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Title: Lymphangioleiomyomatosis: Functional and morphological characteristics in 36 patients

Dr. Marina 13344 Kameneva kmju@mail.ru MD [1], Prof. Ivetta 13345 Dvorakovskaya kmju@mail.ru [1] and Dr. Olga 13346 Baranova dr_baranova@mail.ru MD [1]. [1] Institute of Pulmonology, Pavlov's State Medical University, St. Petersburg, Russian Federation, 197 089 .

Body: Aim: To investigate the functional and morphological features of LAM. Material and methods. The 36 cases of LAM had been diagnosed in women (m.age 41.9±12.6 yr). The CT, pulmonary function tests, conventional histological techniques, immunohistochemistry (IHC) tests with monoclonal antibodies to actin, desmin, HMB-45, estrogen (Ers) and progesterone (Prs) receptors were performed. Results: The microscopic picture of early stages of the disease was characterized by multicentric foci of small smooth muscle proliferates in the alveolar septa, the walls of blood vessels with deformation of their lumen. IHC study confirmed the smooth muscle nature of proliferates. There was mild airway obstruction (FEV1 69.6±18.5%Pred). With progression of the disease, tendency to increase and fusion of foci with cyst formation, destruction, violation of the lung tissue architectonics was determined that significantly changed lung mechanics. Along with the smooth muscle proliferates desmoplastic was observed. Desmin expression indicates the relationship between desmoplastic and smooth muscle proliferates and depends on the stage of morphogenesis. The sharp change in the parameters characterizing airway obstruction (FEV1 45.2±16.9%Pred), increasing the RV (RV/TLC 176±16.9%Pred) and decreased lung elasticity (CR 0.19±0.07 kPa/l) were the functional markers of this process. A reduction of DLco was observed in all patients. Conclusion: A reduction of lung elasticity is a significant functional marker of changes of normal lung tissue architectonics. The character and severity of functional impairment in LMA are determined by morphological changes in the lungs and should be considered in determination of treatment strategy.