題名:Effect of Lung Lavage on Oxygenation in a Patient with Pulmonary Alveolar Proteinosis

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摘要:肺蛋白沉著症(pulmonary alveolar proteinosis)爲一不明致病因且罕見的疾病，可能由於先天顆粒白血球巨噬細胞刺激因子(granulocyte-macrophage colony-stimulating factor)失常、吸入有害物質、血液疾病或肺部感染等原因，造成肺泡腔內沉積過量的表面活性磷脂及脂蛋白，且合併肺間質發炎或纖維化，使病患肺順應性下降並影響氣體交換，而產生呼吸困難、咳嗽、運動喘及發紺等症狀，全肺灌洗是目前唯一有效的治療方法但無法根治。本個案被診斷爲肺蛋白沉著症已兩年並接受數次全肺灌洗治療，此次因感冒而惡化，藉由其治療過程探討全肺灌洗的方法、其對氧合功能的成效及病患後續居家呼吸照護的重要性。

Pulmonary alveolar proteinosis is a rare syndrome of unclear etiology, which is characterized by intra-alveolar accumulation of lipoproteinaceous material with minimal interstitial inflammation or fibrosis and resulted in decreasing lung compliance and impairing gas exchange. It may relate to congenital abnormality of granulocyte-macrophage colony-stimulating factor (GM-CSF) pathway activity, inhaled toxic substance, and certain hematological or pulmonary infectious diseases. The patients usually present short of breath, cough, progressive exertional dyspnea, and cyanosis. Whole lung lavage is the only effective treatment but unable to cure the disease. This patient was diagnosed as PAP for 2 years and received several times of whole lung lavage. The acute exacerbation was induced by influenza this time. In this report, we will discuss the procedures of whole lung lavage and its effects on oxygenation, and the importance of long term respiratory care for PAP patients.