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Rachid Attou & Pascal Reper

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Case Report

Complicated pneumothorax and congenital lung cystic malformation

Rachid Attou, Pascal Reper

Critical Care Department, Brugmann University Hospital, Université Libre de Bruxelles, Belgium

Congenital cystic adenomatoid malformation, also named congenital pulmonary airway malformation (CPAM), is a congenital lung abnormality which is uncommon in adults. The usual radiological appearance of CPAM is a cystic space-occupying lesion. We present one case of CPAM with unusual clinical and radiological findings, a complicated spontaneous pneumothorax with intracystic haemorrhage with successful conservative initial treatment, despite acute haemodynamic instability.

Keywords: CPAM, Pneumothorax, Cystic haemorrhage, Acute respiratory distress

Introduction

Congenital pulmonary airway malformation (CPAM) is a developmental abnormality that has been described for the first time as a disease or entity by Chin and Tang in 1949. It was classified into three subtypes in 1977, and was later expanded into five types with a new name as CPAM by Stocker in 2002.[1] In total, 80–85% of cases are recognized in the first 2 years of life; adult presentation is uncommon.[2] Most CPAMs in adults involve unilateral lobes of the lung, and may be complicated with pulmonary bacterial infections and abscesses.[3–5] We describe one CPAM case in an adult with unusual clinical and pathological findings, complicated with spontaneous pneumothorax and intracystic haemorrhage.

Case Presentation

A 25-year-old man was admitted to hospital for acute respiratory distress with acute chest pain. On clinical examination, pulmonary auscultation was asymmetric. Heart rate was 110 beats/min. Blood pressure and temperature were normal. Chest X-ray showed a complete left pneumothorax (Fig. 1); a chest tube with pulmonary drainage was easily inserted, which led to clinical improvement of the patient's condition. A few minutes later, blood began to appear in the chest tube (total 140 ml). Computed tomography (CT) scan of the chest showed a big cystic formation of 14 cm in the left lower lobe (Fig. 2) with signs of intracystic bleeding following thoracic drainage and re-expansion of the lung. Radiological findings were consistent with type I CPAM according to Stocker's classification.[1] The patient was admitted into intensive care unit (ICU) for survey; his situation was uncomplicated with conservative initial treatment. Haemodynamic situation remains stable but fluid volemization was necessary. Four hours later, the haemoglobin level was 3 g lower than when the patient was admitted in the hospital. The patient was later discharged of the ICU without further complications or medical intervention confirmed by follow-up. Definitive surgical treatment was considered on a long-term basis.

Discussion

CPAM is a rare congenital abnormality. Symptomatic presentation in adult life is extremely uncommon. The usual radiological appearance of CPAM is a cystic space-occupying lesion.

The classical histological change of CPAM is that normal pulmonary alveoli are replaced by cysts composed of adenomatoid hyperplastic bronchioles. Based on the size and number of the cysts, this lesion was initially classified into three groups. These types present the same lesion, but tend to vary on the radiological presentation. Type I CPAM accounts for 70% of cases, which is characterized by single or multiple cysts more than 3 cm in diameter lined by pseudo-stratified ciliated columnar epithelium, along with mucous cells which are considered to potentially mutant to adenocarcinoma. Type II lesion is consisted of multiple terminal bronchiolar-like uniform cysts smaller than 2 cm in diameter, lined by cuboidal to columnar epithelium. Type III CPAM usually involves an entire lobe of lung and has a spongy-like appearance, constructed by bulk gland-like structures. Another two types were proposed in 2002.[1] The new classification system with added type 0 and IV

Correspondence to: P Reper, Critical Care Department, Brugmann University Hospital, Free University Brussels, VanGehuchten 4, 1020 Brussels, Belgium. Email: pascal.reper@chu-brugmann.be

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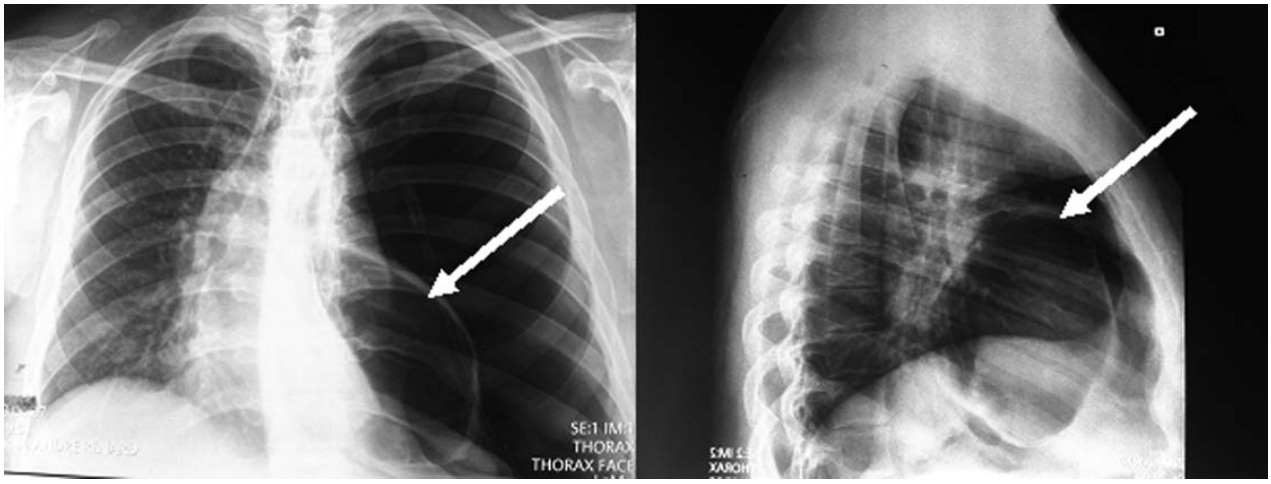

Figure 1 Complete left pneumothorax with cystic formation suggesting congenital pulmonary airway malformation (CPAM) (see arrow).

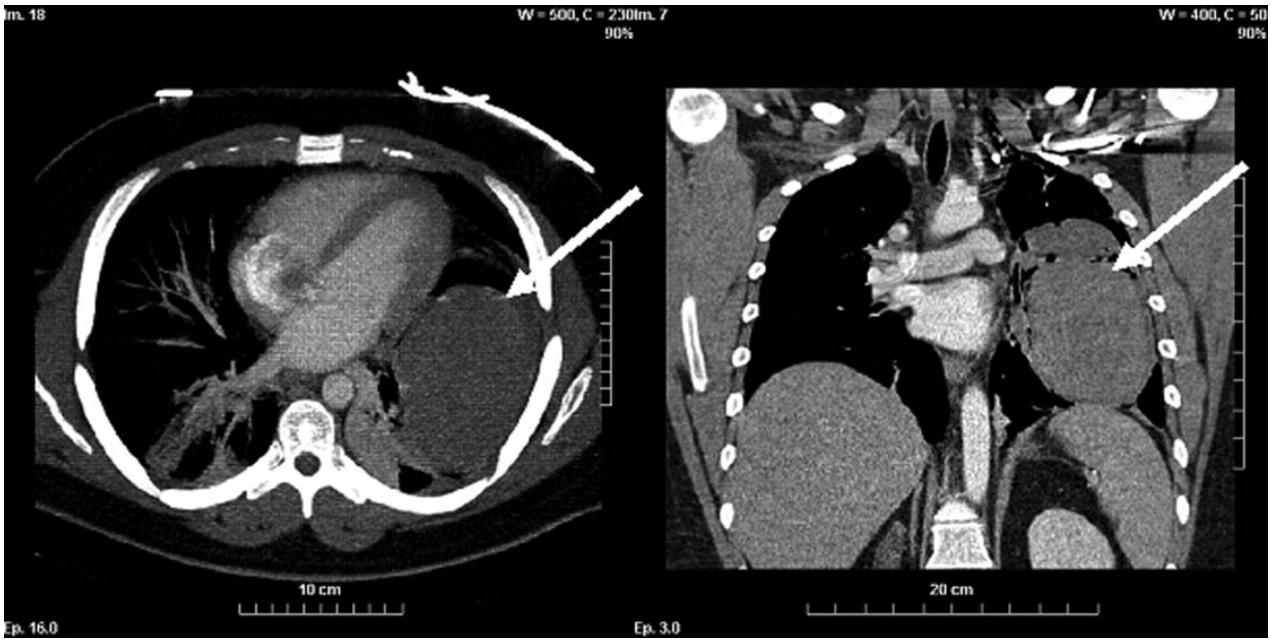

Figure 2 CT scan of the chest: big cystic formation (14 cm) in the left lower lobe with intracystic bleeding following thoracic drainage and reexpansion of the lung (see arrow).

was not widely applied because type 0 is very difficult to differentiate from bronchogenic cyst, and the similarities between type IV and cystic pleuropulmonary blastoma may result in misdiagnosis.[3] Late-onset CPAM in adults may be more complicated on radiographic images due to recurrent infections. In this report, the patient was classified into Stocker's type I CPAM for its uniform size of the cyst and regular proliferation pattern of the bronchiolar epithelium larger than 3 cm in diameter.

In our report, intracystic haemorrhage was confirmed by CT scan examination. By far, bacterial infection is the most frequently reported CPAM complication which causes acute fever and lung abscess.[4] To our knowledge, an intracystic haemorrhage with pneumothorax within CPAM has not yet been documented. Presentations of spontaneous pneumothorax may vary from that of bacterial infection. It may either produce a fulminant invasive pulmonary infection or quietly coexist for year in the human host without symptoms. Aspergillosis may also display cystic cavities or one solid mass on the CT scan that resemble lung tumour or tuberculoma. In our case, cystic haemorrhage could be caused by adenomatoid hyperplasic bronchioles producing a complex CT scan image in the case of pneumothorax. However, multicysts with uniform size and adenomatoid hyperplastic bronchial structures revealed by gross and histological examinations led to the diagnosis of cystic malformation.

The extensive involvement of the lesion increased risk and difficulty of the surgery. In our description, the initial pneumothorax was complicated with intracystic haemorrhage, but the ICU survey and medical follow-up of the patient remained uncomplicated. According to international guidelines, prevention of recurrence is as important as the initial treatment. The probability of recurrent pneumothorax

is high, infection or recurrent bleeding could occur in the future with potential catastrophic consequences, and a carcinoma may arise from the cyst as a long-term complication. Therefore, definitive surgical treatment must be considered out of acute phase in order to minimize surgical risks.

The disease might have some kind of familial relevance. Until now, the exact mechanisms of CPAM remain unknown, but Roberts *et al.* suggested that this lesion might be familial and related to chromosome abnormalities. Although the karyotype analysis was refused by the patient in this case, the possibility that this congenital lesion might be heritable or related to chromosome or genetic malfunction could not be completely ruled out.

Conclusions

CPAM is a rare congenital abnormality. Symptomatic presentation in adult life is extremely uncommon. The usual radiological appearance of CPAM is a cystic space-occupying lesion. Patients with underlying cystic lung disease can develop spontaneous complications such as pneumothorax due to pressure-volume changes. We report a case in which spontaneous pneumothorax further associated with intracystic haemorrhage was the presenting manifestation of CPAM of the lung in a previously healthy and asymptomatic young adult showing acute chest pain and respiratory distress in the emergency room. The patient was successfully initially conservatively treated.

We believe that this is an alternative to emergency surgery in complicated CPAM, even if initial fluid volemization is necessary. Further surgical treatment is indicated to prevent further catasptrophic complications such as carcinoma or recurrent bleeding.

Competing Interests

The authors declare that they have no competing interests.

Authors' Contributions

All authors have written, read, and approved the final manuscript.

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