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Lymphangiomyomatosis: Radio-logic-Pathologic Correlation¹

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Lymphangiomyomatosis (LAM) is an uncommon interstitial lung disease that exclusively affects women, usually during their reproductive years. LAM is characterized pathologically by abnormal proliferation of LAM cells in the lungs and in thoracic and retroperitoneal lymphatics. Thirty-three cases of LAM were reviewed retrospectively for clinical and radiologic findings. Twenty-eight (85%) of 33 women (aged 21-62 years; mean, 37.5 years) were symptomatic. Radiographs ($n = 32$) demonstrated reticular opacities in 21 (66%) patients, large lung volumes in 17 (53%), pleural effusion in 14 (44%), and pneumothorax in 13 (41%). High-resolution CT ($n = 15$) and conventional CT ($n = 3$) showed 2–5-mm bilateral thin-walled cysts in all patients and cysts that were 6-12 mm or larger in patients with severe lung involvement. CT depicted diffuse lung involvement by cysts in nine (50%) patients, relative sparing of lung apices in seven (39%), and relative sparing of lung bases in two (11%). Pleural effusion and pneumothorax were seen at CT in four (22%) and three (17%) patients, respectively. Four cases of tuberous sclerosis complex-associated LAM (TSC-LAM) (women aged 27-50 years; mean, 35.7 years) were similarly reviewed. Three (75%) were symptomatic. Radiographs ($n = 4$) demonstrated reticular opacities in three (75%) and large lung volumes in two (50%). All high-resolution CT ($n = 3$) and conventional CT ($n = 1$) studies showed 2–5-mm bilateral thin-walled cysts and cysts that were 6-12 mm or larger in two patients with severe lung involvement. Pleural effusion and pneumothorax were demonstrated at CT in three (75%) and two (50%) patients, respectively. LAM and TSC-LAM affect symptomatic women who often exhibit reticular opacities and large lung volumes at radiography and bilateral uniform small thin-walled cysts at CT. Large (>12 mm) cysts occur in patients with severe cystic lung involvement. Pneumothorax and pleural effusion are common associated findings.

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