Lymphangioleiomyomatosis: A Rare Indication for Lung Transplantation

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Lymphangioleiomyomatosis is a rare pulmonary disease with a poor prognosis. Lung transplantation, however, remains the only treatment option for patients with advanced disease. We describe a patient with LAM who was referred to lung transplantation.

Patient Description

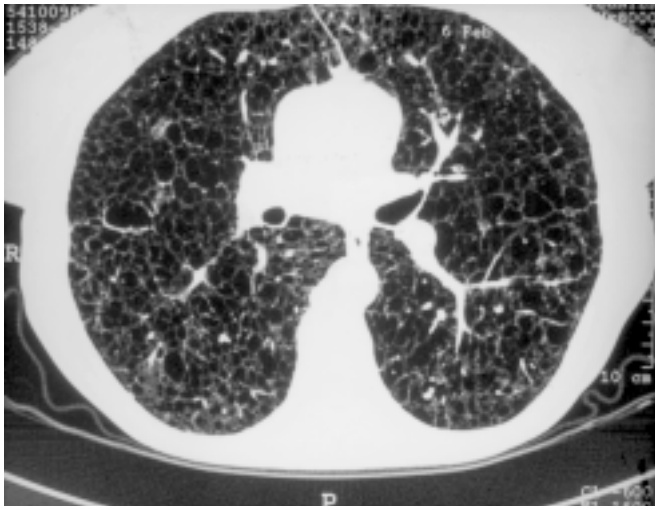
A 45 year old woman was referred to our pulmonary clinic for possible lung transplantation. She had slowly progressive dyspnea and non-productive cough during the last 6 years. Her medical history revealed recurrent episodes of pneumothorax that had necessitated pleurodesis with bleomycin 2 years ago, and right nephrectomy due to angiomyolipoma 20 years ago.. Physical examination revealed a respiration rate of 24/minute and normal breath sounds. Pulmonary function showed severe airway obstruction with forced expiratory volume in 1 second of 18% of the predicted (0.49 L), total lung capacity of 7.3 L (150% of the predicted) and diffusing lung capacity of 33% of the predicted (18 ml/min/mmHg). High resolution computed tomography of the thorax [Figure] demonstrated numerous thin walled air-filled cysts diffusely distributed throughout all lung zones. The typical high resolution CT, the clinical presentation and the history of renal angiomyolipoma confirmed the diagnosis of lymphangioleiomyomatosis. Trial of treatment with medroxyprogesterone acetate failed. Following clinical deterioration and decline of pulmonary function, the patient was referred for lung transplantation.

LAM = Lymphangioleiomyomatosis

Comment

LAM is a rare pulmonary disease resulting from proliferation of a unique smooth muscle-like cell, the LAM cell, involving the small airways, the pulmonary microvasculature and the lymphatic structures. Most patients are premenopausal women who seek medical attention for worsening dyspnea or pneumothorax, chylous fluid collections, or a complication of renal angiomyolipoma [1,2]. Despite the presence of hormonal receptors on the LAM cell, therapeutic manipulation of hormone status has not had much success in halting the disease tendency to progress [3]. For patients with advanced LAM, lung transplantation offers the only hope at present [4]. The pleurodesis that the patient underwent in the past, however, may complicate the LTX. The 1 and 2 year survival rates after LTX for LAM were 69% and 58%, respectively. These rates compare favorably with those for LTX overall [4]. In summary, LAM is a rare pulmonary disease with poor prognosis. LTX, however, remains the only treatment option for patients with advanced disease.

LTX = lung transplantation



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