MULTICYSTIC LUNG SYNDROME: RARE CASES OF REGRESSION

V. K. Gavrysyuk, A. V. Strafun, N. M. Musienko, N. V. Pendalchuk, N. D. Morska

Abstract

Multicystic lung syndrome (MLS) is defined by a presence on chest computed tomography of multiple, round parenchymal attenuations, having distinct borders with normal parenchyma and < 2 mm thick wall. Pneumothorax, usually relapsing, and respiratory failry are the major clinical manifestations of MLS. CT findings can make certain contribution into recognition of the disease but diagnosis is largely based on appropriate symptoms, extrapulmonary signs of systemic disease and/or congenital disease.

There has been described three major causes of MLS — lymphangioleiomyomatosis — LAM (sporadic or associated with tuberal sclerosis complex), pulmonary Lanherhans cell histiocytosis (LCH) and in recent time — Birt-Hogg-Dube) syndrome, associated with gene FLCN mutations.

Other causes of MLS include lymphoid disorders of lung, especially in Sjögren's syndrome, non-amyloid immunoglobulin deposition disease; infections; malignancy, mainly sarcoma metastasis; desquamative interstitial pneumonia, hypersensitivity pneumonitis, cryptogenic organizing pneumonia with atypical cavity forming regression etc.

The cases of regression of MLS in other diseases are rare. In most cases according to literature data regression is documented based on functional improvement which is not confirmed by CT data.

The article presents the brief description of two clinical cases of MLS with demonstration of positive CT changes.

Presented cases of MLS regression confirm the effectiveness of glucocorticosteroid treatment, which help to restore bronchial passability by means of reduction of compression caused by pulmonary infiltration in patient with organizing pneumonia and involution of granulamatous process in patient with hypersensitivity pneumonitis. In both cases the valvular mechanism of cyst genesis has been interrupted.

Key words: multicystic lung syndrome caused by organizing pneumonia and hypersensitivity pneumonitis, glucocorticosteroid therapy, regression.

Ukr. Pulmonol. J. 2023;31(4):35–40.

Volodymyr K. Gavrysyuk
National institute of phthisiology and pulmonology named after F. G. Yanovskyi NAMS of Ukraine
Chief of Department of Interstitial Lung Diseases
Corresponding member of NAMS of Ukraine
Doctor of medicine, professor
10, M. Amosova str., 03680, Kyiv, Ukraine
Tel./fax: 38 044270-35-59, gavrysyuk@ukr.net