Experience of using mTOR inhibitor in lung transplant recipients with lymphangioleiomyomatosis

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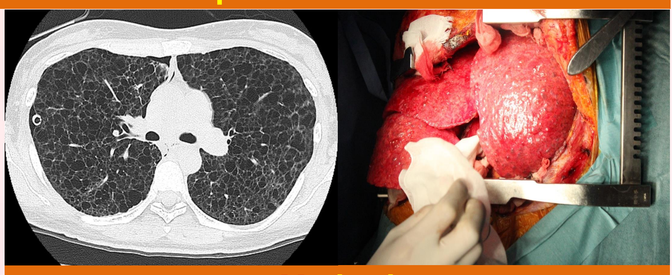
Background and Purpose

Lymphangioleiomyomatosis (LAM) is a rare pulmonary disease, sometimes resulting in lung transplantation (LTx). Drug therapy using mTOR inhibitor (sirolimus /everolimus) can alleviate tumor progression and decline in lung function in LAM patients, but can also cause serious complications like dehiscence in bronchial anastomosis after LTx.



Method

We retrospectively reviewed 14 LTx cases with LAM at Okayama University Hospital.

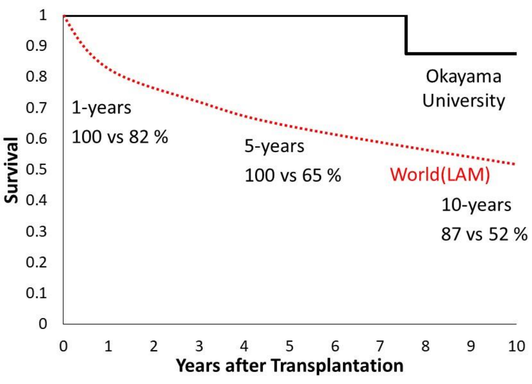
Result

	Age at LTx / sex	LTx type	Double / Single	Pre LTx mTOR inhibitor	Post LTx mTOR inhibitor	Starting months	Bronchial complication after LTx	Other Complication after LTx
Case 1	23F	LDLLTx	Double	—	sirolimus	72 months later	—	Recurrence of LAM
Case 2	30F	LDLLTx	Double	—	Sirolimus	96 months later	—	Recurrence of LAM
Case 3	30F	CDLTx	Double	—	Everolimus	120 months later	—	Progression of renal AML
Case 4	46F	CDLTx	Single	—	—	—	—	Pneumothorax of native lung
Case 5	38F	CDLTx	Single	—	Sirolimus	120 months later	—	Progression of LAM
Case 6	32F	LDLLTx	Double	—	—	—	—	Recurrence of LAM
Case 7	32F	LDLLTx	Double	—	Sirolimus	3 months later	—	chylothorax
Case 8	40F	CDLTx	Single	—	—	—	—	chylothorax
Case 9	27F	CDLTx	Double	—	Sirolimus	36 months later	—	Chylothorax, Progression of renal AML
Case 10	22F	CDLTx	Double	—	Sirolimus	36 months later	—	Progression of renal AML
Case 11	39F	CDLTx	Double	—	Sirolimus	3 months later	—	chylothorax
Case 12	23F	CDLTx	Double	Everolimus	Everolimus	6 months later	—	Progression of renal AML
Case 13 (Re-LTx case2)	45F	CDLTx	Double	Sirolimus	Sirolimus	6 months later	—	—
Case 14 (Re-LTx case5)	48F	CDLTx	Single	Sirolimus	Sirolimus	6 months later	—	—

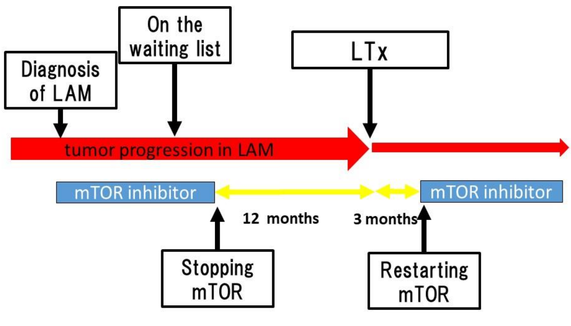
LDLLTx: living donor lobar LTx, CDLTx: cadaveric LTx, AML: angiomyolipoma

- 14 cases, 12 female recipients including two re-transplant, were identified.
- Four underwent LDLLTx and ten underwent CDLTx.
- Three of 14 underwent single LTx and suffered late complications in their native lung (over-inflation/ lung cancer/ aspergillus).
- There were no complication of bronchial anastomosis.

Overall Survival (LAM=14)



Our strategy



The post-LTx survival rate was 100% at 5 years and 87.5% at 10 years with a median follow-up of 110 months (range 1-203 months).

In our strategy, mTOR inhibitor was stopped about one year before LTx (or on listing) and was started after 3 months post LTx if necessary.

Relevant Financial Relationship Disclosure Statement

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Conclusion

- LTx is a feasible therapeutic option for patients with advanced LAM.
- mTOR inhibitors can provide optimal management in LTx recipients especially with progression of recurrent pulmonary LAM/ angiomyolipoma and complicated chylothorax.