European Respiratory Society
Annual Congress 2012

Abstract Number: 4517

Publication Number: P3620

Abstract Group: 1.5. Diffuse Parenchymal Lung Disease

Keyword 1: Pulmonary hypertension **Keyword 2:** Interstitial lung disease **Keyword 3:** Lung injury

Title: Clinical study of pulmonary hypertension complicating pulmonary Langerhans cell histiocytosis

Ms. Fei 27521 Li lftutu@yeah.net [1], Prof. Huaping 27522 Dai daihuaping@sina.com MD [1], Ms. Liying 27523 Peng ply0501075@163.com [1], Prof. Mulan 27524 Jin kinmokuran@163.com MD [2] and Prof. Chen 27525 Wang cyh-birm@263.net MD [3]. [1] Department of Respiratory and Critical Medicine Care, Beijing Chao-Yang Hospital, Beijing, China, 100020 ; [2] Department of Pathology Medicine, Beijing Chao-Yang Hospital, Beijing, China, 100020 and [3] Beijing Hospital, Health Ministry, Beijing, China, 100730 .

Body: Objective To study the clinical features of pulmonary hypertension (PH) complicating pulmonary langerhans' cell histiocytosis (PLCH). Methods Medical records of 11 PLCH patients were reviewed from June 2006 to June 2011. Results 4 of the 11 PLCH patients presented PH with more severe clinical presentations. The major symptoms were laboring dyspnea and diminished exercise capacity (NYHA functional class IIIor IV), which were complicated with the sigh of right heart failure. The cystic change and pulmonary artery / main artery > 1 were predominate on chest high-resolution computed tomography. Right heart enlargement was also found. As to pulmonary function presentation, the patients displayed severe carbon monoxide diffusing capacity impairment and significant hypoxemia. The pulmonary artery systolic pressure were highly increased. Besides the typical histopathological features presented, involvement of both arteries and veins was also observed. Oxygen and symptomatic therapy were essential in the part of clinical treatment. 3 of the 4 PLCH-PH received corticosteroids or chemotherapeutic agents, but the effect was not ideal. Only one case of PLCH-PH have a stable condition. Conclusion Pulmonary hypertension is a common complication of PLCH and seems to predict a poor prognosis. It is necessary to derive an early diagnosis and prevention.