PULMONARY ALVEOLAR PROTEINOSIS: THE CASE OF SUBTOTAL PULMONARY PARENCHYMA INVOLVEMENT

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Abstract

Pulmonary alveolar proteinosis (PAP) is a rare pulmonary disease, characterized by the intraalveolar accumulation of surfactant-like lipoprotein substance. There are three forms of the disease: autoimmune (formerly known as idiopathic), secondary and congenital PAP.

The disturbances of surfactant metabolism (protein-lipoid complex secreted by type II pneumocytes) play the crucial role in the development of the disease. Surfactant decreases a surface tension of alveoli and prevents its collapse at the end of the expiration and takes part in protection against infections as well.

Surfactant is inactivated through the transformation into the surface-inactive aggregates, which are mainly absorbed by type II pneumocytes. The rest of surfactant are catabolized by alveolar macrophages (AM). This process is regulated by cytokines, especially granulocyte-macrophage colony-stimulating factor (GM-CSF) being the most important of them. Disturbances of interrelation between GM-CSF and cellular receptors blocks the signals to effector cells for surfactant deactivation. This is resulted in excessive accumulation of surfactant in alveoli.

General lavage of the lung is a standard method of symptomatic management of PAP. Contemporary pathogenesis treatment uses two approaches — increasing the clearance of surfactant by means of exogenous GM-CSF administration and decreasing the level of GM-CSF antibodies using the rituximab and plasmapheresis.

Due to the use of broncho-alveolar lavage (BAL) 5-year survival rate of th patients reaches 95 %. The mortality depends on the area of lung lesion and reversely correlated with the timely diagnosis and treatment.

We report the case of subtotal lung parenchyma lesion associated with severe respiratory failure in 46 years old patient. Three series of BAL (5 procedures in each with 1-2 months interval) were performed using the limited volume of lavage fluid due to the severity of the patient's condition. The results was an improvement of clinical condition, resolution of respiratory failure, significant reduction of the size of the lesions, improved lung transparency on CT scan, restoration of lung diffusion and total lung capacity.

Conclusion. Therapeutic BAL is an effective method of PAP management. The outcome of clinical case presented and our many years experience confirm not only the effectiveness of conventional BAL with reduced lavage volume application but also a safety of fiberoptic bronchoscopy, especially in patients with subtotal involvement of lung parenchyma.

Key words: pulmonary alveolar proteinosis, subtotal pulmonary parenchyma involvement, pulmonary bronchoalveolar lavage, efficiency.

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