THE CASE OF LYMPHANGIOLEIOMYOMATOSIS, ASSOCIATED WITH TUBEROUS SCLEROSIS COMPLEX

O. K. Yakovenko, R. B. Sydor, O. V. Solovey, T. N. Galkevych, I. V. Kornievych

Abstract

Lymphangioleiomyomatosis (LAM) — is a rare lung disease, affecting mainly women of child-bearing age. LAM is characterized by progressive infiltrative growth of smooth-muscle-like cells (LAM-cells), leading to cystic destruction of lung parenchyma, and airways, blood and lymphatic vessels obstruction.

In about 15 % of all cases LAM is associated with tuberous sclerosis complex (TSC) — a hereditary systemic dysplasia, caused by compromised development of ectodermal germ layer, leading to combined skin tuberous growths and brain, eye, heart, kidney and lung lesions.

The report presents brief characteristics, diagnosis criteria of TSC and LAM, as well as the clinical case of association of these two conditions.

Key words: tuberous sclerosis complex, lymphangioleiomyomatosis.

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Oleh K. Yakovenko
Volyn regional clinical hospital
Chief of pulmonology dpt.
MD, PhD
Lutsk, 21, Grushevsky ave., volyn_pulmo@ukr.net