A 34-year-old nonsmoking African-American woman with a history of childhood asthma presented to her internist with 25 h of right-sided chest pain and shortness of breath. The initial physical examination was remarkable for a respiratory rate of 28; heart rate, 140 bpm; and absent breath sounds on the right. A portable chest radiograph revealed a large right pneumothorax. She was treated with tube thoracostomy with resolution of the pneumothorax.

Two weeks after hospital discharge, a follow-up posteroanterior and lateral chest radiograph better revealed bilateral hilar adenopathy and diffuse increased interstitial markings (Fig 1). These coarse reticular infiltrates were more predominant at the bases. A computed tomography (CT) scan was performed that showed innumerable thin-walled cysts throughout the lungs again with a basilar predominance (Fig 2, top). They ranged in size from a few millimeters to about 2 cm. The intervening lung parenchyma was normal; no nodules were seen. Enlarged lymph nodes were seen throughout the mediastinum and in both hilar regions (Fig 2, bottom). There was only minimal adenopathy in the anterior mediastinum. There was no effusion or pneumothorax. The adenopathy was thought typical for sarcoid but the presence of diffuse pulmonary cystic disease is not a typical feature of sarcoid.

Pulmonary function tests revealed moderate airflow obstruction with normal lung volumes and diffusing capacity. Forced vital capacity (FVC) was 2.83 L (82% pred), the FEV₁ was 1.64 L (57% pred),

(Figure 1. Posteroanterior chest radiograph shows diffuse interstitial infiltrates and bilateral hilar adenopathy.)

(Figure 2. Top. Adenopathy is present in the azygos esophageal recess as well as both hilar regions. Bottom. Lung windows show multiple cysts throughout both lungs.)
Diagnosis: Coexistence of Sarcoid and Lymphangiomyomatosis

Pathologic evaluation of the lung revealed cystically dilated spaces lined by smooth muscle, with other areas of the lung showing random proliferation of smooth muscle along the alveolar septae, blood vessels, and lymphatic spaces. Immunohistochemical analysis revealed strong positivity to smooth muscle actin and intense nuclear staining for progesterone receptors and weak nuclear staining for estrogen receptors. This is typical of lymphangiomyomatosis (LAM). The lymph node was completely replaced by multiple conglomerate noncaseating granulomata consistent with sarcoid. Stains for organisms were negative. The lymph node did not show smooth muscle proliferation.

Pulmonary LAM is a rare disease seen almost exclusively in women of child-bearing age. The primary underlying disease is smooth muscle proliferation along the bronchioles, veins, and lymphatics, leading to pneumothoraces, hemoptysis, and chyloous effusions. On plain films of the chest, LAM typically appears as a diffuse reticulonodular infiltrate more predominantly in the bases. This progresses to a fine honeycomb pattern throughout the lungs, including the costophrenic angles. While lymph node involvement is commonly seen histologically, this is not a prominent feature radiographically. However, some smaller series have noted a significant amount of thoracic lymphadenopathy. The adenopathy that has been reported is often mild to moderate in size and extent.

On high resolution computed tomography (HRCT), the interstitial changes are better characterized as numerous, diffusely distributed, thin-walled cystic airspaces ranging in size from 2 mm to 5 cm with normal intervening parenchyma. The cyst walls range in size from 1-4 mm. Nodularity is not a feature of LAM, and septal thickening is minimal to absent. Lung volumes are usually increased or normal. Chyloous effusions and pneumothoraces are commonly seen.

Sarcoid may also affect women of child-bearing age. It is more common in African-Americans. It classically shows paratrabecular and hilar lymphadenopathy on roentgenographic examination, and it may also have parenchymal disease. This most often appears as nodular or reticulonodular infiltrates, but miliary or alveolar patterns are not unusual. Cystic changes in the lungs are associated with end-stage fibrosis and architectural distortion, most commonly seen in the mid to upper lung fields. On HRCT, small nodules are seen adjacent to the pleura and as irregular thickening of bronchial walls and vessels. Interlobular septa may be thickened. The distribution tends to be patchy rather than diffuse with normal intervening lung parenchyma. Honeycombing or lung cysts are usually subpleural. Cysts vary in size from 3 mm to 2 cm with thickened walls. Cystic changes are almost always associated with fibrosis distorting the central architecture of the bronchi and vessels.

Our patient had many of the features of LAM. She presented with a pneumothorax and dyspnea. Chest radiograph showed normal lung volumes rather than decreased volumes as would be expected with idiopathic pulmonary fibrosis or end-renal-stage sarcoid. CT of the thorax showed numerous thin-walled cysts diffusely throughout the lungs with a basilar predominance and normal intervening parenchyma. Eosinophilic granuloma, which can give a picture of lung cysts almost identical to LAM, often has an upper lobe predominance with sparing of the costophrenic angles. In addition, there are usually small nodules throughout the lung parenchyma in eosinophilic granuloma lung disease, which was not seen in this patient. Mediastinal lymphadenopathy, however, is an uncommon finding in LAM and is usually not as extensive in its involvement. On the other hand, extensive mediastinal lymphadenopathy is a
very common feature of sarcoid. Cystic changes in
the lungs are also seen with sarcoid, but these are a
result of fibrotic end-stage disease with associated
architectural distortion. There was no evidence of
fibrosis in this patient. The final diagnosis of both
LAM and sarcoid, while unusual, accounts for the
findings better than any single diagnosis. We believe
this is the second case report of these diverse findings
in the same patient.

In summary, a pattern of diffuse thin-walled cysts
in the lungs with normal intervening parenchyma
associated with mediastinal adenopathy is an unusual
presentation for LAM or sarcoid alone. Although a
single diagnosis is often sought to explain all CT and
radiographic findings the coexistence of these two
disease entities may be more common than previ-
ously recognized.

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