Unilateral Hyperlucent Lung due to Bullous Disease*

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A 18-year-old white woman was admitted complaining of right-sided chest pain of five days duration. The pain was sharp and acute in onset, pleuritic in nature, and felt diffusely throughout the right chest. She had low grade fever and generalized weakness. The patient denied shortness of breath, cough or bloody sputum. There was no history of asthma or pneumonia, and she was a nonsmoker. Her past medical history was unremarkable, but her father underwent a right pneumonectomy at age 16 for an undefined congenital problem.

Physical examination showed her vital signs to be stable. There was tracheal deviation to the left side. Her right chest was hyperresonant and her breath sounds were diffusely diminished without rales, rhonchi, or wheezes. Left chest examination results were unremarkable.

Admission chest x-ray examination revealed a shift of the mediastinum to the left and multiple air-filled bullae occupying almost all of her right chest cavity, except for a subsegmental infiltrate at the right base (Fig 1A, B).

Admission arterial blood gas levels included pH 7.40, PCO2 42, PO2 90, with 95 percent oxygen saturation while breathing room air.

During the evening after admission, she developed shortness of breath, and a right-sided chest tube was inserted. During the next several days she developed an enlarging right pleural effusion associated with tachycardia and a fall in hemoglobin level (Fig 2). The second chest tube was placed posteriorly but drained little fluid, so she underwent thoracotomy.

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Roentgenogram of blood, forming many of the bullae.

**FIGURE 2.** With a chest tube in place, there was rapid accumulation of blood, filling many of the bullae.

**Diagnosis: Congenital giant bullae of the right upper lobe**

At surgery, thoracic deep retroesophageal and retrosternal recesses from long-standing cystic disease in her right thorax were found. Several large right upper lobe bullae containing sero-sanguinous fluid were present. Compressed but otherwise normal middle and lower lobes were found. On expansion, these lobes barely filled 20 percent of her chest cavity. Although her bronchi were of normal caliber, her pulmonary vessels were vestigial. Right pneumonectomy was performed and she recovered uneventfully.

Cystic disease of the lung may be acquired or congenital. Congenital cysts originate from pulmonary remnants which separated from the main bronchial branches early in life. They are usually identified by a smooth epithelial lining. In most cases, congenital cystic disease affects both lungs. Unilateral cystic disease is most often due to lobar emphysema, bronchogenic cyst, congenital cystic adenomatoid malformation or unilateral hyperlucent lung (Swyer-James or Macleod’s syndrome).

Lobar emphysema is caused by maldevelopment of a lobar segmental bronchus. This anomaly causes gradual development of massive air trapping of the involved lobe, leading to compression of adjacent lung and mediastinal structures. Symptoms develop during the first six months of life in 81 percent of patients. Treatment is emergency lobectomy.

Bronchogenic cysts originate from embryonic lung bud tissue which separates from the remainder of the parenchyma before the bronchi are formed. Bronchogenic cysts usually present as middle or posterior mediastinal masses in children. Symptoms are secondary to infection, which occurs when a bronchial communication is present. Treatment is simple excision.

Cystic adenomatoid malformation represents abnormal bronchial maturation with overgrowth of mesenchymal elements. The affected lung tissue is enlarged, meaty and multicystic but lacks cartilaginous elements. Infants present with signs of respiratory distress due to air trapping and mediastinal compression. Older children usually present with chronic pulmonary infection. The recommended treatment is pneumonectomy.

Unilateral hyperlucent lung (Swyer-James or Macleod’s syndrome) presents with unilateral hyperlucent lung due to air trapping with associated bronchiectasis. There is hypoplasia of the pulmonary artery and hyposegmentation of the bronchi on the affected side. Peripheral pulmonary arteries are diminutive, and although the etiology is unclear, it is thought to be due to infection probably of viral origin and arrested pulmonary maturation in intrauterine life or early infancy. Many patients remain asymptomatic and when symptoms do occur, they are usually secondary to infection.

Proper management of congenital pulmonary cystic disease depends on the patient’s symptoms. Asymptomatic patients are observed, but when symptoms develop, removal of the cyst or cysts is indicated, as most patients respond poorly to attempts at percutaneous drainage. Resection is limited to diseased lung, conserving normal lung when possible. If remaining functional lung does not expand to fill the chest, pneumonectomy may be required.

**References**

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