CASE REPORT

Congenital Pulmonary Airway Malformation Type 4: A Case Report

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Abstract

Congenital pulmonary airway malformation (CPAM) is a very rare pulmonary disease in children. It is often misdiagnosed as a persistent and localized pneumothorax without any further pathological diagnosis. Malignancy and frequent airway infections are major concerns in these patients. Surgical excision is recommended to make a definite diagnosis and exclude hidden malignancies, and is also the treatment of choice. Herein, we report a 1-year-old girl presenting with an acute airway infection. Localized radiolucency was incidentally noted in the left upper lung field on a chest radiograph. No change in the radiolucent lesion was noted after three months of observation, so she received surgical excision. Histological examination disclosed type 4 CPAM without a malignant component thereby avoiding the possibility of repeated airway infection. After surgery her general condition was good, and no new respiratory symptoms were noted after follow-up for one year. (J Pediatr Resp Dis 2013;9:48-52)

Key words: child, congenital pulmonary airway malformation, cyst, radiolucent

INTRODUCTION

Congenital cystic lung disease in children is rare with various clinical presentations. Pulmonary sequestration, congenital pulmonary airway malformation (CPAM), congenital lobar emphysema, and bronchogenic cysts are the four major congenital cystic lesions found in the lungs. They can present at any age and the clinical appearances vary from immediate postnatal respiratory failure to an occasional finding on chest radiography. The diseases may result from compromised interactions between embryologic mesodermal and ectodermal lung components during development. They share similar embryologic and clinical characteristics, and are also known as bronchopulmonary foregut malformations.1

Congenital pulmonary airway malformation (CPAM) is relatively rare, with a reported incidence between 1:25,000 and 1:35,000.2 About 15-50% of cases of congenital cystic lung diseases are reported to be CPAM.2-5 The pathogenesis is unknown, however malformations and abnormal proliferations of different sites of the airway are the basis of CPAM classification.6,7 Increased cell proliferation and decreased apoptosis may cause malignant transformations, and pleuropulmonary blastoma and bronchioloalveolar carcinoma in particular have been reported.2,6,8-11 It is important to make a pathological diagnosis by surgical excision in congenital cystic lung diseases to determine the prognosis. The timing of surgery needs to be carefully considered, especially with asymptomatic patients such as the current case.
CASE PRESENTATION

A 1-year-old girl was brought to our outpatient department suffering from productive cough and fever for 1 week at a local hospital. Neither dyspnea nor tachypnea was noted. She was born after an uneventful 37 weeks of gestation. Her birth body weight was 3025 g and prenatal ultrasonography and postnatal physical examinations showed no abnormalities. She had not been hospitalized after birth for any reason before this presentation.

She was admitted to the hospital with a fever of 38.3°C, tachycardia with heart rate 134/min, and bilateral coarse breathing sounds over the lower lung field but relatively decreased sounds over the upper lung field. After admission, a chest radiograph was taken which showed a large radiolucent area in the left upper lung field. However, no mass effect of mediastinal shifting or signs of right lung atelectasis were noted. Bronchopneumonia with bacterial infection and lobar emphysema was then considered. Needle tapping once with 10 ml of air was performed, and she was given amoxicillin and clavulanic acid treatment for 7 days. She was then discharged after a normal physical examination with afebrile and smooth breathing status. No pathogens were yielded during this hospitalization. Due to the presence of the persistent radiolucent lesion over the left upper lung field for 3 months without clinical respiratory symptoms, she was transferred to our outpatient department for a second opinion.

On examination at our hospital, the radiolucent lesion was noted in the left upper lung field without mass effect to the adjacent tissue and with no change in size from the findings at the local hospital (Fig. 1). Presuming that the emphysematous tissue of the left upper lung was congenital lobar emphysema or a bronchogenic cyst, a segmental resection was performed after 3 months of observation. A gross histological examination (Fig. 2) showed a large cyst measuring 5 x 4 x 1.5 cm in size. There were multiple cysts of various sizes within the thin wall compressing the normal pulmonary parenchyma. Microscopically (Fig. 3), a type 4 CPAM lesion composed of peripheral large cysts with thin walls and a flattened alveolar-type epithelial lining (type I alveolar lining cells) was seen. There were few mesenchymal cells, no cuboidal or columnar cells, and in particular an absence of ciliated cells and...
mucus cells. In addition, there was no sarcomatous proliferation or other immature tissues.

The post-surgical condition of the patient was good, and subsequent chest radiography showed normal results without residual or recurrent cystic lesions (Fig. 4). No new respiratory symptoms or repeated pulmonary infection episodes were recorded during one year of follow-up.

**DISCUSSION**

Congenital cystic adenomatoid malformation (CCAM) was first described by Ch’in and Tang in 1949, and is now also known as CPAM due to the new classification system established by Stocker et al.6 There are five types of CPAM including tracheobronchial (type 0), bronchial/bronchiolar (type 1), bronchiolar (type 2), bronchiolar/alveolar (type 3), and distal acinar (type 4), classified according to the location of the developmental site of malformation.6

Diverse clinical presentations ranging from asymptomatic to life-threatening have been reported. The severity of disease is based on the “timing” of the malformation and the balance of proliferation and apoptosis. For neonates, the size of the CPAM is typically correlated with the severity of clinical problems during pregnancy (hydrops, preeclampsia, polyhydramnios) and diagnosed by antenatal ultrasonography. The bigger the cystic lesion the more severe the respiratory distress presented at birth. For infants and children, malformations communicate with the tracheobronchial tree build valvular mechanism causing hyperinflation of the cystic zone during inspiration and less deflation during expiration. Enlarged cysts compressing the normal pulmonary structure lead to the respiratory symptoms. Glandular tissue proliferation may cause mucin accumulation and recurrent infections. The majority of clinical presentations of CPAM in different ages have been reported as respiratory distress and failure in infants, and repeated airway infections in children.1,4,13-15 In adulthood, the most common presentation is also repeated airway infections. To the best of our knowledge, there have been fewer than 60 reports of CPAM in patients aged 18 and over. A series of airway conditions have been described including lung abscesses, pyopneumothorax, and hemopneumothorax. A common observation is that a diagnosis in adults is usually delayed.13

Early surgical resection has been reported to be an effective and safe treatment and diagnostic method.
for CPAM in infants, children and even in adults.\textsuperscript{3,16,17} A surgical complication rate of less than 10\% was reported in one study on 47 infants, with only one lethal complication due to severe pulmonary hypertension post left side pneumonectomy.\textsuperscript{3} Lujan et al. reported on recurrent pneumonia and the misdiagnosis of spontaneous pneumothorax in children.\textsuperscript{14} In the present case, a persistent and localized radiolucency during the first episode of lower airway infection was incidentally discovered. Early surgical resection was performed smoothly without complications thereby preventing recurrent pulmonary infections. To avoid the risks of repeated pulmonary infections and air tapping, we recommended radical surgery of the lesion once a diagnosis has been established.

The association between CPAM and malignancy has also been well documented. Malformation and proliferation cause hamartomas over the tracheobronchial tree. Type 1 CPAM may involve malignant transformation of mucinous bronchioloalveolar carcinoma,\textsuperscript{8,9,18} and type 4 CPAM requires examination of the entire lesion to exclude pleuropulmonary blastoma by confirming whether or not sarcomatous differentiation is present in the solid parts.\textsuperscript{2,6,11,19} Even though malignant transformation is rare, the prognosis is very different from benign cystic lesions. Surgical resection is the gold standard of management for CPAM for both pathological diagnosis and treatment. However, emergency surgery incurs a higher rate of surgical complications due to prolonged chest tube drainage and wound infection.\textsuperscript{20} Early pulmonary resection during the asymptomatic period is recommended due to almost no reported cases of surgical mortality and satisfactory long-term outcomes.\textsuperscript{3,20-22} In the current case, surgery was performed during the asymptomatic period with no surgical complications, and type 4 CPAM was diagnosed without malignant transformation. The long-term prognosis of early surgical resection will be evaluated in future studies.

In conclusion, CPAM in children is a rare disease with various clinical presentations and having the risk of future malignant transformation. Early pulmonary resection for asymptomatic CPAM is required and recommended to make a definite diagnosis and determine the prognosis. Malignancy and relapse are infrequent when radical surgery is not postponed.

\section*{REFERENCES}