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Title: Characterization of 76 patients with lymphangioleiomyomatosis from a Brazilian reference center

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Body: Introduction: Lymphangioleiomyomatosis (LAM) is a rare disorder affecting childbearing women. There was no previous epidemiological study from South America characterizing subjects with LAM. Objectives: To describe the clinical features, quality of life (SF-36 questionnaire), pulmonary function tests (PFT) and VEGFD data, and also six-minute walk test (6MWT) performance of LAM patients followed in a Brazilian reference center. Methods: Data from 76 women with LAM followed in the Pulmonary Division of the University of Sao Paulo from 2008 to 2011 were reviewed. Results: The mean age was 42 ± 11 years. The average age at diagnosis was 38 ± 9 years, whereas the mean time from first symptom to diagnosis was 23 months. The diagnosis of LAM was confirmed by tissue biopsy in 88% of cases. 14% of patients had tuberous sclerosis, 50% renal angiomyolipoma, 61% previous pneumothorax and 15% previous chylothorax. Dyspnea was a complaint of 69% of subjects and 20% were ex-smokers. Impaired quality of life was found, with worse scores in physical and emotional domains. Mean FEV_1 and D_LCO were, respectively, 74±26%pred and 65±28%pred. The most common abnormalities on PFT were obstructive pattern (58%), reduced D_LCO (53%) and air trapping (22%). The mean distance walked was 493 ± 118 m, while the mean Borg dyspnea score and the minimum SpO_2 at end 6MWT were, respectively, 3 ± 3 and 88±9%. Dessaturation ≥4% was found in 53% of patients during 6MWT. In 60% of subjects, VEGFD serum level was increased. Conclusions: In a Brazilian sample of LAM patients, besides the typical results in PFT, quality of life impairment, increased VEGFD serum levels and dessaturation during 6MWT were also important findings.