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Hemoptysis in a Young Adult with Congenital Cystic Adenomatoid Malformation of the Lung: Case Report

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Authors' contributions

This work was carried out in collaboration between all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Congenital cystic adenomatoid malformations (CCAM), also named congenital pulmonary airway malformation s (CPAM) are multicystic masses of segmental lung tissue with abnormal bronchial proliferation. There is failure of normal bronchoalveolar development with a hamartomatous proliferation of terminal respiratory units in a gland-like pattern (adenomatoid). CPAM can be divided into five types, according to clinical and pathologic features. [1] CPAM type 1 is the most common, and has good prognosis. The widespread use of antenatal ultrasound examination has resulted in an increase in the prenatal diagnosis of CPAM [2]. The diagnosis may not be made for many years or even until adulthood when the abnormality is small. With recurrent chest infection it does become apparent [3,4]. This case report documents young male patient with hemoptysis, and

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right lower lobe t variable sized cysts for which he underwent right thoracotomy with right lower lobectomy. The histopathology tissue report showed feature of congenital pulmonary airway malformation, Type 1(CPAM1).

Keywords: Hemoptysis; congenital cystic adenomatoid malformations.

1. PRESENTING CONCERNS

28 year-old Saudi male presented to our institution on 24th of Jan 2017, for evaluation of productive cough with blood tinged sputum, and fever for two weeks. He had a history of pulmonary tuberculosis TB status post treatment with (rifampicin, isoniazid, pyrazinamide, ethambutol, moxifloxacin and pyridoxine) three years back. In addition, he had a history of rood traffic accident (RTA) six months back with chest trauma. Computed tomography images showed multiple variable size cysts in the right lower lobe suggestive of post traumatic pneumatocele. (Fig. 1). He was treated conservative and discharge from hospital without complication.

2. CLINICAL FINDINGS

The patient appeared well-nourished and comfortable at rest. His vital signs were as follows: temperature, 38.3 C; pulse, 86 beats/ min; BP, 117/81 mm Hg; respiratory rate, 22 breaths/min, and oxygen saturation, 100% on room air. Chest examination findings included decreased breath sounds at the posterior right lower lung field. Cardiovascular abdominal, lymphatic, and genitourinary examinations were unremarkable.

2.1 Diagnostic Studies

Laboratory data on presentation were as follows: WBC count, 14^3/micro L; hemoglobin count, 10.7 g/dL. Liver function test and basic chemistry values were unremarkable. Sputa were negative for acid-fast bacilli (AFB).Chest radiograph, and Computed tomography images were notable for right lower lobe variable sized cysts (Figs. 2,3).

2.2 Therapeutic Focus and Assessment

The patient was given intravenous antibiotic, moxifloxacin, and Piperacillin/ tazobactam.He was referred to thoracic surgery. They did lobectomy for right lower lobe, and an intercostal tube was inserted at right upper lobe (Fig. 4). He was transferred to the intensive care unit. Chest Computed tomography image was done showed, large pleural collection in the right horizontal fissure, with air fluid level and air pockets (Fig. 5).

The histopathology of surgical biopsy revealed cystically dilated space lined by respiratory epithelium, ulcerated lining in many areas, cystically dilated bronchiolar spaces are also identified. The bronchioles are devoid of cartilage. Luminal secretions were seen. Intervening alveoli are noted, show moderate congestion, and mild inflammation. Feature are of congenital pulmonary airway malformation, Type 1 (Figs. 6, 7). Patient was improved and discharge on 12th of Feb 2017.

3. DISCUSSION

Congenital pulmonary airway malformation (CPAM) has incidence of 1:25000 to 1:35000 and there is a male predominance. About 15%-50% of cases of congenital cystic lung disease are reported to be CPAM [5].

Exact etiology of CPAM is not known, it is to be considered as hamartomatous malformation and abnormal proliferation of the pulmonary tissue at different sites.

The lesion might be familial and related to chromosome abnormalities. Deletion/duplication of chromosome 18 is associated with CCAM [6]. CPAM Type 0-Acinar dysplasia/agenesis is rare malformation largely incompatible with life. Lungs are small, firm with diffusely granular surface [5].

CPAM can be separated into five types, based on clinical and pathologic features [7].

CPAM Type I- It is operable with good prognosis, and accounts for nearly 65% of cases. Grossly, lesion is predominantly cystic type (measuring 3-10 cm in diameter) surrounded by smaller cysts [5]. CPAM Type II- mainly seen in first year of life, and it accounts for 10%-15% of cases. It has poor prognosis and most time associated with other congenital

anomalies like extralobar sequestration [5]. CPAM Type III-It accounts for 5% of cases. It can be seen in first few days to months of life, and has male preponderance. CPAM Type IV-It is hamartomatous malformation of the distal acinus, and can be seen in 10%-15% of cases with an age range of newborn to 4 years [5].

Adult cases have been diagnosed incidentally presenting as mass lesions on chest radiograph. Cases have also been diagnosed after presenting with recurrent infection [8], pneumothoraces [8], haemoptysis [10], mycetoma [9], and bronchioloalveolar carcinoma [11].

Congenital lobar emphysema can be distinguished from CPAM by the presence of bronchovascular markings extending to the periphery of the involved lobe and by atelectasis of adjacent tissue [12]. It is difficult to distinguish CPAM from a sequestration of the intralobar variety, but a systemic blood supply would favor sequestration [13].

The relation between CPAM and malignancy has been well documented. Malignant transformation of mucinous bronchioloalveolar carcinoma was seen in Type I CPAM [14,15]. Type II CPAM may involve malignant transformation- Rhabdomyosarcoma. pleuropulmonary blastomas must be considered

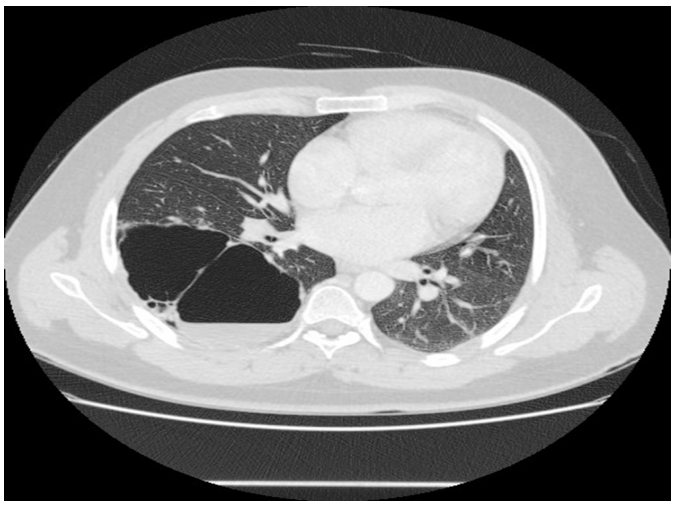

Fig. 1. Multiple variable size cysts are noted in the right lower lobe some of them show air fluid level, the largest measures 11x7x10 cm on its maximum diameter

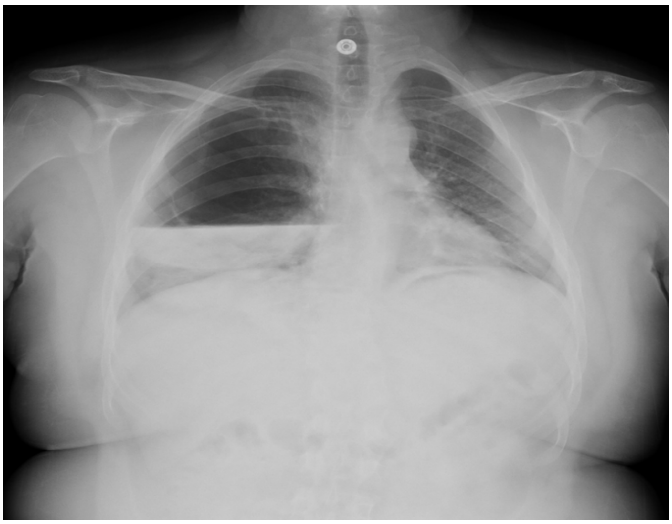

Fig. 2. Right lower lobe variable sized cyst

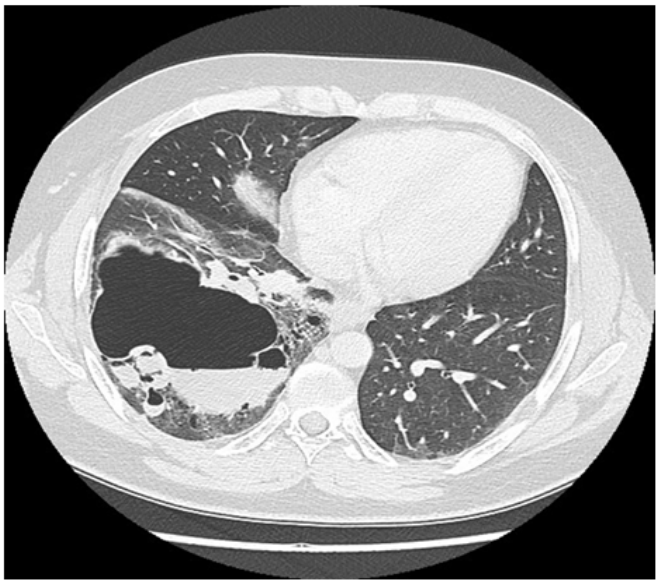

Fig. 3. A follow up study of right lower lobe cystic changes reveled worsening of fluid component with changes, suggestive of hemorrhage with or without infection. Additional the surrounding ground glass density is noted in the basal segments of the right CPAM should be considered

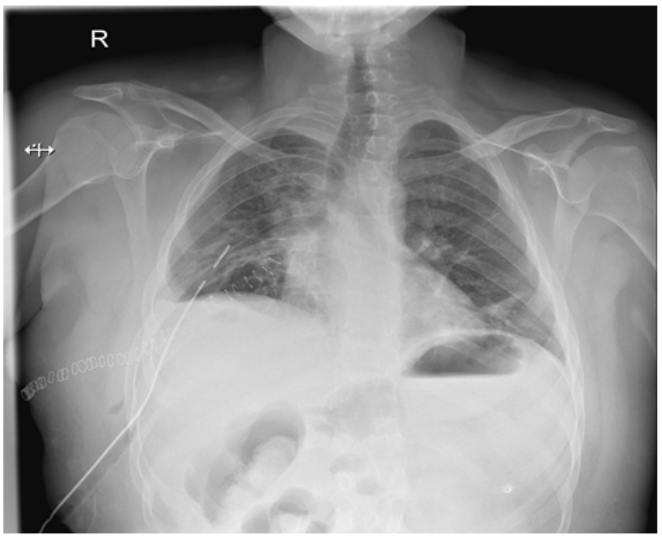

Fig. 4. Status post Right lower lobectomy with ICT

in any child presenting with cystic lung lesion especially Type III CPAM. Lobectomy should be considered for localized disease associated with recurrent infection and hemoptysis.

Surgical intervention in asymptomatic infants is associated with fewer complications and better outcome. The chances of survival can improve up to 50-60% with surgery. In CCAM lesions, betamethasone is an effective treatment. Peranteau et al. found that Survival was 100% with single course of prenatal steroids (betamethasone) in hydropic fetuses with [16,17].

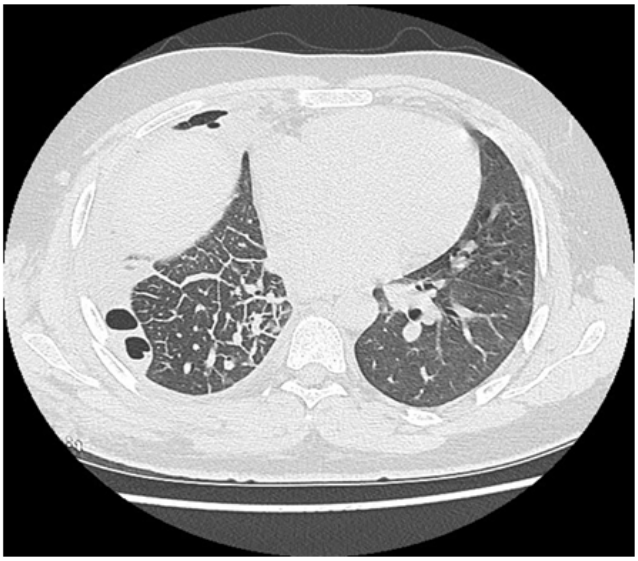

Fig. 5. Right lower lobectomy. Posterior displacement of the middle lobe. Large pleural collection seen in the right horizontal fissure, with air fluid level and air pockets. Right middle lobe small consolidation

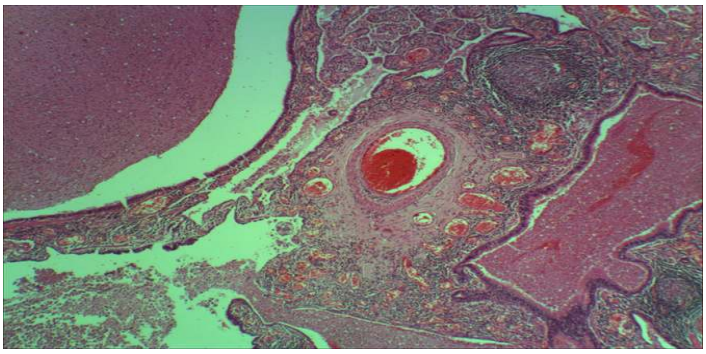

Fig. 6. Multiple variably sized cysts, devoid of cartilage in its wall, H & E, x4

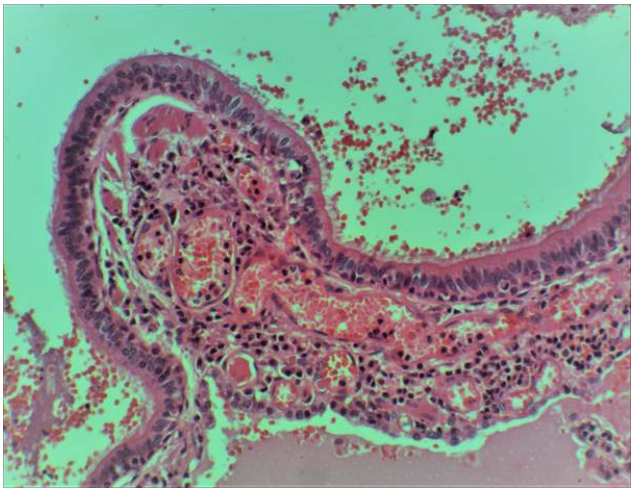

Fig. 7. The cysts are lined by ciliated bronchial type epithelium, H & E, 20X

4. CONCLUSION

CPAM in adults may have atypical clinical and radiological manifestation. Diagnosis is important for treatment, and to prevent secondary complications as hemorrhage and recurrent infection. For the possible development of malignancies strict follow-up is necessary.

CONSENT

Written informed consent was obtained from the patient for publication of this Case Report and any accompanying images.

ETHICAL APPROVAL

As per international standard ethical approval has been taken from local ethical committee the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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