

## Congenital cystic adenomatoid malformation of lung in adult complicated by aspergilloma presenting as recurrent hemoptysis- 3<sup>rd</sup> case report in world literature

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**Abstract:** CCAM is a congenital anomaly of lung, usually uncommon in adults. Presentation in adults is mostly as acute fever and lung abscesses, secondary to bacterial infections. We present a case of 28 year old man having CCAM of left lung, complicated by aspergilloma, presenting with recurrent hemoptysis. Patient underwent curative resection and is symptom free in follow up.

**Keywords:** CCAM, CPAM, Aspergilloma, fungus ball, recurrent hemoptysis

### I. Case report

A 28 years-old unmarried, nonsmoker, Indian male, pursuing his Ph.D course in Soil and Agriculture, presented with the complaints of recurrent hemoptysis and chest pain for the last 2 yrs. Occasional fever with breathlessness, and anorexia was also present. There was no history of TB in family. On examination patient had PR=96/min, BP=112/74 mmHg, RR=26/min. His respiratory system revealed no tracheal shifting, breath sounds were decreased in the lower left lung fields with few crepitations. Rest of the systemic examination were within normal limits

Biochemical tests showed the raised TLC. Sputum smear examination by gram and Ziel Nelson staining for AFB were negative. Viral markers (HbsAg, HIV, Anti-HCV) were non reactive. X-ray showed a thin walled cavity in the left lower lobe [figure 1]. His recent CT Scan showed multiple thick walled cavitation in the left lower lobe with presence of mildly enhancing soft tissue density nodules and air crescent within the cavitation suggestive of fungal ball [figure 2], with evidence of fibrobronchiectasis changes [figure 3] in the left lower lobe. The patient underwent left lower lobectomy via posterolateral thoracotomy. Macroscopic examination of the specimen revealed a mass of 8x6x4 cm [figure 4], on sectioning the mass multiple cysts were noted. The largest cyst was of 2cmx2cm in size and multiple small cysts were of 3mm-5mm in size. Microscopic examination revealed large dilated bronchi lined by normal columnar ciliated epithelium and were filled with degenerated material. The number of alveoli were very few. The stroma between the bronchiole was infiltrated by chronic inflammatory cells. A significant discovery of aspergillus hyphae was made in the mass and highlighted by Grocott's methamine silver stain. Both histological and radiological findings were consistent with Type 2 CCAM.

### II. Discussion

CCAM is a developmental anomaly of lung. 80-85% of cases are detected in first two years of life, adult presentation is very uncommon<sup>[1]</sup>. CCAM was first described as a distinct disease by Chin and Tang<sup>[2]</sup>. It was classified into 3 subtypes in 1977<sup>[3]</sup>, and expanded into 5 subtypes with a new name as congenital pulmonary airway malformation by Stocker in 2002<sup>[4]</sup>. Classification is based on the number and size of the cysts. Type 1 CCAM is characterized by single or multiple cysts more than 3 cm in diameter. They account for 70% of CCAM cases. Type 2 lesion is characterized by multiple terminal bronchiolar-like uniform cysts smaller than 2cm in size, lined by cuboidal to columnar epithelium. Type 3 CCAM usually involves an entire lobe of lung and has a spongy like appearance. The new classification system added Type 0 and Type 4 in year 2002<sup>[4]</sup>, but this classification is not widely applied because type 0 is difficult to differentiate from bronchogenic cyst, and there are similarities between type 4 and pleuropulmonary blastomas. By far bacterial infection is the most frequent reported CCAM complication which causes acute fever and lung abscess<sup>[5]</sup>. To our knowledge only 2 cases are reported to have aspergilloma as a complication of CCAM<sup>[6,7]</sup>. Aspergillus primarily affects the lungs, causing 4 main syndromes: 1) allergic bronchopulmonary aspergillosis [ABPA], 2) chronic necrotizing Aspergillus pneumonia also termed chronic necrotizing pulmonary aspergillosis [CNPA], 3) aspergilloma, and 4) invasive aspergillosis.

The incidence of pulmonary aspergillosis range from 0.016% to 17%<sup>[8,9]</sup> in various studies. The most common presenting symptom is haemoptysis which is reported to be seen in 50-80% of patients<sup>[10]</sup>. Most patients will experience mild haemoptysis but massive and life threatening haemoptysis may occur particularly in patients with underlying PTB<sup>[9]</sup>.

The characteristic radiographic picture of a pulmonary aspergilloma is of one or more round or ovoid intracavitary mass. A change of position of aspergilloma within the cavity with change in position of the patient is an interesting and a variable sign<sup>[10]</sup>. However this sign is nonspecific because other entities like haematoma, neoplasm, abscesses, hydatid cyst and Wegener's granulomatosis also demonstrates this feature. Thickened pleura may be the earliest sign on x ray chest. CT scan is considered to be more accurate technique than conventional chest radiograph in defining fungus balls, particularly in fibrotic and distorted lung fields.

Surgery is the primary choice for treating CCAM and aspergilloma<sup>[11,12]</sup> systemic antifungal and steroids have shown limited results. Other therapies include intracavitary instillation of antifungal agents, embolisation of bronchial arteries for hemorrhage .

### III. Conclusions

CCAM in adults may be complicated by aspergillomas , and surgery is the treatment of choice for the appropriate patient. Radiological and clinical findings need to be confirmed by pathological diagnosis in case of cystic lesions of the lung

Proper informed consent was obtained from the patient for publishing the details in the article

### References

- [1]. Rosado-de-Christenson ML, Stocker JT: Congenital cystic adenomatoid malformation . Radiographics 1991, 11:865-866.
- [2]. Ch'in KY, Tang MY: Congenital adenomatoid malformation of a lung with general anasarca. Arch Pathol(Chic) 1949, 48:221-229
- [3]. Stocker JT, Madewell JE, Drake RM: Congenital cystic adenomatoid malformation: classification and morphologic spectrum. Hum Pathol 1977, 8:155-177
- [4]. Stocker JT: Congenital pulmonary airway malformation: a new name and an expanded classification of Congenital cystic adenomatoid malformation of the lung. Histopathology 2002, 41:424-431
- [5]. Khan NU, Jones MT, Greaves M: Congenital cystic adenomatoid malformation of an entire lung in a 33 year old man a case report and review of literature. Br J Radiol 2008, 81:e276-278
- [6]. Yonker LM, Mark EJ, Canapari CA: Aspergilloma in a patient with an occult congenital pulmonary airway malformation. Pediatr Pulmonol 2011, doi:10.1002/ppul.21538
- [7]. Feng et al.: Congenital cystic adenomatoid malformation of lung in adults: 2 rare case report and review of the literature. Diagnostic Pathology 2012, 7:37
- [8]. Glimp RA, Bayer AS. Pulmonary aspergilloma; diagnostic and therapeutic consideration. Intern Med 1983; 143:303-308.
- [9]. Faulkner S L, Vemon R, Brown PP, et al. Hemoptysis and pulmonary aspergilloma operative versus nonoperative treatment. Anu Thorac Surg 1978; 25: 380-392.
- [10]. Roberts C M, Citron K M, Strickland B. Intrathoracic aspergilloma: role of CT in diagnosis and treatment. Radiology 1987; 165:123-8.
- [11]. Andren SM, Bhattacharjee M, Keenan DJM, et al. Squamous cell carcinoma occurring in the wall of a chronic aspergilloma. Thorax 1991; 46:542.
- [12]. Butz RO, Zetima JR and Leinmger RJ. Ten years experience with mycetoma in patients with pulmonary tuberculosis. Chest 1983; 87: 356.

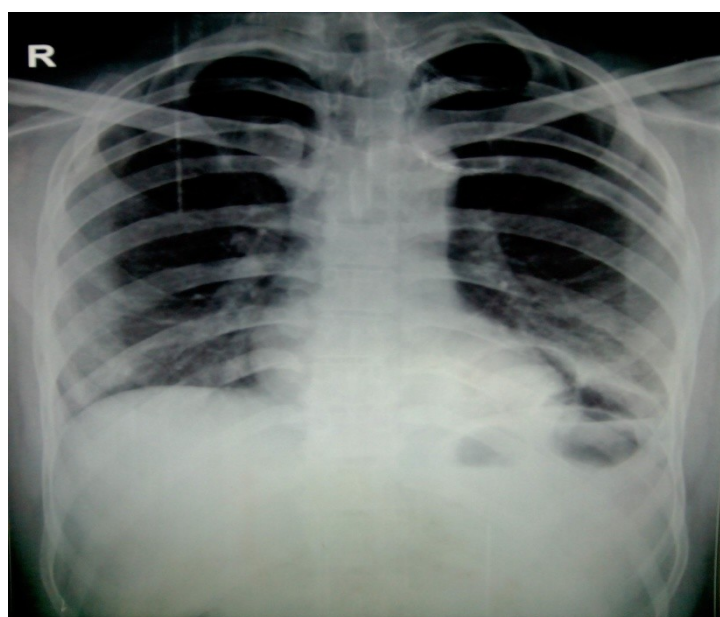


figure 1. X ray chest showing multiple cavitary lesions in the left lower lung

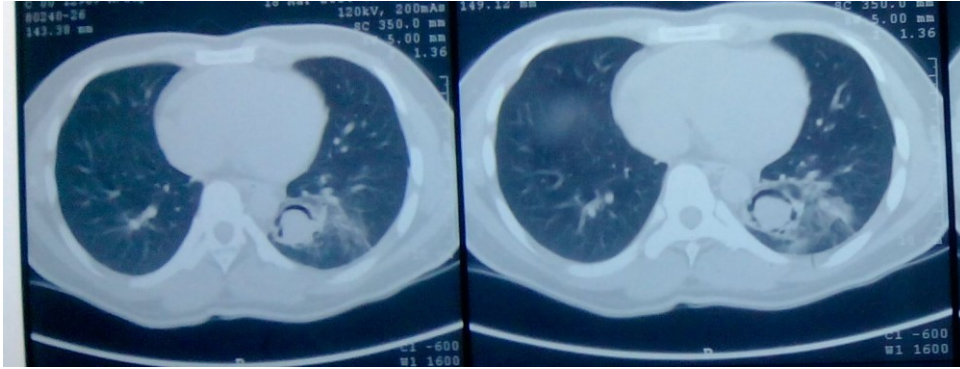


figure 2 fungus ball is visible in the cavity in left lung with surrounding crescent of air.

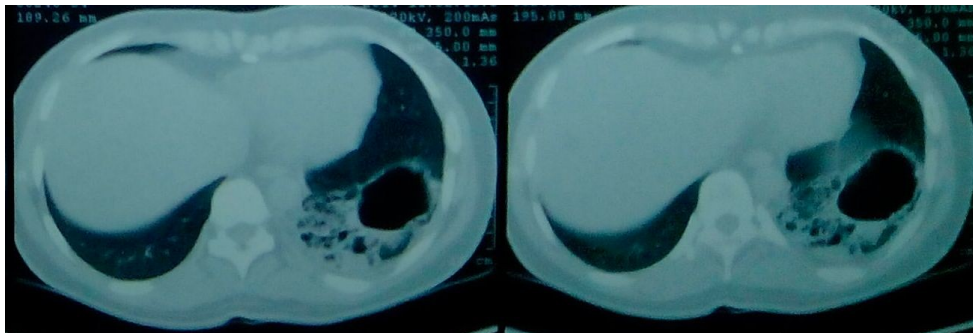


figure 3 multiple cysts of variable sizes in the left lung

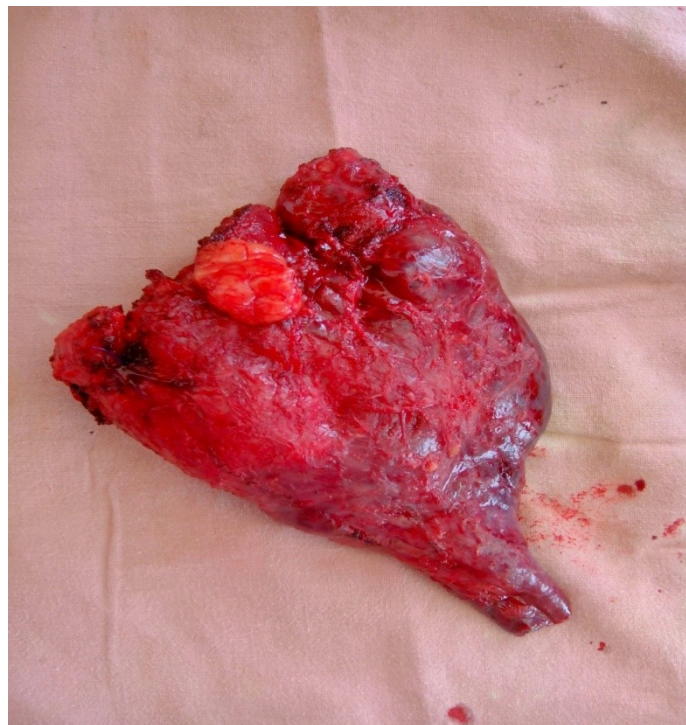


figure 4 resected left lower lobe of the lung