115 年奇美醫院胸腔內科臨床病例討論會

1 時間：115 年 07 月 21 日 PM: 4:00-5:00
2 課程活動題目:Alveolar proteinosis
3 主持人：柯獻欽
4 地點： 奇美醫學中心 10 樓空橋討論室
5 聯絡人：黎安騏（06-2812811 #57132）
6 摘要：

Pulmonary alveolar proteinosis can be understood as a syndrome of altered surfactant homeostasis, leading to a pathologic accumulation of surfactant. Because surfactant homestasis is complex, there are many potential points of disruption- overproduction of phospholipids byt type II pneumocytes, impairment of clearance of phospholipids by macrophages, or both. As a consequence of this disruption, floccular periodic acid-Schiff (PAS) positive proteinaceous and lipid rich material is deposited within the alveoli. There are two forms of alveolar proteinosis described: a "primary" or idiopathic form occuring in the absence of an identifiable associated disease or exposure (possibly related to an immunologic disturbance), and a "secondary" form provoked by or associated with another condition. Secondary alveolar proteinosis has been described in association with: 1) infections of the lung; 2) hematologic malignancies and other conditions altering a patients immune status (AIDS); and 3) exposure to inhaled mineral dusts (silica, aluminum, titanium) and chemicals (insecticides).

Patients generally present between the age of 20-50 years with progressive shortness of breath and mild, usually non-productive cough. Weight loss, fatigue, and malaise may also be present. However, up to 30% of patients will be asymptomatic even with florid CXR findings. Males are affected more than females (2 to 4:1). Physical exam findings are usually non-specific, but clubbing (25%) and cyanosis (21%) can be seen. An elevated serum lactate dehydrogenase level is the most common associated laboratory abnormality. Patients also have elevated levels of lung surfactant proteins A and D in both the serum and BAL fluid. The intrapulmonary shunt fraction while breathing 100% oxygen is typically elevated (average 20%). Most patients have restrictive lung disease with decreases in total lung capacity, forced vital capacity, forced expiratory volume in 1 second, and diffusion capacity for carbon monoxide. The disorder usually follows a variable course with exacerbation's and remissions. Spontaneous improvement or recovery occurs in 25-50% of

cases. Previously up to 30% mortality within several years had been reported, but the actual mortality may be less than 10%. Treatment is whole lung bronchopulmonary lavage to remove abnormal surfactant which can lead to improved oxygenation as alveoli re-expand, but this is not required in all cases. After treatment, marked improvement in the disease and radiographic findings are usually apparent. About 80% of patients respond favorably to lung lavage. Lung transplant can be used in selected cases, but the disorder can recur in the transplanted lung.

Concomitant superinfection can occur, classically with *Nocardia*, but more recently described with *Mycobacerium avium-intracellulare* and *pneumocystis carinii*

X-ray:

CXR: On CXR there are small acinar nodules which may coalesce to form areas of consolidation- typically in a "bat-wing" configuration which may mimic pulmonary edema, although the heart size is normal and pleural effusions are typically absent. The involvement is occasionally asymmetric or unilateral. Lymphadenopathy and pleural effusion are rare (although an effusion can be seen following lavage). Changes typically resolve slowly over weeks to months. Later there is diffuse reticulogranularity and changes consistent with interstitial lung disease.

Computed tomography: HRCT demonstrates areas of patchy ground glass opacification or consolidation and peripheral or central ill-defined nodular opacities. Smooth interlobular septal thickening is also seen and intralobular interstitial thickening is common in association with the ground-glass opacifications. The septal thickening produces an underlying polygonal pattern referred to as "crazy paving." The "crazy paving" pattern can also be seen in cases of mucinous bronchoalveolar cell carcinoma, lipoid pneumonia, ARDS, and drug-induced pneumonitis.