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# Lymphangioleiomyomatosis: a radiological diagnosis

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## DESCRIPTION

Lymphangioleiomyomatosis (LAM) is a rare disease which predominantly affects young females and is commonly found in association with tuberous sclerosis.<sup>1</sup> The clinical features result from progressive cystic destruction of the lungs and the accumulation of LAM cells within the lungs and axial lymphatics.<sup>2</sup> The gold standard for the diagnosis of LAM is a tissue biopsy which shows nodular infiltration by abnormal smooth muscle cells, termed LAM cells. Not all patients with LAM require tissue biopsy for a definitive



**Figure 1** Computerised axial tomography scan of the chest showing multiple well-defined thin-walled bilateral lung cysts consistent with lymphangioleiomyomatosis.

diagnosis as the disease has a characteristic computerised axial tomography (CAT) appearance in the majority of cases. The author reports a 34-year-old non-smoking African-American female who presented to the clinic with progressive shortness of breath (Figure 1). CAT scan chest showed multiple well-defined thin-walled bilateral lung cysts which are randomly distributed throughout the lungs with normal intervening lung parenchyma consistent with LAM. There were no infiltrates, worrisome nodules, or pleural effusion. LAM needs to be differentiated from other chronic pulmonary diseases which present with a cystic lung appearance. Langerhans cell histiocytosis which closely mimics LAM is predominantly characterised by nodules and cyst walls of variable thickness and tends to spare the basal parts of the lung near the costophrenic angles.<sup>3</sup> Centrilobular emphysema can be differentiated from LAM by absence of well-defined walls and the distribution of the vessels relative to the cystic spaces where the vessels can cross cystic spaces.<sup>3</sup>

**Competing interests** None.

**Patient consent** Obtained.

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