HYBRID LESION OF CONGENITAL CYSTIC ADENOMATOID MALFORMATION AND BRONCHOPULMONARY SEQUESTRATION: A CASE REPORT

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Abstract

A 7-year-old well and healthy boy was hospitalized for the first time for severe respiratory distress. He was diagnosed with congenital cystic adenomatoid malformation (CCAM) and bronchopulmonary sequestration (BPS). CCAM and BPS are rare congenital pulmonary diseases and their presentation concurrently as a hybrid lesion is even rarer. CCAM is usually diagnosed prenatally however it may regress postnatally and the patient may be asymptomatic. BPS usually presents after birth or later in childhood with recurrent lung infections. We highlight this case of hybrid lesion of CCAM and BPS with an atypical clinical presentation which was lifesaving surgically with diagnosis made via radiographic imaging.

Keywords: Bronchopulmonary Sequestration, Congenital Cystic Adenomatoid Malformation, Congenital Pulmonary Disease, Hybrid Lesion, Lung Mass

Introduction

Congenital cystic adenomatoid malformation (CCAM) and bronchopulmonary sequestration (BPS) are congenital pulmonary diseases that have the same embryological character and clinical presentation. However, a hybrid of both CCAM and BPS is very rare. CCAM is usually detected during an antenatal scan and it may regress postnatally with appropriate treatment. On the other hand, BPS can be present at birth or in older children with varying presentation and clinical symptoms. We highlight a case of hybrid lesion of CCAM and BPS as this is an extremely rare condition that is surgically treatable.

Case Report

A 7-year-old boy with no known medical illness presented with a week's history of cough. He also complained of left upper abdominal pain. Subsequently, he developed acute onset of shortness of breath and required ventilator support. There was reduced air entry at the left lung on auscultation. Otherwise, the rest of the physical examination findings were normal. The biochemical investigations noted that the infective parameters were raised. The white cells count was 49 x 10^9/L, C-reactive protein (CRP) was 150 mg/L, and erythrocyte sedimentation rate (ESR) was 60 mm/hr.

Further history was obtained from his guardian. Antenatally, the pregnancy ultrasonography screenings were carried out in a district clinic, with no significant abnormalities detected. Postnatally, he was born full-term without complications in a district hospital. He was discharged well without neonatal care unit admission. His development was up to age and he was doing well in daily activities. He had several episodes of upper respiratory tract infection with

a few visits to a local clinic. Otherwise, he had never been hospitalized. On admission, the chest radiograph showed left hemithorax opacity, with the trachea and mediastinum shifted to the right (Figure 1). Ultrasonography over the left hemithorax noted a large heterogeneous mass occupying the mid and lower parts of the left hemithorax with left pleural effusion. Subsequent contrast-enhanced Computed Tomography (CECT) of the thorax revealed a heterogeneously enhancing mass occupying the left hemithorax. Multicystic lesions noted within the mass (Figure 1). This mass had no communication with the left bronchial tree. Opacified vessels were seen traversing within the mass. The feeding artery was noted to be a branch of the abdominal aorta while the venous return was noted to be continuous with the left pulmonary vein (Figure 2). Diagnosis of BPS was made based on the abnormal blood supply to the left hemithorax lesion. However, with the presence of multicystic component within the mass, CCAM was also considered. Thus the diagnosis of hybrid lesion of CCAM and BPS was made. The less likely differential diagnosis was lung malignancy.

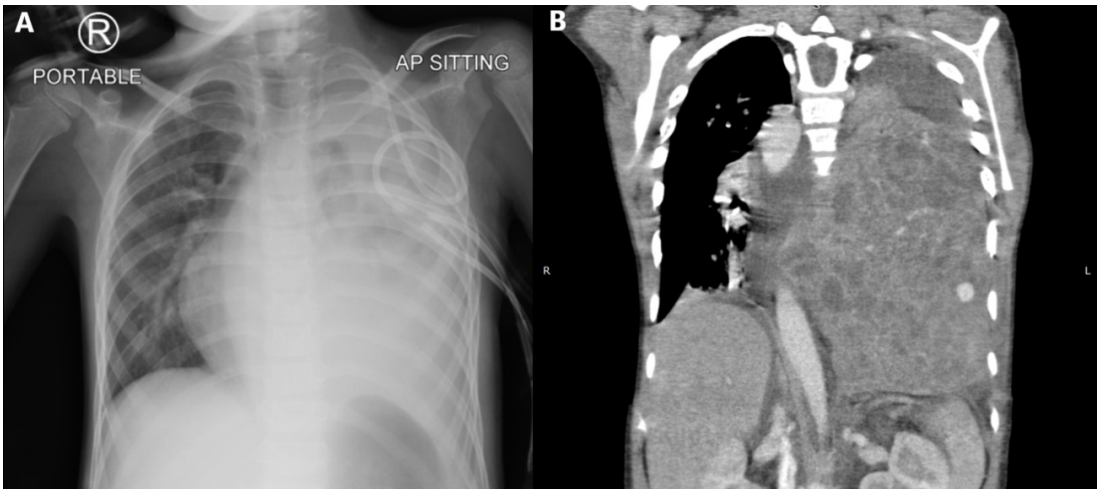

Figure 1: Chest radiograph (A) and contrast-enhanced Computed Tomography in coronal view (B). Large mass occupying left hemithorax causing homogeneous opacity of the left hemithorax on the radiograph (A). Huge heterogeneous mass was seen in the computed tomogr aphy (B) with evidence of multicystic component within.

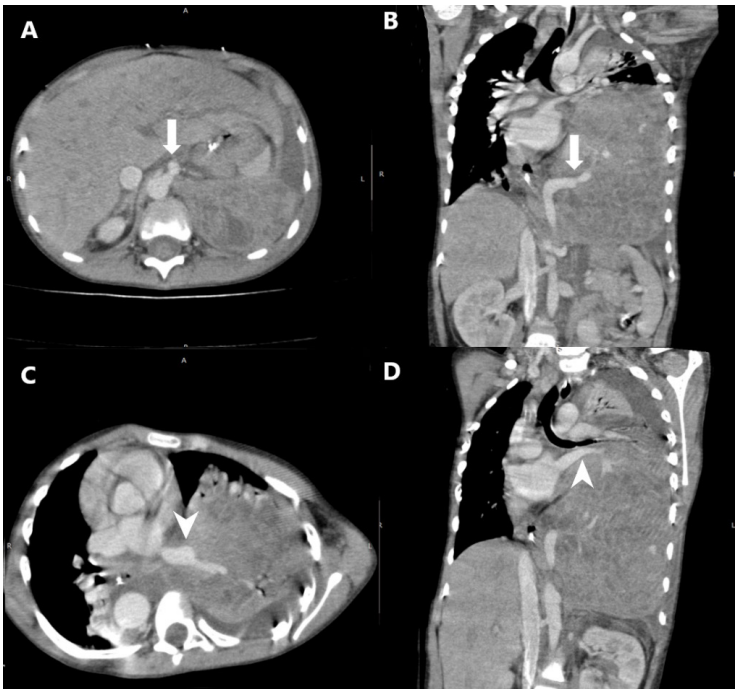

Figure 2: Contrast-enhanced Computed Tomography of the thorax in axial (A and C) and coronal (B and D) view. Multicystic mass at the left hemithorax with arterial blood supply from the aorta (arrow in A and B), while venous return to the pulmonary vein (arrowhead in C and D).

The child subsequently underwent resection of the lesion. Intra-operative findings confirmed the radiological findings of BPS. Histopathology further confirmed the diagnosis of cystic adenomatoid malformation (Figure 3). The child was discharged home well. He was noted to be well on subsequent clinic follow-up.

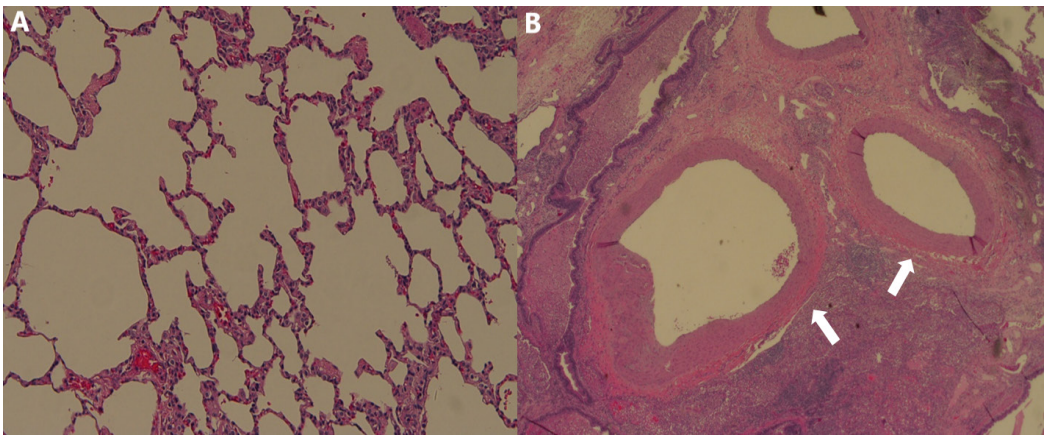

Figure 3: Histopathology slides (A and B) of the congenital cystic adenomatoid malformation showing multiple cysts formation lined with ciliated epithelium (arrow).

Discussion

Both CCAM and BPS are congenital pulmonary diseases (1). Both these diseases are of similar embryological development pattern and clinical presentation (2, 3). A hybrid lesion is known as a combination of both CCAM and BPS where an abnormal systemic blood supply and venous return are seen within a CCAM lobe or a sequestrated lung with a CCAM component (2).

CCAM was first described by Ch'in and Tang in 1949 (4). It is characterized by multicystic malformation at the terminal bronchioles replacing normal alveoli (3). It makes up about 25-30% of congenital pulmonary disease (1). Three different types of CCAM were mentioned by Stocker et al. (5) which was further classified by Adzick et al. (6) into macrocystic (type 1) and microcystic (type 2). It has no preference for the side of the lung, sex, or ethnicity (7). CCAM is mostly detected antenatally via ultrasonography (1, 7, 8). The progression of the disease may vary. It may cause severe respiratory distress postnatally where the patient will need urgent surgical treatment (8). On the other hand, it may also regress postnatally and the patient may be asymptomatic for life.

BPS is a portion of the lung parenchyma that has no communication with the tracheobronchial tree (7, 8). It makes up about 0.15% to 6.4% of all congenital pulmonary diseases (9). BPS has a separate blood supply from the aorta with the venous return either to the pulmonary vein or the vena cava. It can be further divided into intralobar (ILS) and extralobar sequestration (ELS) (3, 7, 9). ILS is situated within a lung lobe with the same visceral pleura as the normal lung, where ELS is separated from the normal lung with separate pleura (9). The clinical presentation of BPS varies with age. It may occur at birth or later in life. At birth, a child will develop respiratory distress. For an older child, it may be asymptomatic at the beginning however the child will be presented with recurrent lung infections and respiratory distress. ILS may present at any age, with the prevalence at about 20 years old (9). ELS is more commonly detected during the antenatal or neonatal period (9, 10).

As for our patient, he was not known to have any medical illness. Antenatal ultrasonography screenings were carried out during the gestational period in a district clinic where no significant abnormality was detected at that time. Due to the limitation in a district clinic, no detailed antenatal ultrasonography screening was done. He was born term without any complications. He was never admitted to the neonatal care unit for respiratory distress. Postnatally, he was well, and development was appropriate for age. He was at an older age upon his first presentation of severe respiratory distress although he had multiple episodes of upper respiratory tract infection previously. He had a short history of pulmonary symptoms which deteriorated very quickly requiring ventilator support. Correlating with the usual clinical presentations for both CCAM and BPS, the diagnosis of CCAM for our patient was not obtained antenatally or at a younger age. However, our patient's clinical presentations are corresponding to BPS where the older child may have recurrent lung infections.

Radiological imaging techniques are important for obtaining a diagnosis. A plain chest radiograph is a useful screening tool of the thoracic region as this is a relatively low dose of radiation and fast imaging technique. In this case, the plain chest radiograph significantly showed the abnormal homogeneous opacity of the left hemithorax with mediastinal shifted to right (Figure 1A). Ultrasonography technique is a useful non-radiation technique to assess the lesion, especially in children. The ultrasonography of the left thorax for our patient showed large heterogeneous mass at the mid and lower part of the left hemithorax with

left pleural effusion. However, this technique is limited with lower sensitivity in assessing the lungs.

CECT thorax is a superior radiological imaging technique that provides more detailed information before surgical intervention and proper planning. However, one of the disadvantages of CECT is that it requires the child to be cooperative during the scan, or else sedation might be required. The complication of the contrast media and sedation administration also need to be considered for the safety of the patient. CECT provides multiplanar images of the thorax and the characteristics of the lesion can be studied in detail. The cystic component of the lesion can be well seen in the CECT. Also, the vascular supply of the lesion can be identified. The arterial supply for our patient is a branch from the aorta where the venous return is to the pulmonary vein (Figure 2). Furthermore, the radiographic diagnosis of intralobar sequestration for our patient was confirmed surgically. The type 2 CCAM was confirmed with evidence of cystic airspaces lined with ciliated epithelium histopathologically (Figure 3).

Management for CCAM varies according to the clinical condition of the patient and the progression of the disease. Urgent surgery will be conducted if the child develops respiratory distress postnatally, or else close follow up for the child is needed to monitor for the progression of the disease. No surgical intervention is needed if the CCAM regresses over time. Conversely, management of BPS is surgical intervention. Early diagnosis and surgery for BPS are crucial to prevent overgrowth of the sequestrated lung which may restrict the growth of the normal lung segments causing respiratory distress or heart failure in the long run. As for hybrid lesion of CCAM and BPS, the definitive treatment is surgery.

Conclusion

Diagnosis of hybrid lesion of CCAM and BPS is difficult when relying solely on clinical history. Radiological imaging techniques are helpful tools to obtain the correct diagnosis and proceed to appropriate treatments that are life-saving for the patient.

Acknowledgement

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Competing interests

The authors declare that they have no competing interests.

Ethical Clearance

Written consent was taken from the patient's guardian for the case report.

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