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Title: The role of vascular endothelial growth factor D in diagnosis of polycystic lung diseases

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Body: The study was aimed at determining the prospective discriminative value of serum vascular endothelial growth factor D (VEGF-D) concentration in reliable diagnostic differentiation of patients with lymphangioleimyomatosis (LAM) from subjects with other polycystic lung diseases (PLD). Methods: Serum VEGF-D level was measured by enzyme-linked immunoassay in 37 patients with PLD including 8 women with LAM, 27 patients (13 women and 14 men) with pulmonary Langerhans cell histiocytosis (PLCH), 1 woman with lymphocytic interstitial pneumonia (LIP) and 1 woman with tuberous sclerosis (TSC) without LAM. Results: Serum VEGF-D concentration was significantly higher in LAM patients [median 1979 pg/ml (interquartile range (IQR) 468-3897pg/ml) in comparison to subjects with PLCH [median 534pg/ml (interquartile range (IQR) 126-791pg/ml], TSC without LAM-387pg/ml and LIP-233pg/ml. The VEGF-D cut-off level of 790 pg/ml provided significantly discriminative consistency for LAM diagnosis with specificity of 96.3% and sensitivity of 87,5%. Conclusions: A serum VEGF-D level exceeding 790 pg/ml in a woman with typical cystic changes on high-resolution computer tomography (HRCT) scans is diagnostically specific for LAM.