CASE REPORT

Congenital pulmonary airway malformation (CPAM) with initial presentation in an adult: a rare presentation of a rare disease

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SUMMARY

Congenital pulmonary airway malformation (CPAM) is a rare congenital abnormality with unknown exact aetiology or clear genetic association. It is characterised by a failure of bronchial development and localised glandular overgrowth. Typically, it is diagnosed on prenatal ultrasound, only infrequently in children, and even less commonly in adults. We present a case of a 25-year-old man, with no previous lung diseases who presented with right-sided chest pain, fever and cough suggestive of pulmonary infection. Chest imaging, including CT scan, showed a large focal cystic mass within the right lower lobe along with ground glass opacities suggestive of CPAM. He was started on intravenous antibiotics. Bronchoscopy showed a large amount of pus in the right lung and bronchoalveolar lavage confirmed the microbiological diagnosis of methicillin-resistant *Staphylococcus aureus.* He improved with antibiotic treatment. He was discharged with 6-week course of antibiotics and follow-up afterward.

BACKGROUND

Congenital pulmonary airway malformation (CPAM) is a rare congenital abnormality that presents in 0.004% of all pregnancies, and constitutes ~25% of all congenital pulmonary anomalies.[1–3] It is characterised by failure of bronchial development and localised glandular overgrowth with no known aetiology or clear genetic association.[4–7] CPAM occurs equally across races and genders.[8] Typically, it is diagnosed prenatally[7] and only infrequently in children and adults.[1] When it presents in adults, the most common presentation is recurrent and resistant infections, even though some of these lesions are also found incidentally on chest imaging in asymptomatic patients.[4 5 8 9] CT scan, in general, is a reliable method for diagnosis and classification.[10] Stocker's criteria are used to classify CPAM from type 0 through type IV. This is ordered by the affected region of the lung from most proximal to most distal.[11–13] Type I CPAM is the most prevalent and is associated with the most favourable prognosis.[6] In adult-diagnosed CPAM, chronic inflammation from recurrent infections may cause alteration in the radiological and histological presentation.[4 14] In such cases, biopsy is often necessary to rule out malignancy.[1 9]

CASE PRESENTATION

A 25-year-old man, with no medical history, presented to the emergency department with right-sided chest pain, fever and cough for around 12 days. He denied any previous symptoms of respiratory infections even as child. He is a current smoker, 10 cigarettes daily since age 15, used to snore and smoke heroin but quit 5 years ago. He had two teeth extractions in the past but no other surgeries. Patient's uncle died of pneumonia at age 40, otherwise no other significant family history of lung diseases.

At the emergency department, he had fever 103.0°F (39.4°C), tachycardia 110–150 bpm, blood pressure 89/53, respiratory rates 18 and oxygen

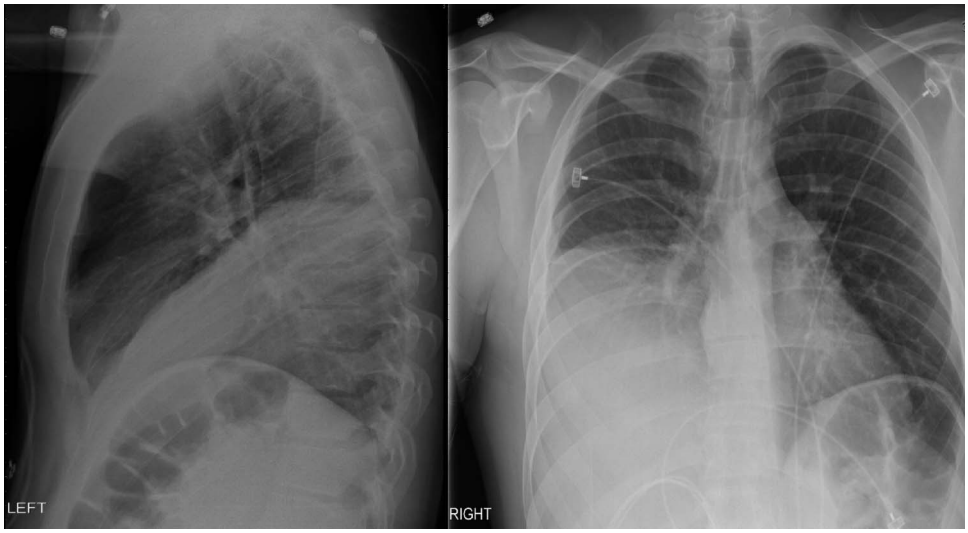

Figure 1 Two-view chest X-ray at presentation showing near-complete opacification of the right lower lobe.

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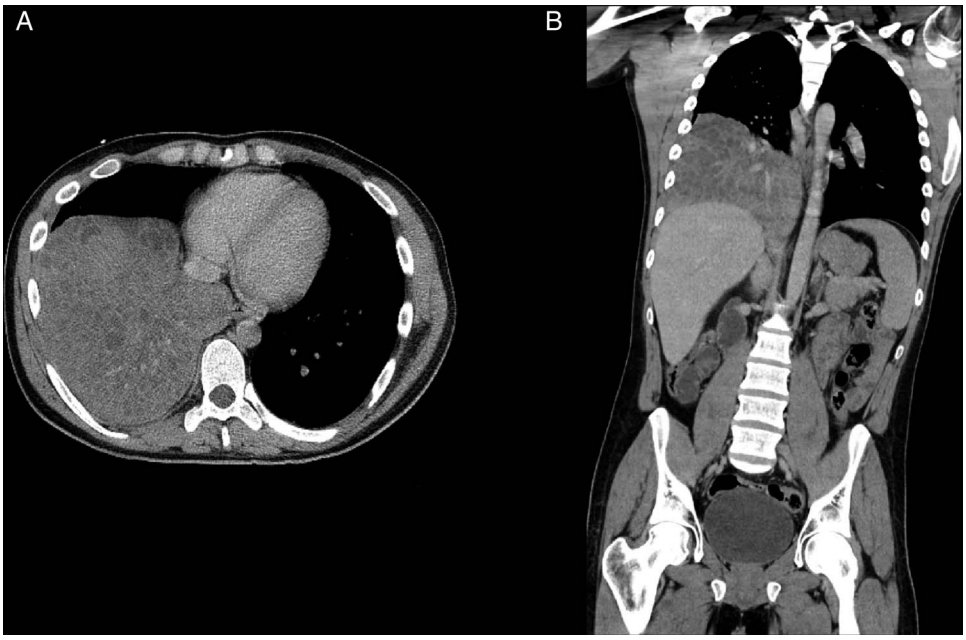

Figure 2 Chest CT with IV contrast at presentation showing a large focal cystic mass within the right lower lobe along with ground glass opacities, suggestive of congenital pulmonary airway malformation type 3 given the appearance of microcysts and the involvement of an entire lobe.

saturation >94% on room air. Chest examination revealed shallow breaths, decreased breath sounds in right interscapular area and dullness to percussion in the same area.

INVESTIGATIONS

Complete blood counts showed white cell counts of 36.7K/mm^3. Two-view chest X-ray showed near-complete opacification of the right lower lobe (figure 1), this was followed up by chest CT with IV contrast that showed large focal cystic mass within the right lower lobe along with ground glass opacities (figure 2). Radiologist suggested the mass to be CPAM, which was previously named as congenital cystic adenomatoid malformation (CCAM) and more consistent with type 3 given the appearance of microcysts and the involvement of an entire lobe.

TREATMENT

He was admitted to the intensive care unit with the diagnosis of infected CPAM with suspected right lower lobe abscess and sepsis and was started on broad spectrum intravenous antibiotics, including vancomycin, piperacillin–tazobactam and levofloxacin to cover and double cover *pseudomonas*.

Bronchoscopy was performed on hospital day 3 and showed large amounts of pus coming out from the right lower lobe segments and there were no gross abnormalities in the anatomy of the right lower lobe segments though deeper subsegments could not be examined due to the pus that could not be completely suctioned. Bronchoalveolar lavage and Brush sampling were positive for methicillin-resistant *Staphylococcus aureus*. Accordingly, the patient's antibiotics were de-escalated to vancomycin alone.

OUTCOME AND FOLLOW-UP

In the following few days, he started to improve clinically and his white cell counts dropped to 26.7K/mm^3 (figure 3). He was transferred to medical floor. The patient developed red man syndrome as a side effect from vancomycin. The infectious disease team was consulted, and they switched the patient to IV

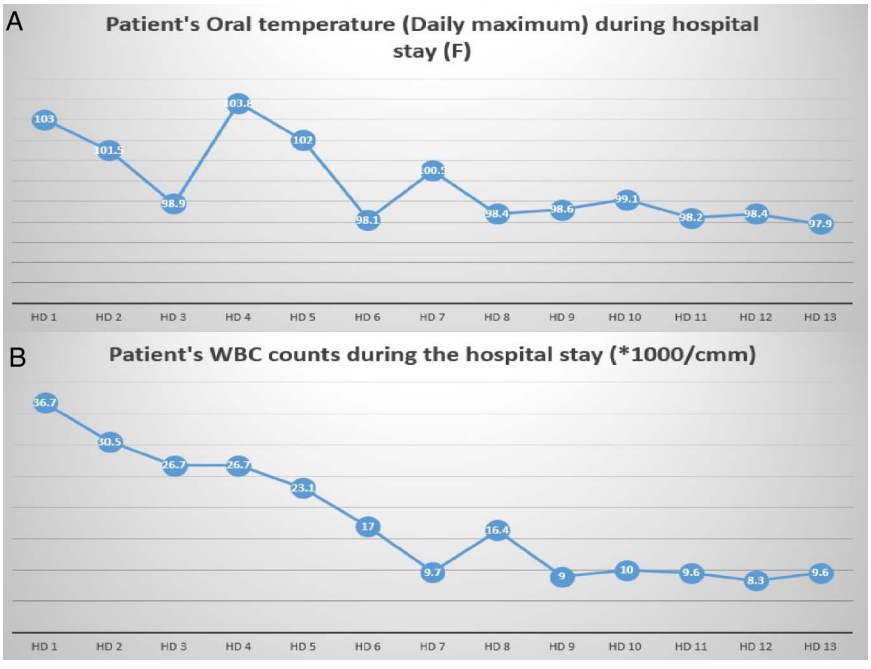

Figure 3 The trend in patient's oral temperature in Fahrenheit (A) and white cell counts (B) throughout hospital stay.

clindamycin with plans to discharge him on oral clindamycin for 6 weeks, followed by follow-up CT scan of the chest. Unfortunately, the patient missed his next appointment despite our attempts to contact him, so the follow-up CT scan of the chest was not performed.

DISCUSSION

CPAM, which was previously named as congenital cystic adenomatoid malformation (CCAM), is a rare congenital abnormality that presents in 0.004% of all pregnancies,[1 2] and constitutes ~25% of all congenital pulmonary anomalies.[3] Its aetiology is poorly understood.[4 5] It is characterised by a failure of bronchial development and localised glandular overgrowth.[6 7] Its prevalence appears to be equal across race and gender,[8] and it lacks a clear genetic association.[4] Typically, it is diagnosed on prenatal ultrasound;[7] however, in the literature, it is also infrequently diagnosed in children and even less commonly in adults.[1]

When this anomaly is diagnosed after birth, there are a number of likely complications. The most common presentation in adults is recurrent and resistant infections. However, some of these lesions are also found incidentally on chest imaging in asymptomatic patients.[4 5 8 9] Other common presentations include pneumothorax, dyspnoea and haemoptysis.[5 8 9] Late presentation is typically diagnosed, and fairly reliably classified, by CT scan of the chest.[5 10] In adults, biopsy is often necessary to rule out malignancy.[1 9]

By Stocker's criteria, CPAM has been categorised from type 0 through type IV—ordered by the affected region of the lung from most proximal to most distal.[11] Type 0 involves the primary bronchi or trachea. This type is not frequently discussed in the literature, as those affected do not survive gestation.[12] Type I is the most prevalent, accounts for 60–70% of cases, and is associated with large (up to 10 cm) cystic lesions of the bronchi.[11] These patients have the most favourable prognosis.[6] Type II is characterised by bronchiolar involvement with many small (<2 cm) cysts.[11 12] This type carries a risk of a variety of other developmental abnormalities—most commonly other intrathoracic, abdominal or skeletal malformations.[11] Type III involves the distal bronchiolar and alveolar regions, and is associated with large lung masses with subcentimetre cysts.[2 11 12] Finally, type IV is associated with the most distal acinar region of the lung, with large cysts similar to type I.[11 12] Histologically, these cysts are made up of the same cell types as the region from which they were derived, from pseudostratified ciliated columnar in type I, transitioning to more cuboidal epithelium in types II and III, and finally squamous cells in type IV.[11 12] There has also been a variety of cross-over patterns described in the literature, among the CPAM classifications[10] and other congenital malformations, with bronchial atresia and bronchopulmonary sequestration being highly associated with CPAM.[7 10 13]

In adult-diagnosed CPAM, chronic inflammation from recurrent infections may cause alteration in the radiological and histological presentation.[4 14] This chronic inflammatory process is also thought to be the stimulus for malignant transformation.[8] This is a more common manifestation in type I disease, and is particularly associated with bronchoalveolar carcinoma, as well as adenocarcinoma.[7 8] Malignancy may often be detected in patients in early adulthood.[1 7] For this reason, resection is generally recommended, regardless of the patient's symptomatology.[5 7]

In our case, we presented a type III CPAM which is an uncommon type of CPAM. In this type, it is very uncommon to be asymptomatic until adulthood. Our patient has never been diagnosed with any lung infections until he presented at age of 25.

Learning points

- Congenital pulmonary airway malformation (CPAM) is a rare congenital abnormality which presents typically prenatally, only infrequently in children and adults.
- The most common presentation in adults is recurrent and resistant infections. However, some of these lesions are also found incidentally on chest imaging in asymptomatic patients.
- CPAM are classified according to the affected region of the lung from most proximal to most distal.
- In adults presenting with CPAM, biopsy is often necessary to rule out malignancy.

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Patient consent Obtained.

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