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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FRI0614

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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FRI0615

EVALUATION OF COMPLIANCE AND RELATED FACTORS IN COLCHICINE TREATMENT IN FAMILIAL MEDITERRANEAN FEVER PATIENTS

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Background: Familial Mediterranean Fever (FMF) is an autoinflammatory disease requiring long-term treatment. Increasing the compatibility with colchicine treatment in patients with FMF is the first step for preventing amyloidosis. Patients' beliefs about medicines and treatment may affect treatment adherence and treatment success.

Objectives: The aim of this study was to determine adherence to colchicine treatment and related factors in FMF patients. In addition, patients' beliefs about colchicine, which are one of the important factors affecting the treatment adherence of patients, were evaluated.

Methods: Total of 179 patients with FMF was included in this study. The demographic and clinical features and MEFV gene mutations were recorded. The treatment adherence of the patients was assessed using by Compliance Questionnaire on Rheumatology (CQR). The Beliefs About Medicines Questionnaire (BMQ-T) was used to assess patient's beliefs about colchicine. The relationship between compliance of treatment and clinical characteristics of patients were assessed.

Results: One hundred thirteen (63.1%) of the patients were male. The mean age of patients was 34.5 ± 12.7 years and mean delay in diagnosis was 6.7 ± 8.4 years. The mean dose of colchicine was 1.37 ± 0.43 mg/day and, the percentage of patients using colchicine regularly was 66.5%. Adherence to treatment was higher in patients with concomitant diseases than those without comorbidities (p = 0.028). In addition, treatment compliance was higher in married patients compared to single patients (p = 0.013). The colchicine dose used in compatible patients was higher than in non-compatible patients (p = 0.033) (Table 1). We also found that as the BMQ-T Specific Necessity scores increased, compliance with treatment increased. On the other hand, as the BMQ-T General Overuse and General Harm scores increased, non-compliance with treatment increased (Table 2).

Table 1. Relationships between socio-demographic and clinical characteristics of patients and CQR score (n = 179)

Characteristics	CQR score		
	≥80% n=29	< 80% n=150	p
Age (years) median (25th–75th percentile)	37.0 (28.5–44.5)	29.0 (24.0–44.0)	0.099 ^a
Gender (n,%)			0.164 ^b
Male	15 (13.3)	98 (66.7)	
Female	14 (21.2)	52 (78.8)	
Marital status, (n,%)			0.013 ^b
Married	21 (22.8)	71 (77.2)	
Single/widowed/divorced	8 (9.2)	79 (90.8)	
Comorbidities, (n,%)			0.028 ^b
Yes	12 (26.7)	33 (73.3)	
No	17 (12.7)	117 (87.3)	
Delay of diagnosis (years)			0.920 ^a
Median (25th–75th percentile)	3.0 (0.5–10.0)	3.0 (1.0–9.0)	

Table 1. Demographic features and treatment response during anakinra therapy

Patient ID #	Age	Sex	Diagnosis	Duration of pericarditis follow-up (mo)	Prior medications	Number of recurrences before anakinra	Anakinra treatment duration (mo)	Time to corticosteroid discontinuation (mo)	Number of recurrences after daily dose of anakinra
1*	23	M	IRP	129	Colchicine, NSAIDs, CS, HCQ	6	52	9	No
2	32	F	IRP	128	Colchicine, NSAIDs	7	4	NA	No
3*	40	F	IRP	21	Colchicine, CS	6	8	1	No
4	20	M	IRP	11	Colchicine, CS	3	8	2	No
5	25	M	FMF	30	Colchicine, CS	5	15	1	No

F: Female, M: Male, CS: Corticosteroid, HCQ: Hydroxychloroquine, NA: Not applicable

*Dose tapering was unsuccessful in these patients

Colchicine dose, (mg/day) 1.5 (1.25–1.75) 1.5 (1.0–1.5) **0.033^a**
 Median (25th–75th percentile)
 Data represented either as the median (25th–75th percentile) or as the frequency. a: Mann-Whitney test b: Pearson Chi-Square test, CQR, Compliance Questionnaire on Rheumatology

Table 2. Beliefs about Medicines Questionnaire Scale Scores in Adherent and Nonadherent Patients.

Variable	CQR score			
	Discontinuous		Z	p
	≥80% n=29	< 80% n=150		
BMQ-T-Specific Necessity	4.8 (4.1–5.0)	3.8 (3.35–4.25)	-5.260	0.001
BMQ-T-Specific Concerns	2.8 (2.2–3.8)	2.8 (2.4–3.6)	-0.100	0.920
BMQ-T-General Overuse	2.25 (1.75–2.63)	2.75 (2.25–3.25)	-2.936	0.003
BMQ-T-General Harm	1.75 (1.38–2.63)	2.38 (2.0–3.0)	-3.212	0.001

Z; Mann-Whitney test, Data represented as median (25th–75th percentile, CQR; Compliance Questionnaire on Rheumatology

Conclusion: In patients with FMF, it is important to evaluate the compliance with the treatment due to the importance of colchicine to prevent amyloidosis that may occur in patients without treatment. As this study shows it is also important to determine patients' beliefs about medicine in terms of their influence on patients' compliance with treatment.

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ANAKINRA TREATMENT IN RECURRENT PERICARDITIS: SINGLE CENTER EXPERIENCE

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Background: Recurrent pericarditis (RP), however the etiology is unknown in the majority, may be observed in autoinflammatory diseases such as familial Mediterranean fever (FMF) and tumor necrosis factor receptor-1 associated periodic syndrome (TRAPS). Colchicine has long been used to treat pericarditis related to FMF as well as patients with idiopathic recurrent pericarditis (IRP) (1). Alternative treatments have been reported for cases with colchicine resistant RP.Objectives:

Objectives: The aim is to present our data regarding anakinra treatment in recurrent pericarditis either related to FMF or idiopathic, who are resistant to colchicine.

Methods: Patients who had received anakinra with a diagnosis of recurrent pericarditis either idiopathic or secondary to FMF followed in our autoinflammatory disease center between 2014-2018 are evaluated retrospectively. From patients' files, demographic and clinical features, response to other treatment approaches such as NSAID, corticosteroid, colchicine, were evaluated. All patients have been genetically screened for monogenic autoinflammatory diseases (MEFV, TRAPS, MVK, NLRP3, NOD2). Patients who had at least 3 attacks were administered anakinra 100 mg/day. Therapeutic efficacy, as well as side effect profile of anakinra is also assessed.

Results: There were 5 patients (3 male and 2 female) with the diagnosis of RP, 1 was related to FMF and 4 were idiopathic. The mean age of