Case Report

Late Manifestation of Congenital Cystic Adenomatoid Malformation with Lung Abscess: A Case Report

Sanitra Sirithangkul MD*,
Supichaya Chuengchitraks MD*, Dusit Staworn MD*,
Chalida Laohapand MD*, Teerachat Silarat MD**

** Department of Pediatrics, Phramongkutklao Hospital, Bangkok, Thailand*
*** Department of Surgery, Phramongkutklao Hospital, Bangkok, Thailand*

Congenital cystic adenomatoid malformation (CCAM) is a rare congenital cystic abnormality of the lung. Most of cases are usually diagnosed and managed in the newborn period, even though some are asymptomatic and present in childhood or adult.

The authors report a 7-year-old girl who presented with chronic cough, hemoptysis and clubbing of fingers. Physical examination revealed decreased breath sound and dullness on percussion at right upper chest. A chest radiograph showed a large thin- walled cyst with air fluid and a small thin-walled cyst occupied the whole right upper lobe. Computed tomography (CT) of the chest showed two large thin-walled cavities with air fluid level in the right upper lobe with few small cavities nearby. She was given antibiotics and cardiovascular and thoracic surgeon was consulted. The patient underwent right upper lobectomy. Microscopic examination was compatible with CCAM type 1. The post operative course was uneventful and the recovery was complete. She continued to be healthy at a follow-up visit about 8 months postoperatively but chest radiograph showed soft tissue density at right upper lung field. Chest CT findings were compatible with recurrent CCAM. A follow-up chest radiograph at 13 months postoperatively showed significant reduction in size of the lesion.

Keywords: *Congenital cystic adenomatoid malformation, Lung abscess, Surgical resection*

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Congenital cystic adenomatoid malformation (CCAM) is a rare developmental abnormality of the lung. The incidence is 1: 25,000-35,000 pregnancies[1]. It was first described as a distinct pathological entity by Chin and Tang in 1949[2]. It has no association with race, sex, maternal age or familial predisposition[3]. It is characterized by an adenomatous overgrowth of terminal bronchiolar-like structures with subsequent suppression of alveolar growth, resulting from an embryologic insult before the 49th gestation day[4].

Sophisticated diagnostic imaging procedures allow the detection of most congenital malformations. Prenatal diagnosis is possible by ultrasonography after the 16th week of gestation[3,4]. CCAM is the most common type of fetal thoracic mass diagnosed by prenatal ultrasonography[5]. Although ultrasonographic prenatal diagnosis is becoming increasingly sophisticated, diagnostic errors are possible and some patients suffering from a CCAM of reduced size are not diagnosed. These patients remain asymptomatic until the occurrence of respiratory infections; chest radiograph then give rise to the diagnostic suspicion of CCAM. In some cases the CCAM can disappear spontaneously (especially during fetal life), but repeated episodes of pneumonias frequently occur.

The authors report a 7-year-old girl with a late manifestation of CCAM.

Correspondence to: Sirithangkul S, Department of Pediatrics, Phramongkutklao Hospital, Bangkok 10400, Thailand.
Phone: 0-2354-7600
E-mail: sanitras@hotmail.com

Case Report

A 7-year-old girl was referred to our pediatric service because of lung abscess. She had a five-year history of chronic cough and digital clubbing. She had no history of pneumonia and did not have any chest radiographs. This time she presented with hemoptysis for 4 days. On examination, she was afebrile, had no cyanosis, no tachypnea or dyspnea. Marked digital clubbing was present. Her oxygen saturation was 98% on room air. Her weight was 18.6 kg (10th percentile) and her height was 118 cm (25th-50th percentile). She had

normal chest contour and her trachea was in midline. Auscultation of the chest revealed decreased breath sound and diminished tactile fremitus with dullness on percussion at right upper lung. The remainder of her physical examination was normal.

A chest radiograph showed a large thin-walled cyst with air fluid level and a small thin-walled cyst occupied the whole right upper lobe (Fig. 1). Complete blood count: Hb 12.6 g/dl, Hct 36%, WBC 11,500/mm^3, PMN 53%, L 35%, M 10%, E 2%, platelets 393,000/mm^3. Computed tomography (CT) of the chest showed two large thin-walled cavities with air fluid level in the right upper lobe, about 5 cm and 7 cm in diameter, with a few small cavities nearby (Fig. 2).

She was given intravenous antibiotics and cardiovascular and thoracic surgeon was consulted. In order to prevent spilling of pus to other lobes during operation, the cyst was aspirated under fluoroscopy before incision. The patient underwent right upper lobectomy. Macroscopic examination revealed a large cavity sized 6 x 8 cm with anchovy thick fluid inside, occupied most of the right upper lobe (Fig. 3). The pus culture was positive for beta-lactamase producing Haemophilus influenzae. Appropriate antibiotics were given for 4 weeks. Microscopic examination was compatible of with CCAM type 1. The post-operative course was totally uneventful and recovery was complete.

Eight months after surgical resection, she was well and the digital clubbing resolved but chest radiographs showed soft tissue density at right upper lung field (Fig. 4). Chest CT showed a large fluid-filled cavity with a small thin cyst with air fluid level nearby at upper part of right lung (Fig. 5). Both findings were compatible with recurrent CCAM. She continued to be healthy and a follow-up chest radiograph at 13 months postoperatively showed significant reduction in size of soft tissue density at right upper lung field (Fig. 6).

Discussion

CCAM is one of congenital lung malformations, characterized by an overgrowth of terminal bronchiolar-like structures resulting in the formation of cysts of variable sizes within the lung. The vast majority of CCAM described to data are unilobar; the cases of multilobar diseases are much less frequent and the cases of bilateral diseases rarer still[6].

Patients with CCAM can present as neonates with severe, progressive respiratory distress due to cyst expansion or as older children or adults with recurrent pulmonary infections localized to one lobe or hemoptysis. CCAM is most commonly found in the neonatal period and up to 90% of diagnoses are made in the first two years of life[7]. Small lesions cause few symptoms. A predominately solid lung mass is usually found in stillborn and premature infants and is associated with hydrops, ascites and polyhydramnios. On occasion, CCAM may remain asymptomatic and be discovered incidentally on a chest radiograph later in life[3,4].

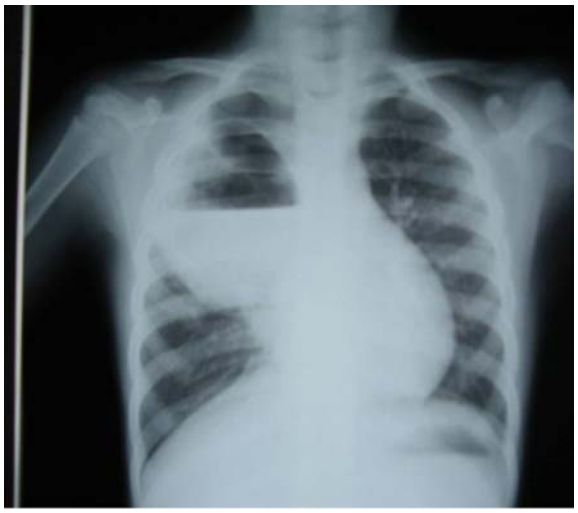

Fig. 1 Chest radiograph showed a large thin-walled cyst with air fluid level and a small thin-walled cyst occupied the whole right upper lobe.

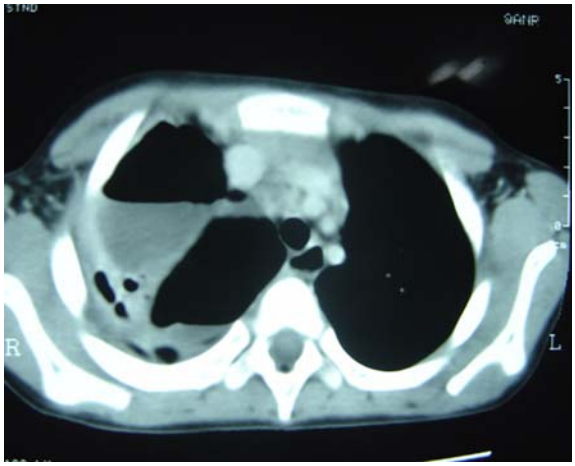

Fig. 2 Chest CT showed two large thin-walled cavities with air fluid level in the right upper lobe, about 5 cm and 7 cm in diameter, with a few small cavities nearby.

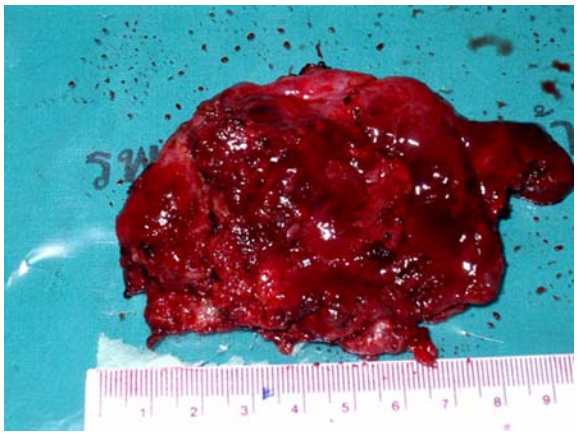

Fig. 3 Macroscopic examination revealed a large cavity sized 6 x 8 cm with anchovy thick fluid inside.

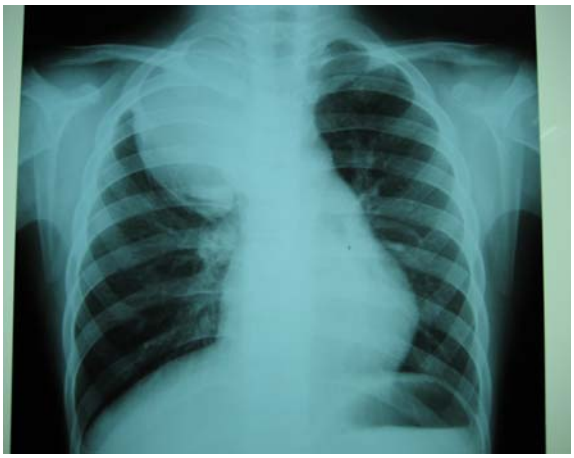

Fig. 4 Chest radiograph at 8 months postoperatively showed soft tissue density at right upper lung field.

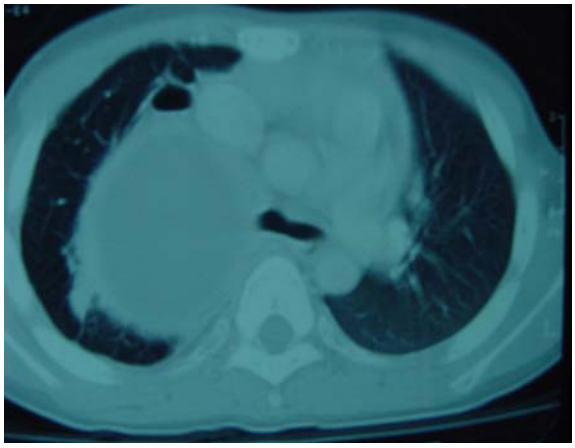

Fig. 5 Chest CT at 8 months postoperatively showed a large fluid-filled cavity with a small thin-walled cyst with air fluid level nearby at upper part of right lung.

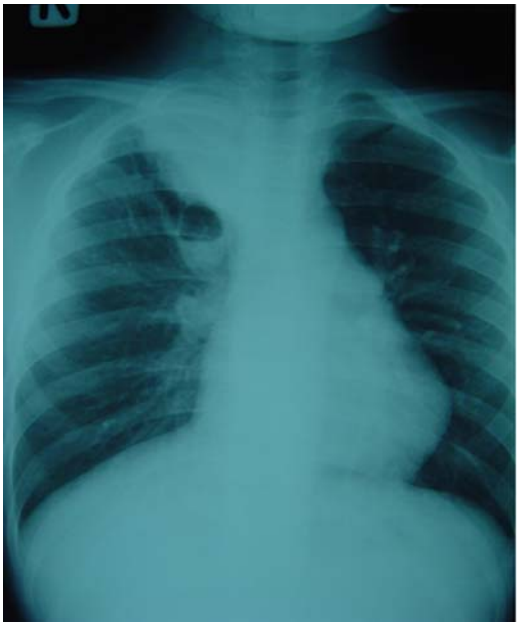

Fig. 6 A follow-up chest radiograph at 13 months postoperatively showed significant reduction in size of soft tissue density at right upper lung field.

Diagnosis of CCAM is suggested from the clinical features and chest radiograph findings. Chest radiographs may show single or multiple cysts or a solid mass. Bronchogenic cyst, pulmonary sequestration, congenital lobar emphysema and congenital diaphragmatic hernia in newborn period should be considered in the differential diagnosis, for which chest CT is often required. Chest CT is useful in confirming the presence of the lesion, determining the extent of the lesion and defining associated abnormalities. CT of CCAM patient shows a mass with a variable number of solid and cystic components[8]. CCAM communicates with bronchial tree at birth and therefore typically contains air soon after birth. The imaging appearance is determined by the size and number of cysts. Lesions are typically solitary with no lobar predilection[9].

Definitive diagnosis of CCAM is made on histologic examination. The classification most widely used is the original classification by Stocker of CCAM type I-III[10]. In 2002, the CCAM classification was redefined and renamed to congenital pulmonary airway malformations (CPAM), type 0-4, and was based on

the hypothesis that each type corresponds to a perturbation along the airway from its proximal (bronchus) to distal segment (bronchiolo-alveolar sac)[11]. The most frequent pathological forms are type 1 and type 2, with the prevalence of 60-70% and 15-20% respectively.

Management of CCAM is straightforward for symptomatic lesions, however, has been controversial for prenatally diagnosed asymptomatic lesions. All babies diagnosed antenatally with CCAM require postnatal imaging with CT irrespective of signs of antenatal resolution[12]. Surgical resection is the standard of care for symptomatic patients with CCAM[13]. The most commonly used technique is lobectomy because segmentectomy or localized resection may result in incomplete resection and/or persistent pneumothorax[14]. Surgical outcomes are excellent[15]. Elective surgery is associated with better outcome than emergency surgery[16]. Asymptomatic lesions have the potential to become infected; undergo malignant transformation (for instance bronchiolo-alveolar carcinoma, rhabdomyosarcoma, pleuro-pulmonary blastoma); or cause hemoptysis or pneumothorax. For all these reasons surgical resection is favored by many centers as the treatment of choice for asymptomatic patients. However, the appropriate management of asymptomatic lesions has remained controversial and management decisions are often influenced by physician preferences reflecting the past experiences of the individual or team[17].

The presented case had a long history of chronic cough and digital clubbing. Her chest radiographs showed two cysts with air fluid level in a large one. She was placed on appropriate antibiotics for lung abscess. The authors suspected of CCAM then chest CT was performed. All clinical and radiological findings indicated CCAM. She underwent right upper lobectomy with totally uneventful postoperative course. During follow-up she was well but chest radiograph and chest CT were compatible with recurrent CCAM. She continued to be healthy and thriving and a recent chest radiograph showed significant reduction in size of soft tissue density at right upper lung field. The authors have decided to observe and follow-up both clinical and radiological findings with a plan to perform surgical resection if she has any symptoms or the lesions are progressively worse.

Conclusion

Late manifestation of CCAM is an infrequent illness which requires a high level of suspicion. It usually presents with recurrent pulmonary infections localized to one lobe or presents as lung abscess. The diagnostic method of choice is computed tomography of the chest. The most common pathological form is type 1. The recommended treatment is surgical resection, in principle a lobectomy, once the diagnosis has been established. There should be a long clinical and radiological follow-up.

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อาการแสดงในระยะหลังของโรค Congenital cystic adenomatoid malformation ที่มาด้วยฝีในปอด: รายงานผู้ป่วย

สนิตรา ศิริธางกุล, สุพิชญา จึงจิตรักษ์, ดุสิต สถาวร, ชลิดา เลาหพันธ์, ธีรฉัตร ศิลารัตน์

โรค Congenital cystic adenomatoid malformation (CCAM) เป็นโรคที่เกิดจากความผิดปกติแต่กำเนิดของปอดซึ่งพบได้น้อย ผู้ป่วยส่วนใหญ่มักได้รับการวินิจฉัยและรักษาตั้งแต่ยังเป็นทารกแรกเกิด ยังมีผู้ป่วยบางรายที่ไม่มีอาการและแสดงอาการตอนโตหรือเป็นผู้ใหญ่ รายงานผู้ป่วยเด็กหญิงไทย อายุ 7 ปี มาด้วยอาการไอเรื้อรัง ไอเป็นเลือดและนิ้วปุ้ม ตรวจพบเสียงปอดเบาลงและเคาะทึบที่หน้าอกข้างขวาบน ภาพรังสีทรวงอกพบถุงน้ำขนาดใหญ่ซึ่งมีสารน้ำอยู่ภายในและถุงน้ำขนาดเล็กอยู่เกือบเต็มปอดกลีบขวาบน เอกซเรย์คอมพิวเตอร์ของปอดพบถุงน้ำขนาดใหญ่ 2 ถุง ที่มีสารน้ำอยู่ภายในและถุงน้ำเล็ก ๆ อยู่ด้านข้าง ผู้ป่วยได้รับการรักษาด้วยยาปฏิชีวนะและปรึกษาศัลยแพทย์ทรวงอก ผู้ป่วยได้รับการผ่าตัดเอาปอดกลีบขวาบนออก ผลการตรวจทางพยาธิวิทยาเข้าได้กับ CCAM ชนิดที่ 1 ไม่พบมีภาวะแทรกซ้อนหลังผ่าตัดและผู้ป่วยฟื้นตัวดี ผู้ป่วยสบายดีแต่ภาพรังสีทรวงอกหลังผ่าตัดเป็นเวลา 8 เดือนพบเงาขาวผิดปกติที่บริเวณของปอดขวาบน เอกซเรย์คอมพิวเตอร์ของปอดพบว่าเข้าได้กับการกลับเป็นซ้ำของ CCAM การติดตามภาพรังสีทรวงอกหลังผ่าตัดเป็นเวลา 13 เดือน พบว่ามีการลดลงอย่างชัดเจนของเงาขาวผิดปกติที่บริเวณของปอดขวาบน