FRI0613

THERAPEUTIC STRATEGIES AND SURVIVAL IN PATIENTS WITH INTERSTITIAL PNEUMONIA WITH AUTOIMMUNE FEATURES

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Background: Recently the term "interstitial pneumonia with autoimmune features" (IPAF) has been proposed to identify patients with interstitial lung disease and autoimmune characteristics, not fulfilling the criteria for specific connective tissue diseases (CTD). Until now, only few data are available about the clinical and serological features of IPAF patients, their survival and the possible evolution in a CTD.

Objectives: Aim of the study was to investigate the therapeutic choices in IPAF patients, their efficacy and safety.

Methods: Fifty-two patients (mean age at diagnosis 65.5±11.0 years, female/male ratio 29/23) were consecutively enrolled and prospectively followed for 45±31.6 months.

Data about therapies, disease onset, serological, clinical and therapeutic features, pulmonary function tests and high-resolution computed tomography were periodically repeated. A worsening of lung function was defined as a reduction of 10% compared to baseline of forced vital capacity (FVC) and diffusion lung capacity of CO (DLCO).

Results: An immunosuppressive therapy was prescribed in 15 patients (namely cyclophosphamide in 4, mycophenolate mofetil in 6 and azathioprine in 6, respectively), while 6 patients were treated with anti-fibrotic therapies (pirfenidone or nintedanib). Thirty-three patients taken corticosteroids, associated to other drugs in 18 patients, and alone in 15. Finally, no therapies were prescribed in 16 patients. At the end of follow-up FVC remained stable in 35% of patients, worsened in 50% and increased in 15%; DLCO remained stable in 25.8% of patients, worsened in 58.1% and increased in 16.1%.

Mean survival was 94.2±8.5 months, and no differences were recorded according to the kind of therapy, while survival was significantly associated to the lung function at baseline (FVC and DLCO were confirmed to be significantly associated to death at multivariate analysis).

The treatment was generally well tolerated. In 7/15 patients treated with immunosuppressive therapy was recorded at least an adverse event, responsible for the treatment discontinuation only in 3 patients.

Conclusion: The therapeutic strategy in IPAF patients reflects the heterogeneity of the disease. Waiting for specific trial in this population, treatment is empirical and based on the baseline features of the patients. Despite well tolerated both anti-fibrotic and immunosuppressive therapies seem to not influence the evolution of the disease.

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CLINICAL AND LABORATORY FEATURES OF MACROPHAGE ACTIVATION SYNDROME IN PATIENTS WITH ADULT STILL'S DISEASE

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Background: Adult Still's Disease (ASD) sometimes develops macrophage activation syndrome (MAS). MAS is a severe, fatal complication, however,

little is known about its clinical courses and relation to initial symptoms, diagnosis and treatment.

Objectives: To clarify characteristics of MAS in patients with ASD and identify risk factors for developing MAS.

Methods: Consecutive ASD patients diagnosed with Yamaguchi's criteria in our hospital from April 2012 through September 2017 were retrospectively reviewed. Clinical symptoms, laboratory data and treatment information were collected from their charts. Patients were divided into two groups according to the complication of MAS and analyzed.

Results: Sixty five patients with ASD with available information were enrolled in the analysis. The mean age at ASD onset was 45.7 years old, and female was 81.5%. Among them, 12 patients (18.5%) developed MAS during the clinical course. Three patients developed MAS at the same time with the diagnosis of ASD, seven patients after the initiation of glucocorticoid treatment, and two patients at relapse. Five of nine who underwent bone marrow examination proved to be complicated with hemophagocytotic syndrome. The median duration from initial symptoms of ASD to fulfillment of Yamaguchi's criteria was shorter in patients with MAS than those without (10.5 days vs 21.0 days, $p=0.077$). Drug allergy occurred more frequently in patients with MAS (50.0% vs 22.6%, $p=0.056$). The maximum levels of ferritin and AST before treatment was higher in patients with MAS than those without (ferritin, 6090ng/mL vs 3434ng/mL, AST 86.5U/L vs 68.0U/L), whereas the maximum levels of CRP was lower in patients with MAS (8.9mg/dL vs 15.9 mg/dL). In nine patients who developed MAS after glucocorticoid treatment, white blood cell count decreased by almost half despite moderate to high dose of glucocorticoid, and the level of ALT increased 10-fold and ferritin increased 2-fold at MAS development. However, the levels of CRP at MAS rather decreased before treatment, suggesting the change in CRP did not reflect MAS development.

Conclusion: The incidence of MAS was 18.5% during the clinical course of ASD. Patients with MAS developed ASD more rapidly than those without. The ferritin and ALT levels predicted and reflected MAS development, whereas CRP levels was not associated with MAS.

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