When mother nature takes your breath away

Annemie JACOBS[1] , Herwig ALAERTS[2], Philip LERUT[3], Mathias LEYS[4]

[1] Respiratory Medicine, UZ Leuven, Belgium
[2] Anatomopathology, [3] Thoracic Surgery, [4] Respiratory Medicine
AZ Groeninge, Kortrijk, Belgium

Introduction

Congenital cystic adenomatoid malformation (CCAM) is a congenital disease in which adenomatous hyperplasia of the bronchiolar epithelium leads to the formation of numerous cysts.

We describe an adult patient in whom pulmonary lobectomy was performed, which confirmed the diagnosis of CCAM (1).

Case Report

A 29-year-old male patient was referred to the department of pulmonology with an ongoing respiratory infection despite treatment with antibiotics. In the past three years, he received multiple courses of antibiotics for recurrent respiratory infections. His medical history revealed haemochromatosis, allergic rhinitis and allergic asthma. He recently stopped smoking and worked as a cook in a nursing home.

On auscultation, inspiratory crackles and squeaks on the right side were registered. Biochemical results showed elevated inflammatory parameters. Chest X-ray showed a liquid-air-filled cystic space in the right lower lobe (Figure 1). Chest computed tomography suggested the presence of a lung abcess (Figure 2). Bronchoscopic alveolar lavage was negative for bacteria or fungi. Cultures for mycobacterium tuberculosis remained negative after 6 weeks. HIV serology and autoimmune serology were negative.

The patient was treated with broad spectrum antibiotics for 6 weeks with significant amelioration of biochemical and imaging results, but a thin-walled cystic structure remained (Figure 2). However, complaints of night sweats and general weakness persisted. Given the history of relapsing respiratory infections and the clinical suspicion of CCAM, a right lower lobectomy was performed four months after treating the pneumonia (Figure 3). Anatomopathological results confirmed the presence of congenital cystic adenomatoid malformation (Figure 4).

The patient fully recovered postoperatively and regained his functionality in daily life after an intensive rehabilitation program.

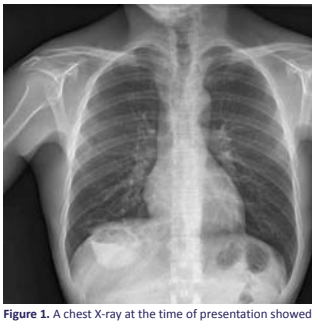

Figure 1. A chest X-ray at the time of presentation showed a liquid-air-filled cystic space in the right lower lobe.

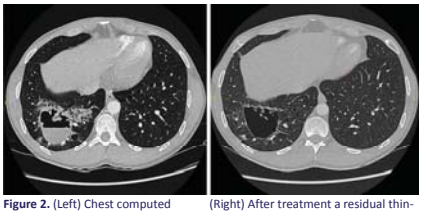

Figure 2. (Left) Chest computed tomography at presentation showed a lung abcess in the right lower lobe. (Right) After treatment a residual thin-walled cystic structure in the right lower lobe persisted.

Figure 3. Right lower lobe resection specimen.

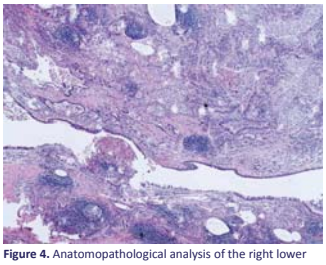

Figure 4. Anatomopathological analysis of the right lower lobe confirms the diagnosis of congenital cystic adenomatoid malformation.

Discussion

CCAM is a rare congenital anomaly. However, it remains the most common malformation of lung development (25%). Its incidence is 1 in 10000 to 35000 pregnancies, and its prevalence is approximately 9/100 000 births. The etiology of CCAM is unknown (2). Bronchopulmonary foregut malformations that are probably caused by an arrest in lung development between the 4th and 7th week of fetal life, are assumed. It does not have any genetic predisposition or gender predilection. It is not affected by maternal factors such as race, age or environmental exposures (3).

CCAMs are commonly diagnosed in the prenatal or neonatal period (4). The detection of CCAM in adulthood, after the age of 18 years, is extremely rare (1). Patients may present with symptoms, including cough, dyspnea, haemoptysis and respiratory distress, or remain asymptomatic (3). Although CCAM is a congenital disease, abnormalities previously not apparent on chest X-ray, can be detected in a later stage, with increased visibility of the lesions after repeated inflammation (1). The imagery is based primarily on CT scan, but the diagnosis remains difficult because of its scarcity (2). The clinical diagnosis, highly oriented by radiological approach, is confirmed by pathological examination.

Other diseases that must be distinguished from this disease include pulmonary sequestration, bronchial atresia, bronchogenic cysts and bronchiectasis (1). CCAM can transform into malignancies, such as bronchioloalveolar carcinoma, adenocarcinoma in aged, and pleuropulmonary blastoma (3). Surgical removal of the affected lobe of the lung is the main treatment in CCAM, even if the patient is asymptomatic (1,5). It also prevents future complications as recurrent infections and potential risk of malignant transformation (3).

References

1. Kagawa H, Miki K, Miki M, Urasaki K, Kitada S. Congenital cystic adenomatoid malformation in adults detected after infection. Respirol Case Rep 2018;6(8):1-4.
2. El Amraoui W, Bentalha A, Hamri H, El Kettani S, El Koraichi A. Congenital cystic adenomatoid malformation-dangers of misdiagnosis: a case report. J Med Case Rep 2017;11(212):1-5.
3. Mehta AA, Viswanathan N, Vasudevan AK, Paulose R, Abraham M. Congenital cystic adenomatoid malformation: a tertiary care hospital experience. J Clin Diagn Res 2016;10(11):SC01-SC04.
4. Smith JA, Koroscil MT, Hayes JA. Congenital pulmonary airway malformation in the asymptomatic adult: a rare presentation. Respir Med Case Rep 2018;25:280-2.
5. Moyer J, Lee H, Vu L. Thoracoscopic lobectomy for congenital lung lesions. Clin Perinatol 2017;44:781-94.

Conclusion

CCAM should be considered in the differential diagnosis of patients with pneumoniae recurring at the same location.

Chest CT is preferable to chest X-ray. The diagnosis is confirmed by biopsy. Surgery should be performed for CCAM, even if the patient is asymptomatic (1).

CONTACT:
annemie.jacobs@student.kuleuven.be

Poster Design: Dirk De Weerdt
www.ddwdesign.be (Po835)