腺瘤畸形的處置是否依表現年齡的不同而有差異。

方 法： 我們藉由回顧病歷的方式，收集了 19 位罹患先天性肺泡腺瘤畸形的病人，並分析及記錄其臨床表現、病理組織結果、診斷方式、病程及預後等。

結 果： 在產前或新生兒時期即被診斷出有先天性肺泡腺瘤畸形的病人數有 7 位，其中 4 位有合併呼吸窘迫的症狀；另 12 位病人是在晚期才被診斷出來，其中 8 位病人的臨床表現爲呼吸道感染。經過平均 4 年的追蹤，4 位新生兒時期確診的病童，其病灶隨著年紀皆有消失或變小的跡象。而在晚期確診的案例中，其皆仍維持原樣。骨骼發育異常是這些案例中最常見的相關異常。

結 論： 我們建議在新生兒時期所診斷出來的先天性肺泡腺瘤畸形可持保守觀察療法，因爲這個時期的畸形腫塊會有較高自行緩解的可能。相反地，在孩童或成人時期才被診斷出來的先天性肺泡腺瘤畸形，應及早予以切除才能預防反覆感染，或因常照 X 光所導致輻射線的暴露，或須長期至醫院追蹤所導致的不便。另外，我們亦建議要檢查其相關的骨骼異常，才能早期治療，改善生活品質。

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關鍵詞： 先天性肺泡腺瘤畸形，先天性肺及呼吸道畸形，囊狀肺疾病，晚期，新生兒

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