PULMONARY ALVEOLAR PROTEINOSIS: EARLY AND LONG-TERM TREATMENT OUTCOMES

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Abstract

The literature data regarding the morbidity, etiology, pathogenesis and clinical symptoms, diagnostics and treatment of rare interstitial lung disease pulmonary alveolar proteinosis (PAP) — are presented in current review. Own experience of diagnosing and management of 18 patients with PAP, including the analysis of long-term (from 1 to 7 years) treatment outcomes is summarized.

In 15 patients the diagnosis of PAP, established using high resolution computed tomography (CT), was verified by pathohistology examination of lung bioptates (trasbronchial biopsy — 13 cases, open lung biopsy — 2 cases). In 3 cases the diagnosis of PAP was confirmed solely by highly specific CT-semiotics and exclusion of alternative diagnoses.

All patients were treated using broncho-alveolar lavage (BAL) at fiber-optic bronchoscopy, performed under local anesthesia.

Upon discharge the following course of disease was assessed based on annual examination of patients, which included CT scan and functional tests. The duration of follow-up period was from 1 to 7 years: 1–3 years — 6 patients, 3 years — 3 patients, 4 years — 4 patients, 5 years — 1 patient, 6 years — 2 patients, 7 years — 2 patients.

In 3 patients the progression of disease, which required new BAL, has been registered in average 2 years after the first hospitalization. In 15 cases there was a regression of disease, maintained by additional BAL, performed on as needed basis (7 patients).

The results indicate that high resolution CT scan of chest is a reliable method of diagnosing, which competes with histological examination of lung bioptate. A key sign of PAP on CT scan is the presence of "crazy paving" pattern with "geographical" distribution of the lesions across lung parenchyma.

To the contrast from classic treatment lavage of lungs, bronchoscopic BAL appears to be safer and better tolerated by the patients, since it requires no general anesthesia with separate intubation of both lungs, minimizing the risk of development of such complications as severe hypoxemia, seizures, pneumothorax, lower respiratory tract infections. At the same time bronchoscopic BAL allows to achieve fast regression of PAP and reliably control the disease.

Key words: pulmonary alveolar proteinosis, treatment pulmonary lavage, early and long-term outcomes.

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