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Original Article

# Cystic adenomatoid malformation of the lung in adult patients: clinicoradiological features and management☆

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ABSTRACT

**Purpose:** Congenital cystic adenomatoid malformation (CCAM) of the lung in adults is very rare. We aimed to evaluate the clinicoradiological features of adult patients with CCAM.
**Methods:** Adult patients diagnosed with CCAM were evaluated for clinicoradiological presentation and management.
**Methods:** Nine patients were diagnosed with CCAM (four were incidentally diagnosed on chest imaging). Most cases appeared on computed tomography as thin-walled multiseptated cystic lesions. Two patients had another concurrent pulmonary developmental anomaly. Five patients were surgically treated and three conservatively.
**Conclusions:** CCAMs might be incidentally diagnosed on routine chest imaging in adults and can be safely treated with minimally invasive techniques.



## 1. Introduction

Congenital cystic adenomatoid malformation (CCAM) is a rare and complex developmental anomaly of the lower respiratory tract that is usually diagnosed either prenatally or in neonates and infants [1–3]. Also known as congenital pulmonary airway malformation, CCAM is considered part of the wide spectrum of bronchopulmonary foregut malformations that also includes, among others, bronchopulmonary sequestrations, bronchogenic cysts, congenital lobar emphysema, and bronchial atresia [4–7]. Its presentation or detection in adulthood is very rare, with fewer than 60 reported cases in the literature [8–12]. Adult patients with CCAM usually present with recurrent lower respiratory infections, nonspecific thoracic symptoms (cough, dyspnea), and hemoptysis [13–15]. However, adult patients with CCAM may be asymptomatic [16]. In fact, with the increasing use of thoracic computed tomography (CT), it is more likely that developmental pulmonary anomalies might be incidentally detected on routine chest radiographs or on CTs performed for other reasons. Encountering a developmental lung anomaly in an adult can be a challenge, since the congenital abnormality may be radiologically mistaken for something more sinister in this age group than in the pediatric population [17].

Although there are a few published reports of results of surgical management of congenital lung malformations in pediatric patients, there are very few data regarding the evolution, natural history, or optimal management of CCAMs discovered in adulthood, with only a few case series on the surgical management of this entity [9–11,18,19]. Based on the few reports of CCAMs in adults that have been published and on the fact that these lesions may be incidentally (and increasingly) detected in asymptomatic patients, we believe that the management of CCAMs in adulthood deserves a different approach than that of the pediatric population. The purpose of this work was, therefore, to analyze our experience with adult patients diagnosed with CCAM regarding the clinical and radiological presentation, as well as the management (including surgical treatment) of this group of patients.

## 2. Materials and methods

All adult (18 years of age or older) patients diagnosed with CCAM at our institution (Ramón y Cajal University Hospital, a 1000-bed referral hospital in Madrid, Spain) between January 1, 2005, and December 31, 2014, were identified from a combined radiological and surgical data bank. Approval from the institutional review board was not required because the present study was retrospective and observational. There were nine patients that were diagnosed with CCAM, six of which underwent surgical resection. Three patients were ultimately managed nonoperatively, although surgery was recommended in two cases (in the remaining nonoperative case, the patient was deemed inoperable

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due to advanced heart disease). A multidisciplinary panel composed of two radiologists, two thoracic surgeons, a pulmonologist, and a pulmonary pathologist assessed and reviewed each case in order to reach a final diagnosis of CCAM. It is controversial whether all types of CCAMs in adults should always be surgically excised, and in fact, two of our patients (both asymptomatic) declined surgical treatment and preferred to be followed up. We believe that in the era of multidetector CT scanners being increasingly used, more asymptomatic cases of CCAM will be incidentally detected by radiologists in adult patients, making the approach and management of these lesions different to that of the pediatric population. The medical records were reviewed for the following information: demographic data, presenting symptoms, radiological findings, operative procedures, histopathology, and postoperative course. The last three variables were assessed only in the six patients that underwent surgical resection. Operative morbidity was defined as any postoperative event prolonging or otherwise altering the postsurgical course, whereas operative mortality was defined as deaths occurring within 30 days after operation (or deaths occurred later but during the same hospitalization).

## 3. Results

The demographics and clinical characteristics of nine patients with CCAM are shown in Table 1. The most common form of CCAM in this study was CCAM type I ($n=5$). Two asymptomatic patients were found to have other simultaneous developmental anomalies of the lung (one patient had an intralobar sequestration in the same lobe as the CCAM, whereas the other patient had an intrapulmonary bronchogenic cyst in the same lung but in a different lobe).

The most common presenting symptoms were cough ($n=5$), recurrent respiratory infections ($n=2$), and hemoptysis ($n=2$), but four asymptomatic patients were diagnosed on routine chest radiographs or CTs performed for other reasons. The median duration of symptoms before operation (in the four symptomatic patients that underwent surgery) was 12.5 months (range, 2 months to 2 years). Asymptomatic patients were incidentally found to have CCAM during workup of a positive tuberculin test ($n=1$), routine preoperative chest radiograph ($n=2$), and nephrolithiasis ($n=1$). The mean duration from the incidental diagnosis of the two asymptomatic patients who underwent resection to surgery was 6 months. Radiologists were the first specialists to make or suggest the diagnosis in all cases. Most cases appeared on CT as thin-walled multiseptated cystic lesions. Figs. 1–5 give examples of the imaging findings of CCAM in our patients.

The details of the management (surgical and nonoperative) of the patients are given in Table 2. All surgical procedures were performed electively. The surgical approach was thoracoscopy for five patients and thoracotomy for one (lobectomy in all cases). The two patients that declined surgery and the patient that was deemed inoperable have all been followed-up to date, and none of them have experienced symptoms or complications related to their CCAM (mean follow-up duration: 41 months).

There were no operative deaths. No intraoperative surgical complications were noted. There was only one postoperative complication (prolonged airleak) in the patient treated with thoracotomy, but it resolved with a chest tube. On pathological examination, three patients were found to be CCAM type I, two patients were found to be CCAM type II, and one to be CCAM type III according to Stocker's classification. The three nonsurgical cases were diagnosed by the multidisciplinary panel to be CCAM type I ($n=2$) and CCAM type II ($n=1$). One preliminary pathology report in one of our patients diagnosed a superinfected bronchiectasis instead of a CCAM, but the definite report confirmed the diagnosis of type 2 CCAM. No associated malignancy was observed in the histological studies of the surgical specimens.

## 4. Discussion

CCAM is a rare and complex congenital anomaly of the lower respiratory tract that results from failure of normal bronchoalveolar development with hamartomatous proliferation of terminal respiratory units in a gland-like pattern (adenomatoid) without proper alveolar formation [20]. CCAM is part of the wide spectrum of congenital pulmonary malformations that also includes, among others, bronchopulmonary sequestrations, bronchogenic cysts, congenital lobar emphysema, and bronchial atresia [5,6,21]. CCAM is usually diagnosed either prenatally or in neonates and infants [1–3] and may cause severe respiratory distress in newborns and infants. Its presentation or detection in adulthood is very rare, with less than 60 cases reported in the literature [8–12,22]. In adults, CCAM commonly presents as recurrent pulmonary infections, nonspecific thoracic symptoms (cough, dyspnea), and hemoptysis [13–15]. However, adult patients with CCAM may be asymptomatic for years and diagnosed on routine chest radiographs or other imaging techniques performed for other reasons [11,16]. Interestingly, four of our patients presented with an abnormal radiograph or CT and no symptoms. We believe that with the increasing use of thoracic computed tomography (CT) in adult patients, more asymptomatic CCAMs may be incidentally detected on routine chest

**Table 1**
Demographics and clinical characteristics of 9 adult patients with CCAM

| Case | Age (y) | Sex | Age at diagnosis (y) | Duration of symptoms before operation (months) | Presenting symptoms | Lobe | CCAM type | Chest x-ray findings | CT findings |
|---|---|---|---|---|---|---|---|---|---|
| 1 | 26 | F | 25 | NA (incidental x-ray finding) | None | RLL | II | Multicystic radiolucency | Multicystic lesion with cysts 2 cm or smaller |
| 2 | 71 | M | 69 | NA | Cough, dyspnea | LLL | II | Subtle LLL radiolucency | Multicystic lesion with cysts 2 cm or smaller |
| 3 | 56 | M | 54 | 16 | Cough, hemoptysis, recurrent respiratory infections | LLL | I | LLL ill-defined opacity | Multicystic lesion with cysts 2 cm or larger |
| 4 | 49 | F | 48 | NA (incidental x-ray finding) | None | LLL | I | LLL radiolucency | Multicystic lesion with cysts 2 cm or larger |
| 5 | 67 | M | 67 | 2 | Cough, hemoptysis | LLL | I | LLL cystic lesion | Multicystic lesion with dominant cyst greater than 9 cm |
| 6 | 32 | F | 30 | 23 | Recurrent respiratory infections, cough | RLL | I | RLL opacity | Multicystic lesion with cysts 2 cm or larger |
| 7 | 42 | M | 42 | NA (incidental x-ray finding) | None | LLL | I | LLL cystic radiolucency | Multicystic LLL lesion with cysts 2 cm or greater. There was an incidental small bronchogenic cyst in the LUL |
| 8 | 43 | F | 42 | 9 | Cough | LLL | III | LLL nodular consolidation | Well-defined LLL mass |
| 9 | 70 | F | 69 | NA (Incidental x-ray finding) | None | LLL | II | LLL radiolucency | Multicystic lesion with cysts 2 cm or smaller. There was an incidental intralobar sequestration in the same lobe |

CCAM=congenital cystic adenomatoid malformation; M=male; F=female; NA=not applicable; RLL=right lower lobe; LLL=left lower lobe.