



Answer to Clinical Quiz

CXR Findings:

- *Reticular shadowing* throughout both lungs affecting mainly the middle and lower zones
- Thin wall cystic spaces
(These are better shown on CT of the lung:- Multiple thin wall cysts evenly distributed over both lungs. The lung parenchyma between the cysts appears normal.)
- *Increased lung volume* bilaterally
- Bilateral pleural effusion

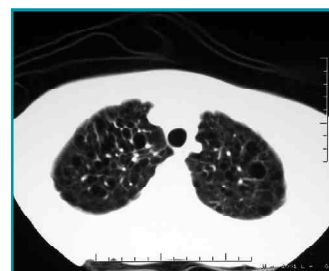


The roentgenographic pattern is basically that of fibrosing alveolitis but with an increased lung volume. In a *young female*, a diagnosis of lymphangio-leiomyomatosis or tuberous sclerosis should be suspected. The presence of pleural effusion is suggestive of the former diagnosis. Radiographically, the two conditions are very similar. Pneumothoraces can occur in both conditions but chyloous effusion is more typical of lymphangio-leiomyomatosis.

DIAGNOSIS: *Lymphangio-leiomyomatosis*

Lymphangio-leiomyomatosis (lymphangiomyomatosis, myomatosis) is an uncommon pulmonary abnormality characterized pathologically by a diffuse proliferation of smooth muscle within airway walls, parenchymal interstitium and lymphatics. It is a disease primarily of young and middle-aged women.

The presenting complaint is usually shortness of breath, sometimes in association with pneumothorax or a history of repeated hemoptysis. Unilateral or bilateral chylothorax is a common manifestation.



Radiologically, the appearance of the lungs in lymphangio-leiomyomatosis is indistinguishable from that of fibrosing alveolitis, with the exception of the effects on lung volume: characteristically, alveolitis manifests as progressive loss of lung volume whereas in lymphangio-leiomyomatosis, the volume tends to be increased. The basic pattern is coarse reticulo-nodular and tends to be generalized. The late pattern is one of honey-combing. High resolution CT scans can show the well-defined cystic spaces ranging from a few mm to 5cm and distributed diffusely throughout the lungs. The wall of these cysts are characteristically thin and uniform.

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