

# Congenital Cystic Adenomatoid Malformation of Lung Mimicking Lung Cancer: A Case Report

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**Abstract:** We report a case of a 66-year-old woman with congenital cystic adenomatoid malformation (CCAM) that presented as a thin-walled cyst on radiological imaging and mimicked lung cancer. The right pulmonary wedge resection was performed through thoracoscopic surgery. Pathologic results proved CCAM which though, uncommon may be misdiagnosed in adults.

**Keywords:** *Congenital cystic adenomatoid malformation; Lung cancer; Misdiagnosis*

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## 0 Introduction

Congenital cystic adenomatoid malformation (CCAM), although rare, is a more common lung lesion in newborns than in adults. We report a case of a 66-year-old woman with a cystic lesion in the right lower lobe which was highly suspicious of lung cancer primarily. The final pathologic diagnosis was CCAM.

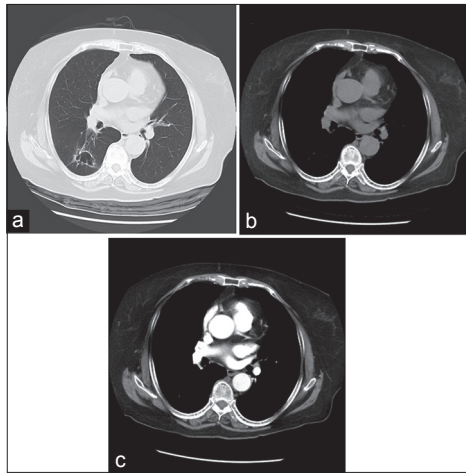
## 1 Case report

A 66-year old Chinese woman, born in Beijing, was admitted with complaints of cough, expectoration, and shortness of breath since for 3 months. She denied fever, night sweats, loss of weight, chest pain, or hemoptysis. Her history was unremarkable with no history of

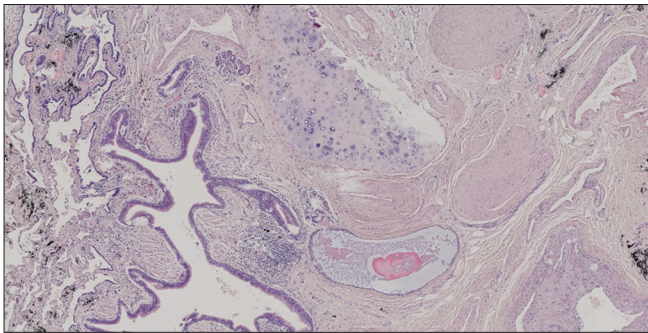
tuberculosis or smoking. Physical examination was normal. Clinically, electrocardiography and routine laboratory tests were normal. Pulmonary function tests showed forced expiratory volume in one second (FEV<sub>1</sub>)/FVC: 71.1% and FEV<sub>1</sub>%pred: 83.2%. Contrast-enhanced computerized tomography (CT) of chest showed 35 mm × 22 mm irregular cystic lesion in the right lower lobe, wall thickness of 0.5–3.5 mm, and smooth inner margin, septum was seen within the cyst. The lesion also showed mild distortion of surrounding lung tissue with no evidence of abnormal arterial blood supply [Figure 1]. Bronchoalveolar lavage fluid cytology showed cells with a heteromorphic nucleus. Right lower lobe thoracoscopic wedge resection was done. The size of the lung tissue removed was 10 cm × 2.5 cm × 1.5 cm. Histopathology [Figure 2] showed cystic tumor of 4.5 cm × 2 cm × 1.5 cm, composed of multiple varying sized cysts lined by pseudostratified ciliated columnar epithelium with few areas of necrosis. All these features were consistent with the diagnosis of CCAM. On follow-up 1 year later, the patient is asymptomatic.

## 2 Discussion

CCAM of the lung is a rare condition, first described by Ch'in and Tang in 1949<sup>[1]</sup>. It is characterized by the presence of an abnormal mass of pulmonary tissue that appears immature, malformed and may show varying degrees of cystic change. It is characterized by an adenomatous overgrowth of the terminal bronchioles with consequent suppression of alveolar growth, the



**Figure 1:** Chest radiography showed a 35 mm × 22 mm cystic lesion in the right lower lung. Lung window setting (a), soft tissue window setting (b), and contrast-enhanced chest computed tomography (c).



**Figure 2:** Histopathology (H and E, ×40), lesion composed of varying sized cysts, lined by pseudostratified ciliated columnar epithelium. The wall of the cysts contains smooth muscle, blood vessels, and cartilage. Relatively normal alveoli may be seen interspersed between the cyst.

formation of intercommunicating cysts lined by cuboidal and ciliated pseudostratified respiratory epithelium. The pathogenesis is not clear, but segmental bronchial atresia has been attributed as the primary pathogenetic defect that results in the development of a CCAM distal to the defect<sup>[2]</sup>. The incidence of CCAM of the lung was found to be 0.0029–0.004% in newborns<sup>[3,4]</sup>. However, on rare occasions, CCAM can be found in adults. The number of female patients compared with male patients was higher<sup>[4]</sup>. In adults, presenting clinical symptoms may include cough, expectoration, chest tightness, and dyspnea. CCAM is often found when they cause recurrent pulmonary infection. Clinical symptoms are not obvious when the lesion is confined to a single lobe or lung segment<sup>[5]</sup>. Radiologic features of CCAM include involvement of single lobe, predominantly lower lung fields with left lung more than right. CCAM imaging pattern is described into three types:

(i) Single or multicystic pattern, (ii) nonhomogeneous mass consistent with the gross configuration of numerous, evenly spaced cysts usually <1 cm size, and (iii) a cystic or solid mass<sup>[6]</sup>. In 1977, Stocker has described three types of CCAM classification on the basis of histopathologic appearance<sup>[7]</sup>. Type I, the most frequently occurring type, was a lesion consisting of single or multiple large cysts (>2 cm in diameter) lined by ciliated pseudostratified columnar epithelium. The walls of the cysts contain prominent smooth muscle and elastic tissue. Mucus-producing cells are present in 32% of the cases, and cartilage in the wall was seen in 10%. Relatively normal alveoli may be seen between the cysts. Type II was composed of multiple small cysts (<1 cm in diameter) lined by ciliated cuboidal to the columnar epithelium and was associated with other anomalies. There were no mucous cells or cartilage. Type III was composed of regularly spaced bronchiole-like structures separated by masses of cuboidal epithelium-lined alveolus-like structures. In our case, CT showed large cystic type lesion with the pathological classification consistent with Stocker Type I. Type I CCAM has good prognosis; however, the malignant transformation was reported in 1% of cases<sup>[8]</sup>, usually occurs to a mucinous bronchioloalveolar carcinoma. At present, lobectomy is considered to be the best method for treatment of CCAM<sup>[9]</sup>.

Lung cancer presenting as thin-walled cyst or cavity has been described in various case reports<sup>[10-14]</sup>. It usually presents as peripheral lung lesion, most of them were adenocarcinomas. These are more common in females than male. CT features suggestive of malignancy include uneven thickening of the thin-walled cavity, irregular margin, septum in cavity, nodule formation, the presence of short spicules, blood vessel convergence signs, signs of lobulation, or pleural indentation. Most important is increasing the size of cyst or cavity with thickening of its wall. Histopathology shows that carcinoma cells grow randomly along the cyst wall<sup>[12-14]</sup>. Hence, the need for surgical resection and pathological examination is reaffirmed.

In our case, the patient was elderly female presented in the 6<sup>th</sup> decade, with no history of recurrent pulmonary infection. Her CT chest showed irregular cystic lesion in the peripheral right lower lobe, with uneven wall thickness and septum within the cyst. As described above, all these features raised the possibility of malignancy. However, diagnosis of CCAM was confirmed with histopathology with no evidence of malignancy.

We report a unique case of pulmonary cystic lesion masquerading pulmonary malignancy, diagnosed as CCAM in a patient presenting unusually in the 6<sup>th</sup> decade. It poses a diagnostic challenge, broadens the thinking of clinicians and increases the understanding of CCAM. Although meticulous imaging may help to differentiate it from malignancy, histopathology is mandatory to confirm the diagnosis.

### 3 Acknowledgments

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### References

- [1] Ch'in KY, Tang MY. Congenital adenomatoid malformation of one lobe of a lung with general anasarca [J]. Arch Pathol (Chic) 1949;48:221-9.
- [2] Moerman P, Fryns JP, Vanderberghe K, Devlieger H, Lauweryns JM. Pathogenesis of congenital cystic adenomatoid malformation of the lung [J]. Histopathology 1992;21:315-21.
- [3] Laberge JM, Flageole H, Pugash D, Khalife S, Blair G, Filiatrault D, *et al.* Outcome of the prenatally diagnosed congenital cystic adenomatoid lung malformation: A Canadian experience [J]. Fetal Diagn Ther 2001;16:178-86.
- [4] Khan N, Jones M, Greaves M. Congenital cystic adenomatoid malformation of an entire lung in a 33-year-old man: A case report and review of the literature [J]. Br J Radiol 2008;81:276-8.
- [5] Pelizzo G, Barbi E, Codrich D, Lembo MA, Zennaro F, Bussani R, *et al.* Chronic inflammation in congenital cystic adenomatoid malformations. An underestimated risk factor? [J]. J Pediatr Surg 2009;44:616-9.
- [6] Peranteau WH, Merchant AM, Hedrick HL, Liechty KW, Howell LJ, Flake AW, *et al.* Prenatal course and postnatal management of peripheral bronchial atresia: Association with congenital cystic adenomatoid malformation of the lung [J]. Fetal Diagn Ther 2008;24:190-6.
- [7] Stocker JT, Madewell JE, Drake RM. Congenital cystic adenomatoid malformation of the lung. Classification and morphologic spectrum [J]. Hum Pathol 1977;8:155-71.
- [8] MacSweeney F, Papagiannopoulos K, Goldstraw P, Sheppard MN, Corrin B, Nicholson AG, *et al.* An assessment of the expanded classification of congenital cystic adenomatoid malformations and their relationship to malignant transformation [J]. Am J Surg Pathol 2003;27:1139-46.
- [9] Muller CO, Berrebi D, Kheniche A, Bonnard A. Is radical lobectomy required in congenital cystic adenomatoid malformation? [J]. J Pediatr Surg 2012;47:642-5.
- [10] Lan CC, Wu HC, Lee CH, Huang SF, Wu YK. Lung cancer with unusual presentation as a thin-walled cyst in a young nonsmoker [J]. J Thorac Oncol 2010;5:1481-2.
- [11] Xue X, Wang P, Xue Q, Wang N, Zhang L, Sun J, *et al.* Comparative study of solitary thin-walled cavity lung cancer with computed tomography and pathological findings [J]. Lung Cancer 2012;78:45-50.
- [12] Qi Y, Zhang Q, Huang Y, Wang D. Manifestations and pathological features of solitary thin-walled cavity lung cancer observed by CT and PET/CT imaging [J]. Oncol Lett 2014;8:285-90.
- [13] Farooqi AO, Cham M, Zhang L, Beasley MB, Austin JH, Miller A, *et al.* Lung cancer associated with cystic airspaces [J]. AJR Am J Roentgenol 2012;199:781-6.
- [14] Yamada S, Noguchi H, Nabeshima A, Tasaki T, Kitada S, Baba T, *et al.* Basaloid carcinoma of the lung associated with central cavitation: A unique surgical case focusing on cytological and immunohistochemical findings [J]. Diagn Pathol 2012;7:175.