

Not all that collapses is a pneumothorax: Congenital pulmonary Airway Malformation in disguise

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Abstract

Congenital Pulmonary Airway Malformation (CPAM) is a rare congenital malformation of the lung characterized by abnormal development of the terminal bronchioles, resulting in cystic or adenomatous lesions within the lung parenchyma, replacing normal lung tissue (1). It has an estimated incidence of 1 in 10,000-35,000 live births, with adult presentation being very rare, and often posing diagnostic challenges (1). We present the case of a 38-year-old man presenting with symptoms mimicking a tension pneumothorax, which was ultimately found to be congenital cystic lung disease. The patient presented with a 10-day history of flu-like symptoms followed by a 2-day history of productive cough and right-sided pleuritic chest pain. On admission, his vital signs were stable with an oxygen saturation of 93-94% on room air. A chest x-ray demonstrated a large right-sided upper lobe pneumothorax with significant mediastinal shift. A chest drain was inserted to manage the pneumothorax, however, there was minimal improvement in symptoms and inadequate drainage from the chest drain. This prompted a transfer to a larger hospital, where a computed tomography (CT) of the thorax was performed. The CT revealed extensive cystic lesions within the right upper lobe, with collapse and effacement of the right middle and lower lobes. These findings suggested an underlying congenital abnormality, most consistent with CPAM. Imaging also revealed drain insertion within a cyst prompting removal of the chest drain. In the days following the admission, a follow-up CT was performed which revealed an enlarged pneumothorax, which was subsequently managed with chest drain reinsertion. This was likely an iatrogenic pneumothorax due to the initial insertion of the chest tube within a cyst. Once the patient was clinically stable, he was discharged with further follow-up testing to assess pulmonary function and consideration for elective pneumonectomy. This case represents the complexities associated with a late presentation and diagnosis of CPAM. Literature supports surgical resection of affected lobes to prevent further complications such as recurrent infections and malignancies (2, 3). Additionally, this unique case highlights the importance of considering a broad range of differential diagnoses in cases of stable pneumothorax presentation, particularly to rule out cystic lung disease. Early use of CT imaging may be useful in differentiating pneumothorax from underlying bullous disease and guiding appropriate management.