Table 2: Heterogenous disease manifestations in three female patients with lymphangioleiomyomatosis and one patient with pulmonary Langerhans cell histiocytosis.

Patient / Radiological findings	Patient 1 / Figure 1A	Patient 2 / Figure 1B	Patient 3 / Figure 1C	Patient 4 / Figure 1D
Age	39-year-old	42-year-old	36-year-old	43-year-old
Previous history	never-smoker sleep-related breathing disorder	occasional smoker for the last 4 years	valvuloplasty for congenital stenosis of the pulmonary valve in childhood	40 pack-years
Symptoms	stress dyspnea spontaneous pneumothorax	hemoptysis and recurrent pulmonary infections over 2 years	severe dyspnea	progressive cough for 4 months negative results for ANA, ENA, anti-ds-DNA, ANCA
VEGF-D* ng/mL	0.522	1.06	3970	N/A
Procedures	VATS for lung biopsy atypical resection with talc pleurodesis.		VATS and wedge biopsy Explorative laparotomy prior to abdominal lymphadenopathy complicated by chyloperitoneum.	
Histopathology findings	nodular proliferate and the cyst wall that strongly expressed HMB-45 due to LAM	LAM was diagnosed without histological confirmation	detected cells expressing HMB-45 LAM confirmed	Bronchial lavage cytological findings: approximately 2% CD1a-positive counted cells due to PLCH
Recommendations	Follow-up evaluations	Follow-up evaluations	Sirolimus, target serum VEGF-D levels less than 10 ng/mL	abstinence from tobacco histological confirmation if progress

LAM: lymphangioleiomyomatosis, PLCH: pulmonary Langerhans cell histiocytosis, HMB-45: Human Melanoma Black-45, *: Reference for VEGF-D value <0.8 ng/mL; this result did not exclude a LAM diagnosis. VEGF-D: vascular endothelial growth factor D, VATS: video assisted thoracoscopic surgery, ANA: antinuclear antibodies, ENA: extractable nuclear antigen, anti-ds-DNA: anti-double stranded DNA, ANCA: anti-neutrophil cytoplasmic antibodies.