Rapid Shrinkage of Retroperitoneal Lymphangioleiomyoma after Sirolimus Initiation

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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 eosin (H.E.) staining, α-smooth muscle actin (αSMA), melanoma antigen gp100 (clone name, HMB45), estrogen receptor (ER) and progesterone receptor (PgR) (Picture 3). The histological diagnosis was lymphangioleiomyomatosis. The abdominal lesion was regarded as retroperitoneal lymphangioleiomyoma, 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 lymphangioleiomyoma (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 lymphangioleiomyoma 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 lymphangioleiomyoma.

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References