Vanishing lung syndrome

Jing Wang PhD(c), Wei Liu PhD MD

A 19-year-old woman was admitted to the emergency room with acute pleuritic chest pain. She had no significant medical history, but was a current smoker. Computed tomography of the chest revealed a giant bulla occupying more than three-quarters of the left hemithorax, compressing the residual lung and displacing the mediastinum toward the opposite side (Figure 1). The images were consistent with vanishing lung syndrome (VLS). At thoracotomy, a giant bulla (at least 20 cm in diameter) was present in the left upper lobe. Bullectomy was performed without incident, allowing re-expansion of the underlying lung (Figure 2). Pathological examination showed an enlarged airspace with paraseptal emphysema on the margin and infiltration of inflammatory cells.

KEY LEARNING POINTS

- VLS is characterized by giant bullae that compress the underlying lung and cause it to seemingly disappear. The bullae are frequently asymmetric or unilateral, and limited to the upper lobes (1,2).
- VLS usually occurs in younger patients and may present with dyspnea, hemoptysis or chest pain (3).
- Paraseptal emphysema underlies most cases of VLS (2), which must be differentiated from other congenital and acquired cystic diseases of the lung.
- Differentiating VLS from pneumothorax is very important. On chest radiography, a pneumothorax will usually cause the lung parenchyma to collapse into a clump toward the hilum. In VLS, the compressed lung usually falls away from the hilum down toward the cardiophrenic angle. Computed tomography often shows fine septations within the bulla or a thin, barely perceptible wall; these may be quite difficult to identify on a standard chest radiograph.
- Treatment is surgical bullectomy (either by thoracotomy or thoracoscopy) and is indicated to relieve pulmonary compression and eliminate the risk of pneumothorax. In the unlikely event of acute distention causing respiratory failure and circulatory collapse, urgent intracavitary drainage should be considered.

REFERENCES
