Lung Transplantation for Lymphangioleiomyomatosis: Fukuoka University experience

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Abstract

Background: Lympahngioleiomyomatosis（LAM）is a rare cystic disease with abnormal proliferation of smooth muscle-like cells, so called LAM cells, in the lung interstitium that results in progressive decline of lung function. Lung transplantation is the definitive treatment for patients with end stage respiratory failure of LAM.

Patients and Methods: Patients with LAM undergoing lung transplantation at The Fukuoka University Hospital during the year of 2005-2015 were retrospectively reviewed.

Result: Fifty four patients including 6 patients with LAM registered as candidates for brain death lung transplantation during the study period. Four of the 6 patients with LAM underwent brain-dead-donor single lung transplantation and 1 patient underwent living-donor one lobe lung transplantation. All patients were females with mean age of 38.5 years. The mean time from diagnosis to lung transplantation was about 8 years. There was one late death with fungal infection and the rate of successful rehabilitation was 80%. The results of lung transplantation with end-stage LAM were highly favorable than other end-stage lung disease.

Conclusion: Lung transplant is a feasible treatment for patients with end-stage LAM, characterized by young without other organ compromise.

Key words: Brain-dead-donor lung transplantation, Living-donor lung transplantation, Lymphangioleiomyomatosis