

Answer: Vanishing lung syndrome

MCQ Explanation:

Vanishing lung syndrome (VLS), or idiopathic giant bullous emphysema, is a rare upper lobe pneumopathy that typically occurs in young, thin male smokers. VLS appears to be a distinct clinical syndrome, characterized by the presence of giant bullae in one or both upper lobes, occupying more than one-third of the hemithorax in young males with a history of tobacco and marijuana abuse.² High-resolution CT scans have been used in the preoperative assessment of patients with VLS to evaluate the extent and distribution of bullous disease.

Patients with pulmonary hemorrhage typically present complaining of cough associated with hemoptysis, fever, and dyspnea. Those with acute respiratory distress syndrome, or noncardiogenic pulmonary edema, would be critically ill with marked hypoxia, unlike this patient. Bronchiectasis is characterized by airway dilation with bronchial wall thickening on high-resolution CT, which is not apparent in this patient.

MCQ 2: Which of the following is the most appropriate next step in management?

Answer: Encourage smoking cessation

MCQ Explanation:

Given the progressive nature of VLS, worsening dyspnea and exercise intolerance are common features. Major complications can include spontaneous pneumothorax and bacterial superinfection. In addition to encouraging smoking cessation, limited bullectomy is the surgical treatment of choice in VLS.¹ The use of long-acting beta agonists or methylprednisolone plays no role in the management of VLS. Furthermore, pulmonary rehabilitation has proven to be of limited benefit in improving gas exchange, pulmonary function, dyspnea or exercise capacity.

References:

L. Roberts, C. E. Putman, et al., "Vanishing lung syndrome: upper lobe bullous pneumopathy," *Revista Interamericana de Radiología*, vol. 12, pp. 249–255, 1987.

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