



UNIVERSITY of
ROCHESTER
MEDICAL CENTER

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DEPARTMENT OF IMAGING SCIENCES

Imaging Sciences Interesting Cases

CASE 16

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CLINICAL PRESENTATION: A 50-year-old female with smoking history presents with shortness of breath and decreasing functional ability. Evaluate for emphysema.

IMAGING FINDINGS: Plain chest radiographs often demonstrate increased interstitial markings in a reticular pattern. It is difficult to appreciate the cysts due to their nearly imperceptible thin walls. CT demonstrates extensive replacement of the lung parenchyma with thin walled cysts.

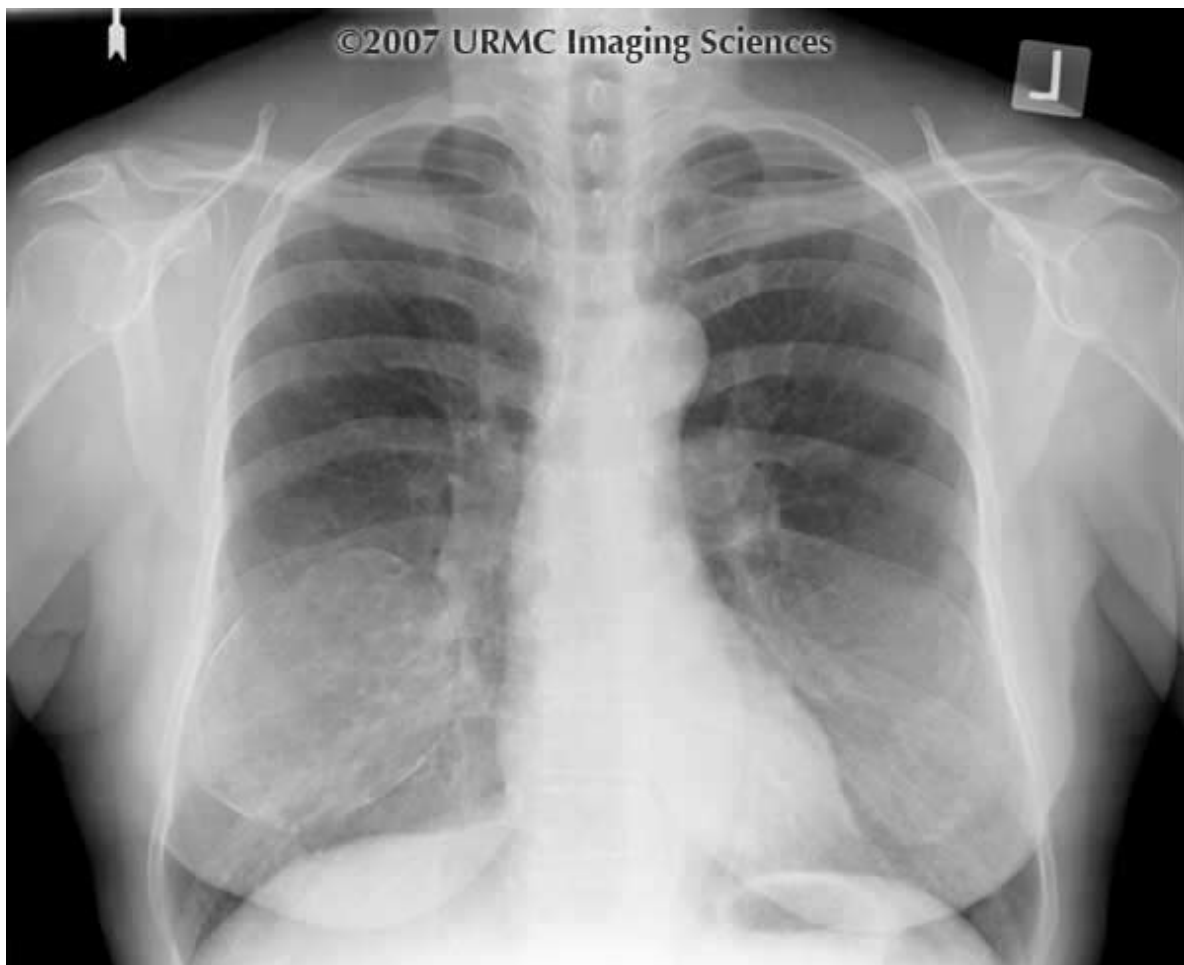


Figure 1: PA chest radiograph demonstrates increased reticular pattern of the lung parenchyma. Bilateral breast implants with calcifications are noted.

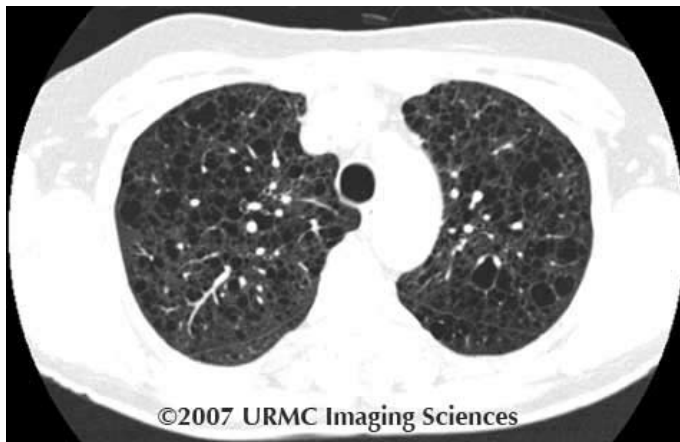


Figure 2: Axial HRCT of the upper lungs demonstrates extensive cystic changes bilaterally.

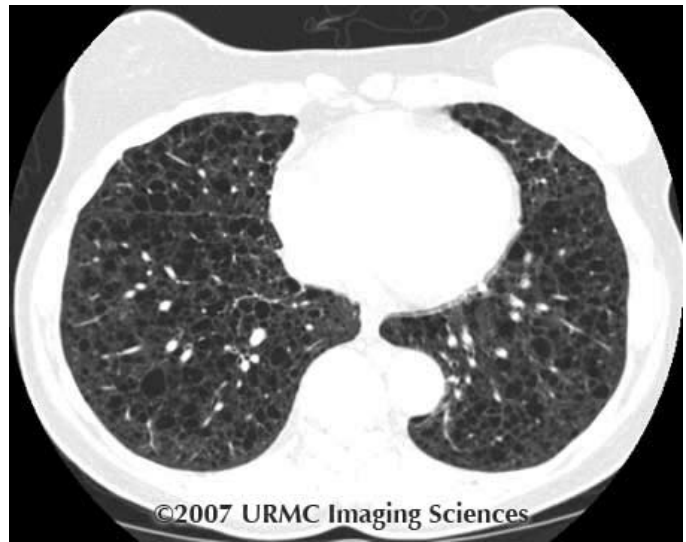


Figure 3: Axial HRCT of the lower lungs demonstrates similar extensive cystic lung disease as in the upper lungs.

DIAGNOSIS: Lymphangiomyomatosis

DISCUSSION: Lymphangiomyomatosis (LAM) is a rare disease characterized by progressive proliferation of atypical smooth muscle in the walls of the bronchi, bronchioles, alveolar septa, pulmonary vessels, lymphatic vessels, and pleura. The proliferation of the atypical muscle cells leads to air trapping and the development of emphysema and cystic air spaces. The parenchymal changes are progressive, eventually leading to diffuse honeycomb appearance with increased lung volumes. It is associated with young females of child-bearing age who are usually between 17 to 50 years of age which suggests that estrogen plays a role in the pathogenesis.

Identical clinical, radiologic, and pathologic pulmonary changes may be seen in 1% of patients with tuberous sclerosis. Although tuberous sclerosis affects both females and males, the pulmonary changes are described exclusively in women.

On chest radiographs, the pulmonary parenchymal changes may be seen as reticular, reticulonodular, military or honeycombing patterns. Other findings include pneumothorax and chylous pleural effusion secondary to involvement of the thoracic duct. On CT, LAM is characterized by bilateral diffuse and extensive cystic air spaces. In mild disease, the spaces measure less than 0.5 cm and those with more extensive disease, the cysts are greater than 1 cm. The cystic walls range from faintly perceptible to 2 mm thick and are surrounded by normal parenchyma.

Differential diagnosis includes emphysema, Langerhans cell histiocytosis, lymphocytic interstitial pneumonia, and pulmonary fibrosis.

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2. Bonelli FS, Hartman TE, Swensen SJ, Sherrick A. Accuracy of high-resolution CT in diagnosing lung diseases. *AJR Am J Roentgenol*. 1998 Jun;170(6):1507-12. [PubMed]