中文題目：於低細胞性骨髓化生不良患者以反覆肺炎表現之肺泡蛋白沉積症

英文題目：Unexpected Pulmonary Alveolar Proteinosis developed in a Patient who Had Hypocellular Myelodysplastic Syndrome and Presented with Recurrent Pneumonia

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Introduction

Pulmonary Alveolar Proteinosis (PAP) is a rare lung disease, characterized as diffuse pulmonary disease where lipoproteinaceous material, primary surfactant and surfactant apoproteins are accumulated in distal airway and alveoli. The clinical course of the disease is variable, ranging from respiratory failure to self-limited, and open lung biopsy is still the gold standard to make the diagnosis. Herein, we presented a rare case who had hypocellular myelodysplastic syndrome and presented with recurrent pneumonia in the past years. She underwent a surgical biopsy and finally PAP was diagnosed.

Case Presentation

A 61-year-old housewife had past medical histories of pancytopenia. She received bone marrow biopsy and it revealed hypocellular marrow (cellularity was about 10-15%, hematopoietic elements composed of trilineage of hematopoietic cells. Positive for immunohistochemical stain with CD34(+) and CD117(+) in myeloid cells.) Hypocellular MDS was diagnosed.

She had several episodes of pneumonia with productive cough, fever and elevated C-reactive protein since June 2012. Chest computed tomography disclosed multiple consolidation patches, ground-glass patches, reticular opacities, nodular opacities and cavities noted in both lungs in Oct 2014. Furthermore, bronchoscopy revealed tracheobronchitis with scanty sputum in the airway. Video-assisted thoracoscopic surgery (VATS) wedge resection of right middle lobe and right upper lobe was performed and the PAP was diagnosed according to the pathologic characteristics of the specimens.

Unfortunately, she suffered from a respiratory failure because of pneumonia progression in Nov 2014. Whole lung lavage was performed twice but no clinical benefit and she died of respiratory failure and septic shock in Feb 2015.

Discussion

PAP has three distinct forms, congenital (mutation in genes encoding for surfactant proteins and granulocyte-macrophage colony-stimulating-factors), secondary (to immunosuppression, hematological cancers or toxic inhalations) and acquired or idiopathic (autoimmune disease, associated with GM-CSF autoantibodies). Secondary PAP (sPAP) often relates to haematologic malignancy and MDS is the most common one such as our present patient. We present a rare case who had underlying hypocellular MDS and developed PAP which presented with recurrent pneumonia. We share this case and make a literature review about the prevalence pathogenesis, diagnosis, treatment and outcome of MDS related PAP.