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Title: Difficulties of diagnosis of pulmonary embolism (PE) in patients with lymphangioleiomyomatosis (LAM)

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Body: Aim: to analyze causes of PE and to describe of clinical, radiological and hemodynamic characteristics of PE in LAM patients. Material and methods. Six cases of PE in patients (pts) with morphologically confirmed LAM were studied. The average age was 52.0±4.7 years. Pts were subjected to clinical and radiological (CT, HRCT, CT-angiography, SPECT with Tc99m) and echodopplercardiography (EDC) researches. Results. The main cause of PE was deep venous thrombosis (DVT) in 5 pts. All pts had predisposing factors for venous thromboembolism. Clinical symptoms were: increasing of dispnea, appearing or increasing haemoptysis, chest pain, cough, tachypnea, tachycardia. The combination of CT-angiography and SPECT revealed signs of PE in all LAM pts. In patient with growing uterus leiomyoma we found signs of PE and the additional intravascular mass. The analysis of EDC data in LAM pts with PE revealed: increasing of systolic pulmonary artery pressure (45.74±2.06 mm Hg), decreasing of E/A right ventricular diastolic filling ratio (1.28±0.05) and E/A left ventricular diastolic filling ratio (1.17±0.21). In all pts deterioration of PFTs data was diagnosed: increase of obstructive limitation (average FEV1 40 % pred.) with significant hyperinflation (average RV 200 % pred.), sharp reduction of diffusing capacity (average DLCOss 36 % pred., average DLCOsb 26 % pred.) and blood gas disorder. Conclusion. The main cause of PE in LAM pts is DVT with the presence of predisposing factors for venous PE. The modern radiologic methods allow to diagnose PE and to suggest that additional intravascular mass in one patient can be associated with LAM cells metastasis leading to formation of blood clots.