Title: High-resolution computed tomography pattern reflects pathophysiologic difference in acute exacerbation of idiopathic interstitial pneumonia

Body: Background: Acute exacerbation (AE) of idiopathic pulmonary fibrosis (IPF) (AE-IPF) shows poor prognosis and high-resolution computed tomography (HRCT) pattern (diffuse pattern) is a significant prognostic factor in AE-IPF (Akira M, 2008 AJRCCM). Aims: We retrospectively compared clinical findings between AE-idiopathic interstitial pneumonias (IIPs) with diffuse pattern and non-diffuse pattern to clarify the difference in pathophysiology and clinical behaviors of the two patterns. Subjects: Seventy cases of AE-IIPs were diagnosed according to the guidelines of Japanese respiratory society in 2004 and classified with radiological and/or pathological findings into two groups: IPF (n=55) and non-IPF (n=15). Methods: Clinical findings were compared between two HRCT pattern (diffuse pattern and non-diffuse pattern). Prognostic significance of each clinical factor was evaluated by univariate and multivariate Cox proportional hazard analysis separately in diffuse and non-diffuse pattern. Results: In diffuse pattern, white blood cell and C-reactive protein was significantly higher and serum Krebs von den Lungen (KL)-6 and P/F ratio was significantly lower than those in non-diffuse pattern. Multivariate Cox proportional hazard regression analysis showed that lower immunoglobulin-G (IgG) and higher KL-6 is significant predictor of a bad prognosis in diffuse pattern. In non-diffuse pattern higher ratio of increase in KL-6 after the AE/KL-6 before AE is a good prognostic factor. Conclusions: Serum KL-6 has a different prognostic value between two radiological patterns in AE-IIPs. The pathophysiology of the two types of AE-IIPs might be different.