

[ PICTURES IN CLINICAL MEDICINE ]

# Rapid Shrinkage of Retroperitoneal Lymphangiomyoma after Sirolimus Initiation

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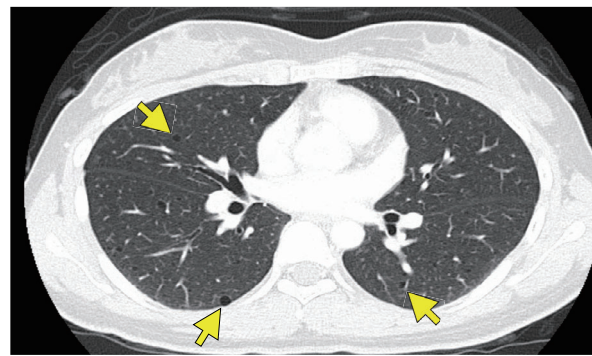
**Key words:** lymphangiomyomatosis, retroperitoneal, lymphangiomyoma, sirolimus, short term

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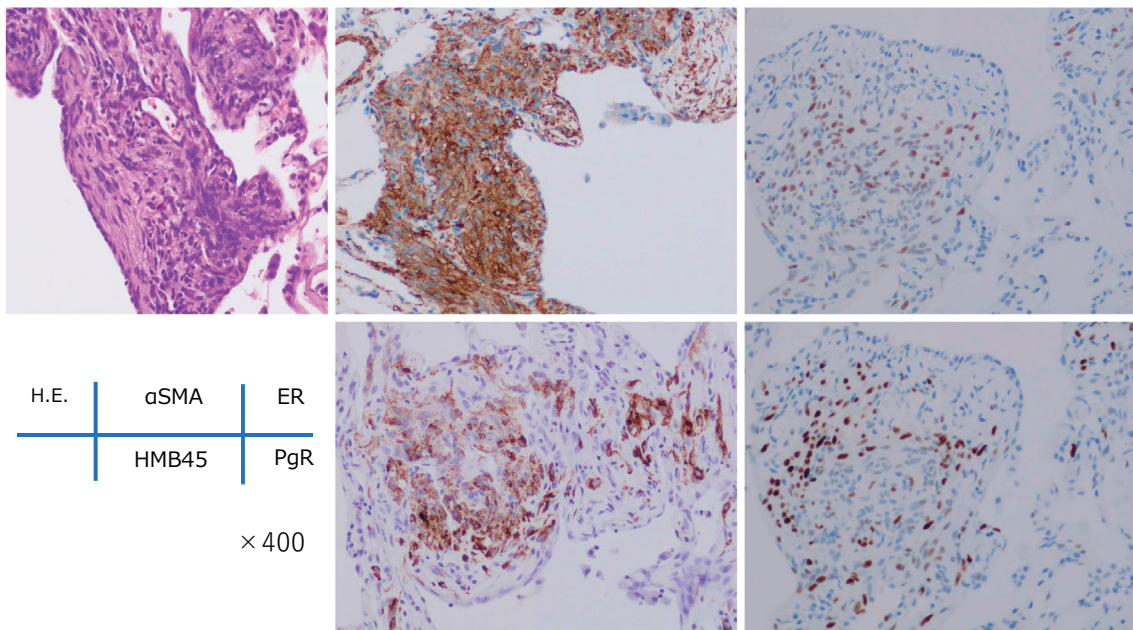
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Picture 1.



Picture 2.

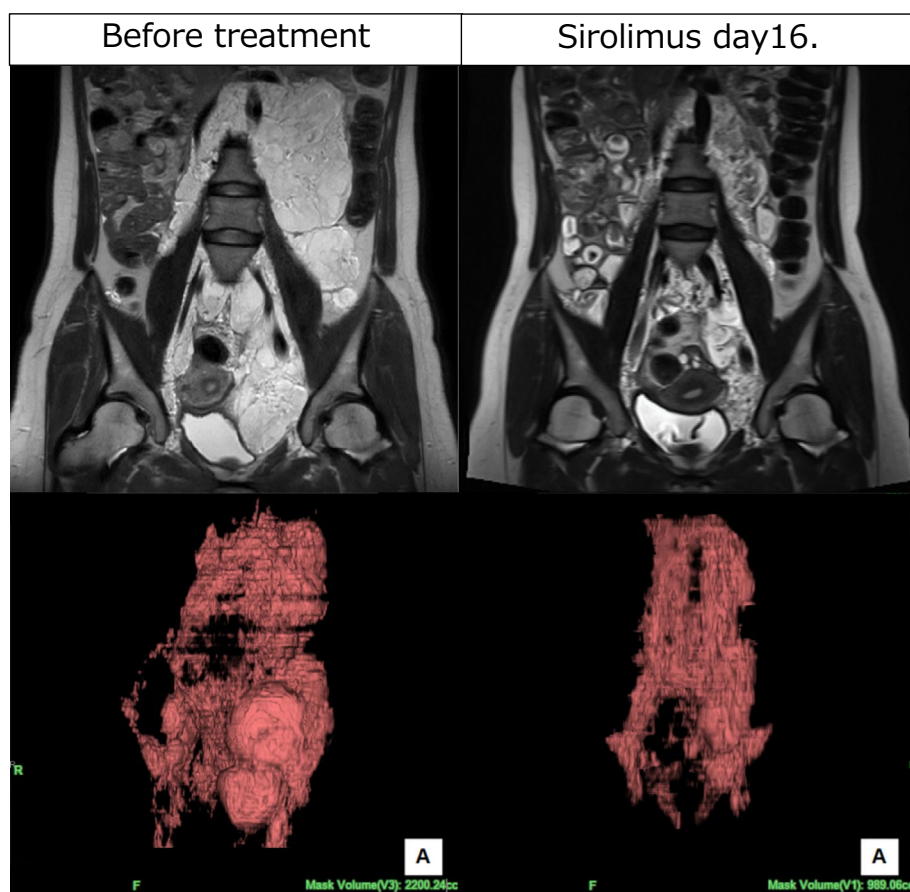


Picture 3.

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**Picture 4.**

A 29-year-old Japanese woman presented with a left inguinal bulge and tenderness. Computed tomography revealed colossal cystic lesions in the retroperitoneal space (Picture 1) and cystic lesions with thin walls in the bilateral lungs (Picture 2). We performed a lung biopsy, and the pathological examination revealed hyperplasia of spindle-shaped cells with Hematoxylin and Eosin (H&E) staining,  $\alpha$ -smooth muscle actin ( $\alpha$ SMA), melanoma antigen gp100 (clone name, HMB45), estrogen receptor (ER) and progesterone receptor (PgR) (Picture 3). The histological diagnosis was lymphangiomyomatosis. The abdominal lesion was regarded as retroperitoneal lymphangiomyoma, although a biopsy was not performed because of the dissemination risk. Treatment with sirolimus (2 mg/day) was started; the abdominal symptoms gradually alleviated. Abdominal magnetic resonance imaging at day 16 revealed a significant reduction in the volume of the retroperitoneal lymphangiomyoma (2,200 to 980 cc) (Picture 4). The trough level of sirolimus in blood at day 18 was 5.2 ng/mL. The effectiveness of sirolimus against retroperitoneal lymphangiomyoma

has been described (1, 2), but there have been no cases in which it was effective within as short a period of time as in our patient. Sirolimus may be effective for obtaining rapid symptomatic improvement of retroperitoneal lymphangiomyoma.

**The authors state that they have no Conflict of Interest (COI).**

#### References

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