MULTIPLE CYSTIC LUNG DISEASES (PART II)

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Abstract

The review presents the analysis of current literature data on multiple cystic lung diseases (MCLD). This group of conditions is defined by the presence on chest CT-scan of the multiple round-shape parenchymal lesions, clearly demarcated from normal parenchyma with < 2 mm thick wall.

There are three major causes of MCLD — lymphangioleiomyomatosis (LAM — sporadic or associated with tuberous sclerosis complex), Langerhans-cell histiocytosis (LCH) of lung and, in last time — Birt-Hogg-Dube syndrome, associated with FLCN gene mutations. Other causes of MCLD may include lymphoid disturbances, especially observed in Sjögren's syndrome, amyloid-free immunoglobulin deposition disease, infections, malignancy, hypersensitivity pneumonitis.

Due to limited role of chest X-ray in detection of thin-wall cysts of small and medium size, previously a morphology of cysts was precisely analyzed by pathologists at autopsy. Breakthrough occurred 15-20 years ago when CT-scan (mainly of high resolution) became available. Currently, highly specific CT features of cysts morphology are well described in LAM and LCH, allowing early diagnosing of these conditions without surgical lung biopsy. Due to a development of CT technique the list of MCLD has expanded significantly. Beside other benefits of emerging lung CT-scan technology in diagnosing of LCH, LAM and other conditions, there is a possibility of objective evaluation of treatment effectiveness and the rate of progression of the disease.

The first part of the review contains a characteristics of two major diseases — lymphangioleiomyomatosis and Langerhans cell histiocytosis of lung. The second part of the review describes Birt-Hogg-Dube syndrome, MCLD in lymphoid disorders, MCLD of an infectious nature, congenital cystic diseases and other nosological forms.

Key words: multiple cyst lung diseases, Birt-Hogg-Dubé syndrome, MCLD in lymphoid disorders, MCLD of an infectious nature, congenital cystic diseases.

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