Congenital Pulmonary Airway Malformation in an Adult: A Case Report and Review of the Literature

Cheng-Chieh Hsu, Yu-Chen Wang*, Wei-Chih Chen

Congenital pulmonary airway malformation (CPAM), previously known as congenital cystic adenomatoid malformation, is a rare anomaly of the lower respiratory tract characterized by cystic adenomatous overgrowth of the terminal bronchioles and airspaces [1-2]. Most cases are found in infants and neonates with respiratory distress. CPAM can be a cause of pulmonary hypoplasia, severe nonimmune fetal hydrops, and fetal death [3]. On rare occasions, CPAM can present in adults, with recurrent pulmonary infections, pneumothorax, hemoptysis, or dyspnea [4]. CPAM has been found to be associated with malignancies. We report the case of a 33-year-old female who was diagnosed with CPAM with recurrent hemoptysis.

Case Report

This 33-year-old woman without systemic disease (except breast augmentation) had had intermittent hemoptysis for many years. Five...
months before this surgical intervention, she was admitted to another hospital for recurrent hemoptysis. The chest plain film (Figure 1A) showed a round opacity with an air-fluid level at the left lower lung. She was given an intravenous antibiotic due to a suspicion of pneumonia. After discharge (2 months before this surgical intervention), she visited our chest clinic for a second opinion, and the chest plain film (Figure 1B) showed a cystic lesion at the left lower lung. She was given medication for symptomatic control, and a chest plain film showed no interval change in the left lower lung lesion (Figure 1C). Since multiple cystic lesions were found by chest X-ray in the left lower lobe (LLL), a computed tomography (CT) with aortography of the chest was arranged for evaluation. The CT scan showed a 6-cm multiloculated cystic lesion in the left lower lobe (LLL) (Figure 2), with a dedicated pulmonary artery and pulmonary vein (Figure 2). There was no definite, engorged abnormal systemic artery from the aorta supplying the cystic portion of the lung. A radiologic diagnosis of CPAM was then made.

Fig. 1. Serial chest plain films: (A) 5 months before this surgical intervention, presenting with hemoptysis; (B) 2 months before this surgical intervention and in a stable condition; (C) pre-operative evaluation.

Fig. 2. Chest computed tomography with angiogram: (left) a multiloculated cystic lesion at the left lower lobe; (right) a dedicated pulmonary artery and pulmonary vein supplying the hypolucent area.
During the period of clinical visits, hemoptysis occurred again, accompanied with chest pain. She came to our emergency department for help. Increased alveolar infiltrates at the LLL were noted on the plain film. The lab data revealed neither leukocytosis nor elevated C reactive protein (CRP). Normal coagulation function was also noted. She was therefore admitted to the floor for empiric antibiotics with levofloxacin and intravenous transexamic acid treatment. The symptoms were relieved gradually. Because of the repeated hemoptysis and pulmonary infection, surgical intervention was recommended after a multidisciplinary team discussion.

Once the condition had stabilized, she underwent pre-operative assessment. The pulmonary exam revealed a normal ventilatory function. The forced expiratory volume in 1 second (FEV\(_1\)) was 2.88 L with 101% of predictive value, the forced vital capacity (FVC) was 3.3 L with 100% of predictive value, the ratio of FEV\(_1\) and FVC was 87%, the total lung capacity (TLC) was 5.31 L with 116% of predictive value, and the provocation test was negative. A video-assisted thoracic surgery (VATS) with lobectomy of the LLL was then performed.

Gross examination of the pathology specimen showed that the lobe of the lung was 13×8×5.9 cm in size. Multiple cystic spaces, measuring about 6×5×3 cm in size, were noted within the lung tissue. The surrounding paren-

---

Fig. 3. Gross photograph showing a lobe of the lung (LLL), measured as 1×8×5.9 cm. Multiple cystic spaces (arrow), measuring about 6×5×3 cm, are noted within the lung tissue.

Fig. 4. Microscopic pathological examination: (left) under a low power view, several dilated bronchioles, irregularly sized and shaped cystic spaces within the lung parenchyma; (right) under a higher power view, pseudostratified, ciliated columnar or cuboidal epithelial cells, suggestive of bronchiole-like structures without accompanying arteries.
chyma showed emphysematous change (Figure 3). Microscopic examination found several dilated bronchioles, and irregularly sized and shaped cystic spaces within the lung parenchyma. These cystic spaces were lined with pseudostratified, ciliated columnar or cuboidal epithelial cells, suggestive of bronchiole-like structures without accompanying arteries. The picture was compatible with type 1 CPAM (Figure 4). The post-operative course was smooth and she was discharged with regular clinical follow-up.

**Discussion**

CPAM is a rare anomaly of the lower respiratory tract characterized by cystic adenomatous overgrowth of the terminal bronchioles and airspaces [1]. It was first described by Chin and Tang in 1949 as a “congenital adenomatoid malformation.” [5] Stocker [6] classified CPAM into 3 groups based on the size of the cysts and their cellular characteristics. Type 1 CPAM is composed of single or multiple large cysts, more than 2 cm in diameter, and lined with a ciliated pseudostratified columnar epithelium. Type 2 CPAM consists of multiple small cysts, less than 1 cm in diameter, lined with ciliated cuboidal to columnar epithelium. Type 3 CPAM is a large, bulky non-cystic lesion producing a mediastinal shift. Two more subtypes were added recently. Type 0 CPAM, also known as acinar dysplasia, is a proximal bronchial anomaly, and type 4 CPAM is a peripheral lung anomaly [7].

CPAM is an uncommon disease; the reported incidence ranges from 1:10,000 to 1:35,000 newborns [8]. In all, 80% to 85% of cases are recognized in the first 2 years of life, and an adult presentation is extremely uncommon [9]. Adult CPAM occurs sporadically [4]. The exact mechanism of the formation of CPAM is still unknown. Some possibilities have been raised, including chromosomal abnormalities like trisomy 18, hereditary renal dysplasia [10], mutation-disrupting TTF-1 (thyroid transcription factor-1) [4], and high levels of HoxB5 (homeobox protein) [11].

CPAMs are usually identified during routine obstetric care due to the increased modern use of ultrasound [12]. CPAM usually presents with acute respiratory distress in neonates and infants [13]. In adults, the most common clinical presentation includes recurrent pulmonary infection, pneumothorax, hemoptysis, fever, and dyspnea. Asymptomatic radiographic abnormalities are the second most common presentation of CPAM in adults [4]. Almost 44% of CPAM patients are found to have lower lobe lung lesions, primarily unilateral [11,14]. CPAM has also been found to be related to malignancies such as adenocarcinoma and pleuropulmonary blastoma [15-16]. It is estimated that approximately 1% of CPAMs, particularly types 1 and 4, transform into malignancy, although the exact incidence is unknown [17]. Mucous cells in type 1 CPAM have a tendency to undergo malignant changes [16].

Chest roentgenogram findings are nonspecific, ranging from a non-visible lesion to consolidation or a mass-like lesion, based on the extension of the lesion. Consolidations are indistinguishable from a lobar pneumonia. An intrapulmonary water density mass with well-defined margins may contain lucent cystic areas or an air-fluid level resembling a lung abscess or pneumatocele [18]. The recommended diagnostic test for CPAM in postnatal life is a CT scan of the chest [19]. In CT radiography, a large cyst or a cluster of multiple air-filled/
fluid-filled cysts, even resembling a solid mass, may be seen [20]. The CPAM may enlarge to produce a mass effect at the adjacent lung and mediastinum, complicated with atelectasis and/or mediastinal shift [9]. Angiography or magnetic resonance angiography can provide images of systemic vascular supply, which may differentiate CPAM from other cystic lung lesions, including bronchopulmonary sequestration [21]. CT scan findings vary depending upon the type of CPAM and the clinical presentation. Types 1, 2, and 4 CPAM are characterized by air-filled cysts. Radiography does not reliably distinguish among them. However, types 1 and 4 tend to appear as a single lesion, with a single or a few larger cysts (up to 10 cm in diameter) [19]. In type 2, the numerous small cysts appear more homogenous (up to 2.5 cm in diameter). Type 4 is often complicated with pneumothorax, or bilateral/multifocal cysts [22]. Type 3 CPAM often appears as a solid, homogeneous mass, usually with a mediastinal shift and/or hypoplasia of the ipsilateral lung [19]. Radiologic differential diagnosis, including pulmonary sequestration, bronchogenic cyst, cystic bronchiectasis, diaphragmatic hernia, and infected tumors, must be made [21].

As CPAM presenting in adulthood is rare, no specific treatment guidelines exist. Surgery is the best option for symptomatic patients [23], to prevent recurrent infection and to eliminate concerns regarding malignancy. For those with asymptomatic CPAM, elective resection is safe and alleviates the risk of symptom development, which may result in a more complicated surgery and recovery [24]. Lobectomy is the most commonly reported procedure because of concern about an incomplete removal of the pulmonary malformation [25] and the development of complications like air leak associated with lung-sparing surgeries [26]. Lung-sparing surgery, in the form of segmentectomy, may be suitable for well-circumscribed lesions, although postoperative recurrences have been described [27]. The resected specimen should always be carefully examined to detect occult malignancy [28]. In extensive CPAM, non-anatomical resection may be the only option, in order to avoid the risk and difficulty of a pneumonectomy [29].

The survival rate at 6 months of age seems to have improved [24]. Prognoses among adult CPAM patients vary, depending upon the pathological features, infection, and the potential for malignant transformation [18].

Conclusion

CPAM is a rare pulmonary disease, that presents with recurrent infections. The causes of the repeated infections need to be investigated. Review of the history and serial images could help differentiate CPAM from other congenital cystic diseases. CT scans are the initial diagnostic choice. Considering the possibility of further infectious episodes and even malignant transformation, surgical resection is suggested.

References

2. 鍾福財、郭志熙、張博瑞等：成人先天囊狀類腺畸形－病例報告及文獻回顧。胸腔醫學；21卷3期 (2006/06/01)：291-7。
4. McDonough RJ, Niven AS, Havenstrite KA. Congenital
Congenital Pulmonary Airway Malformation in an Adult: A Case Report and Review of the Literature


成人先天性肺氣道畸型－病例報告及文獻回顧

許政傑 王予辰* 陳威志

先天性肺氣道畸型（原名為先天囊狀類腺畸型）為罕見之先天肺部囊泡性疾病，主要發生於下呼吸道。大部份的病例為新生兒或嬰兒，乃經過產前超音波檢查或呼吸窘迫的症狀而被發現。然而少部份病人直到成人時期才被診斷，且不一定有症狀。部份的病例可能有反覆性的肺部感染，甚至是衍生出良性腫瘤。因此，建議完全切除病灶，如肺葉切除手術，尤其針對已有症狀的小孩或成人。我們在此報告一例成人之先天性肺氣道畸型，以反覆咳血為表現。經胸腔內視鏡輔助肺葉切除手術後，目前追蹤情況穩定。最後我們針對此病之流行病學、組織病理分類、臨床表現、放射線學變化與成人案例的處置等，回顧並整理過去的文獻以提出此報告。( 胸腔醫學 2017; 32: 80-86)

關鍵詞：先天性肺氣道畸型，先天囊狀類腺畸型，咳血，胸腔內視鏡輔助手術