A Case of Sporadic Pulmonary Lymphangioleiomyomatosis

Sir

Pulmonary lymphangioleiomyomatosis (LAM) is a rare lung disease characterised pathologically by cyst formation and multifocal, nodular proliferation of immature smooth muscle and perivascular epithelioid cells (LAM cells). The following illustrates such a case.

A 33 year old lady, smoker with a 15 pack year history presented to her GP with intermittent right sided pleuritic chest pain and exertional breathlessness when walking up a slight hill. She denied any cough, wheeze, night sweats and weight loss. Her past history include hypothyroidism and liver haemangioma. She has a family history of ischaemic heart disease and works in a launderette. Examination and CXR was normal. Due to an increase in frequency of her intermittent pleuritic chest pain, a CT thorax was organised by her GP which picked up a 4mm cyst in the right midzone of her lung. She was then referred to the respiratory OPD and a follow up CT thorax done showed new ground glass attenuation in both lung. During her work up for an insterstitial lung disease, the only positive finding was a mildly raised anti double stranded DNA but after being assessed by the rheumatologist, there was no evidence of the presence of Lupus. Her pulmonary function test was normal and transbronchial biopsies performed came back as non-diagnostic. She was then referred for a VATS biopsy and thus was diagnosed with pulmonary lymphangioleiomyomatosis. The term sporadic LAM is used for patients with pulmonary LAM not associated with tuberous sclerosis complex. The majority of patients with sporadic LAM are young women of childbearing age. Approximately 30 percent of women with tuberous sclerosis complex are affected by pulmonary LAM. Nearly all patients are symptomatic at presentation, and approximately 70 percent report dyspnoea. Potential complications of LAM include pneumothorax, hemoptysis, chylothorax, chyloperitoneum, chylopericardium, and development of lymphangioleiomyomas of the lymph vessels. High resolution computed tomography of the lung is obtained in all patients suspected of having LAM. Characteristic HRCT findings include diffuse, small, thin-walled cysts. LAM management is largely aimed at easing symptoms (e.g. supplemental oxygen, bronchodilators, pulmonary rehabilitation) and preventing complications (e.g. pleurodesis after a spontaneous pneumothorax, avoiding estrogen therapy).
For patients with progressive, moderate-to-severe lung impairment, clinical trials have shown that sirolimus does provide a beneficial effect. Some patients with LAM eventually require lung transplantation because of progressive respiratory failure. Selection criteria and outcomes are similar to those for other chronic lung diseases. The estimated median transplant-free survival time for pulmonary LAM is 29 years from symptom onset and 23 years from diagnosis. Reflecting on this case, although there is actually no cure for LAM, organisation such as the LAM foundation provides education and support to such patient. Furthermore this foundation also promotes ongoing research for improved treatment and eventually cure for people with LAM.

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References