Case Reports

Recurrent spontaneous pneumothorax associated with bronchial atresia

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Introduction

Bronchial atresia is an apparently rare congenital disorder in which a lobar or segmental bronchus fails to develop normally, resulting in discontinuity between the atretic segment and the remaining bronchial tree (1). This disorder was first described in 1953 by Ramsay and Byron (2) and by 1986, 86 cases had been reported in the English medical literature (3). No case has yet been reported in association with spontaneous pneumothorax.

Case Report

A 20-year old soldier presented initially with a left-sided spontaneous pneumothorax. An intercostal drain was inserted and his lung expanded fully. Four months later, the patient had a second episode of spontaneous pneumothorax which was again treated by insertion of an intercostal drain until the lung re-expanded. He had no relevant personal medical history or family history and had never smoked. On examination he was a muscular, healthy looking male with no signs of systemic illness or connective tissue disorder. Chest examination was completely normal (other than scars from drain insertions) with good air entry over both lung fields. Chest X-ray was initially reported as normal (Plate 1). Pulmonary function tests showed normal spirometry with FVC 82%, FEV₁ 82%, FEV₁/FVC 82% and PEF₅₀ 73% of predicted and with no change post-exercise. Lung volumes were normal.

Plate 1 Postero-anterior chest film revealing an area of 'emphysema' in the midzone of the left lung.

Computerized tomographic scan of the chest revealed a branching mass in the left perihilar region with distal hyperinflation and 'cystic' changes in the distribution of the superior segment of the left lower lobe (Plates 2 and 3). In retrospect, these findings were also apparent on plain postero-anterior chest films. High resolution CT showed no evidence of apical subpleural blebs. Ventilation-perfusion scanning demonstrated a matching defect with delayed wash-in and retention of the inhaled gas in the distribution of the superior segment of the left lower lobe. Fibre optic bronchoscopy and bronchography failed to demonstrate the presence of a bronchial opening to the superior segment of the left lower lobe. The patient was therefore diagnosed as having bronchial atresia of the superior segment of the left lower lobe.

Received 20 July 1995 and accepted in revised form 20 November 1995.

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Plates 2  Axial CT of the chest below the level of the carina. A branching mucus clot is seen in the apical segment of the left lower lobe (arrow) with surrounding 'cystic emphysema'.

Plate 3  Semicoronal CT of the chest demonstrating mucus clot (large arrow) surrounded by emphysema. Residual localized pneumothorax is seen at the apex and lateral aspect of the lung (small arrows).

Discussion

Congenital bronchial atresia is rare (1,4). A number of reviews have analysed the clinical, radiological and pathological features of this condition (1,3–6).

The precise pathogenesis of this disorder is not clear (7,8). During fetal pulmonary development, an insult to the integrity of a bronchus occurs resulting in complete obstruction at or near its origin with maintenance of intact bronchial architecture distal to the obstruction. Mucus secreted into the involved bronchi cannot drain and as water is absorbed, a thick solid mucus clot remains, usually in the shape of the bronchial tree (3). The atretic segment receives no transbronchial airflow but, during inspiration, air enters via collateral drift through the intra-alveolar pores of Kohn (9). This air cannot escape during expiration, resulting in air trapping and hyperinflation of the atretic lung.

As in the present case, most cases of bronchial atresia occur in young adults, in males more frequently than in females (64% vs. 36%) (3). In a review of 86 cases (3), 56% were asymptomatic and the diagnosis was made due to coincidental findings on routine plain chest X-ray. The most common symptom was recurrent infection in 21%. To the authors’ knowledge, no case of congenital bronchial atresia to date has presented as spontaneous pneumothorax.

While it is possible that bronchial atresia and pneumothorax occurred coincidentally in the present patient, the authors believe that this was not the case. It is thought that the bronchial atresia predisposed to recurrent spontaneous pneumothorax. There are several reasons for this assertion. Firstly, in bronchial atresia, the atretic segment is comprised of hyperinflated lung and bullae, some of which are subpleural. Because air can enter this segment during inspiration via the pores of Kohn, but cannot escape during expiration (due to closure of the collateral pores), air is effectively trapped at an increased pressure in the atretic lung segment thereby predisposing to rupture into the pleural space. Secondly, 91% of patients with primary spontaneous pneumothorax are smokers.
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and are asthenic. The present patient is a non-smoker and has no other risk factors for the development of spontaneous pneumothorax. Thirdly, CT demonstrated no evidence of subpleural blebs. This is in contrast to the findings in primary spontaneous pneumothorax in which apical blebs are present in 80% of cases (11). Finally, the pneumothoraces in the present patient occurred on the same side as the atretic segment.

The authors find it surprising that spontaneous pneumothorax is not a more frequent presenting symptom of bronchial atresia, and think it likely that this apparently rare entity is simply not diagnosed in some cases of spontaneous pneumothorax.

In congenital bronchial atresia, chest X-ray demonstrates hyperlucency and a perihilar mass in 80% of patients (3). Computerized tomography is diagnostic in this disorder (12) and there is little need for the performance of more invasive tests. The classical findings on CT scan are those of peripheral segmental or lobar hyperinflation surrounding a perihilar mass which is often branched and represents a mucus plug. In equivocal cases, bronchoscopy, bronchography and ventilation–perfusion scans may confirm the diagnosis. Whether CT scan of the chest should be routinely performed in patients after a single spontaneous pneumothorax is debatable. However, careful examination of a plain poster-anterior chest X-ray specifically looking for evidence of bronchial atresia will show the typical radiological abnormalities in most cases where this entity is present.

References