European Respiratory Society
Annual Congress 2013

Abstract Number: 4892

Publication Number: P5123

Abstract Group: 1.5. Diffuse Parenchymal Lung Disease

Keyword 1: Interstitial lung disease **Keyword 2:** No keyword **Keyword 3:** No keyword

Title: Analysis of multicentric Castleman's disease (MCD) with pulmonary lesions

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Body: Background MCD is a systemic lymphoproliferative disease that often involves pulmonary lesions. Few studies have combined analyses of clinical, radiological, and pathological (CRP) data relating to this rare disease. Aim We aimed to identify the CRP features of this disease via a multicenter retrospective study. Methods We enrolled 40 patients who had MCD with pulmonary involvement throughout Japan. Results The average age at diagnosis was 47 years (men, 13; women, 12). The patients were admitted with symptoms of cough (48%), dyspnea (24%), and fever (18%). Laboratory findings revealed elevated serum levels of IL-6 (24.1 pg/mL), KL6 (736.7 U/mL), CRP (6.68 mg/dL) and IgG4 (394.1 mg/dL). Common radiologic findings included presence of ground glass attenuation (96%), centrilobular nodules (92%), septal thickening (72%), bronchovascular thickening (64%), and cysts (64%). From the pathological point of view, all specimens showed lesions characteristic of marked lymphoplasmacytic accumulation with lymphoid follicles and lymphatic distribution of foci of dense fibrosis. Small cystic lesions were found in 4 lung samples, but no obstructive vasculitis was observed. The mean IgG4+/IgG+ plasma cell ratio was 22%. A pulmonary function test revealed progressive obstructive airflow limitation. Steroid therapy was successful in preventing progression in just 33% of cases, but tocilizumab was successful in 67%. Three patients died due to respiratory failure with progression of diffuse cystic lesions. Conclusion The clinical course of pulmonary involvement of MCD varies from stable disease to progressive obstructive impairment. Tocilizumab appears to be more effective than steroid therapy in controlling the disease.