CHEST

A 22-Year-Old Nonsmoker With Diffuse Cystic Lung Disease

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A 22-year-old previously healthy woman was evaluated in pulmonary clinic for shortness of breath and cough that had been slowly progressive over 3 months. She otherwise reported being fully functional and attended her college graduation a week prior to evaluation. She had no history of smoking, illicit drug use, connective tissue disease, or noxious exposures.

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Physical Examination Findings

Vital signs were as follows: BP, 120/80 mm Hg; pulse, 120 beats/min; respirations, 24 breaths/min; and oxygen saturation as measured by pulse oximetry on ambient air, 95% at rest and 85% on ambulation. Lung examination was significant for moderately diminished breath sounds and scattered rhonchi on both lung fields. There was no clubbing, cyanosis, or edema in her extremities.

Diagnostic Studies

Blood count, metabolic panel, and rheumatologic serologies were all unremarkable. Pulmonary function tests showed a severe restrictive defect with moderate diffusion impairment. Chest radiograph showed diffuse large bilateral cystic changes within both lungs (Fig 1). Subsequent chest CT scan revealed diffuse irregular thin- and thick-walled cysts with upper and middle lung zone predominance (Fig 2). A surgical lung biopsy was recommended for definitive pathologic diagnosis, but the patient deteriorated within 10 days of initial evaluation. She was admitted to the ICU for acute hypoxemic respiratory failure and pneumothorax requiring chest tube placement and mechanical ventilatory support. Empirical high-dose corticosteroids were initiated, but unfortunately she went into pulseless electrical activity arrest and was unable to be resuscitated. An autopsy was obtained. On microscopic examination of the lungs, there was dense cellular

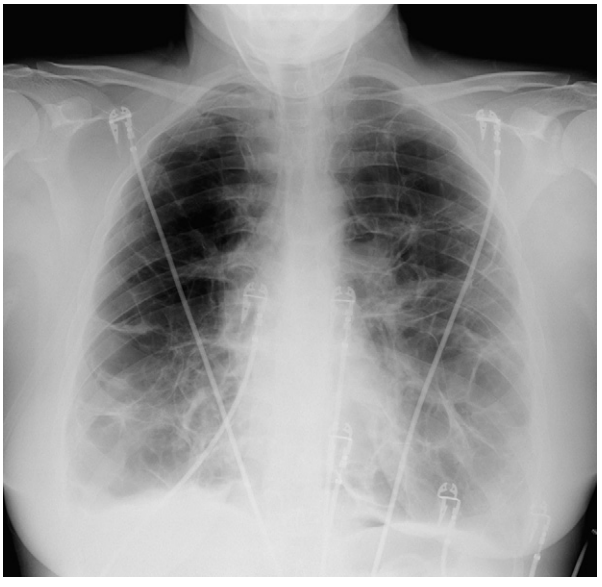

Figure 1 – *Chest radiograph showing diffuse large bilateral cystic changes within both lungs.*

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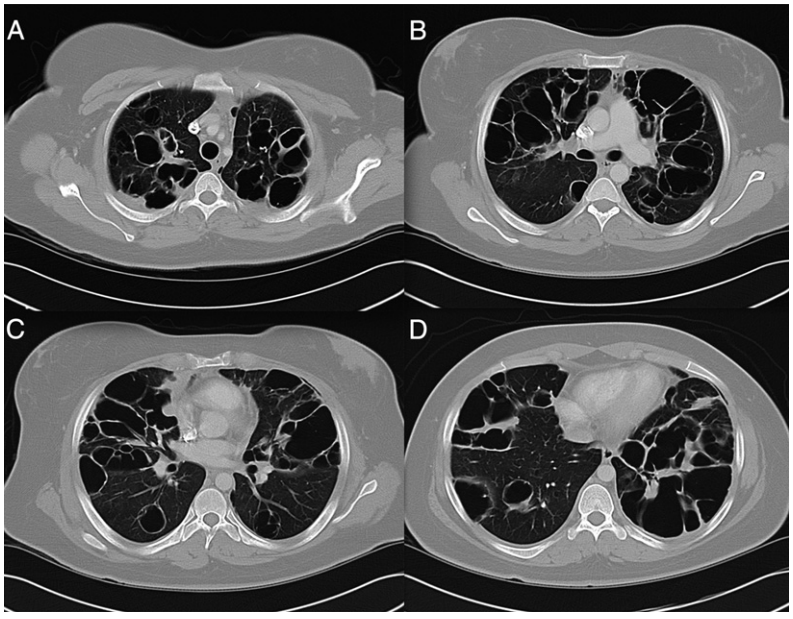

Figure 2 – *A-D, Serial transverse cuts of chest CT scan showing irregular thin- and thick-walled cysts, some large and consisting of several coalescent cysts, present in both lungs, with upper and middle lung zone predominance and relative sparing of the lung bases.*

inflammation within and surrounding bronchi and bronchioles. Immunohistochemical stains showed an inflammatory infiltrate strongly positive for CD1a and S-100 protein (Fig 3).

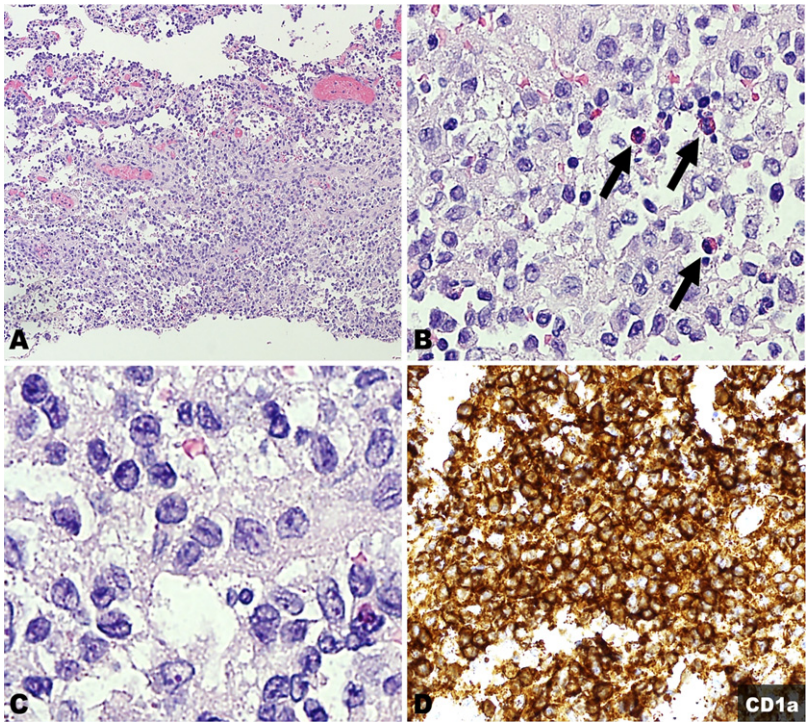

Figure 3 – *A, Low-magnification hematoxylin-eosin (H&E) stain of a pulmonary nodule consisting of mixed inflammatory cells including numerous cells with granular cytoplasm (magnification × 40). B, High-magnification H&E stain showing prominent scattered eosinophils (arrows) (magnification × 200). C, High-magnification H&E stain showing cells with abundant cytoplasm, indented nuclear membranes, and grooved "coffee bean" nuclei (magnification × 400). D, Cells are immunoreactive for CD1a (magnification × 100) and S-100 protein (not shown).*

What is the diagnosis?