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THU0586

CLINICAL CHARACTERISTICS AND RISK FACTORS OF POLYMYOSITIS AND DERMATOMYOSITIS COMBINED WITH INTERSTITIAL LUNG DISEASE IN PATIENTS RESIDING IN THE NORTHEAST SICHUAN PROVINCE IN CHINA

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Background: Idiopathic inflammatory myositis (IIM) is a systemic autoimmune disease characterized by nonsuppurative inflammation of striated muscle, myalgia, and proximal and symmetric muscle weakness. IIM can affect numerous organ systems, such as the skin, lungs, heart, and gastrointestinal tract. Polymyositis (PM) and dermatomyositis (DM) are the most common forms of IIM. Interstitial lung disease (ILD) is a common and severe complication of PM/DM, and concurrent ILD in patients with PM/DM is a major cause of death, with a fatality rate that exceeds 50% [1]. To explore the clinical features and risk factors of PM/DM combined with ILD (PM/DM-ILD) can improve the early diagnosis rate, put early interventions and improve the therapeutic effect.

Objectives: We explored the clinical features and risk factors of PM and DM, combined with ILD to increase early diagnosis and improve treatment outcomes.

Methods: Data were collected from 130 patients with PM/DM, treated at a single center from January 1, 2014 to September 30, 2018. General medical status, clinical symptoms, laboratory indicators, high-resolution computed tomography, therapeutic outcomes, and prognoses were retrospectively reviewed in patients with PM/DM with (ILD group) and without (NILD) ILD.

Results: There was statistically significant difference in age between the ILD group (n=65) and the NILD group (n=65) ($p<0.05$), but no significant between-group differences in PM/DM ratio, sex, or disease duration ($p>0.05$ for all). In the ILD group, initial symptoms were arthritis and respiratory symptoms, and myasthenia for the NILD group. Incidences of Raynaud's phenomenon, dry cough, expectoration, dyspnea on exertion, arthritis, fever, globulin (GLOB), erythrocyte sedimentation rate (ESR), and anti-Jo-1 antibody rate were higher for ILD ($p>0.05$ for all); however, albumin (ALB), creatine kinase aspartate aminotransferase activity ratio (CK/AST) and CK were significantly lower in the ILD group ($p>0.05$ for all). Bivariate logistic regression analysis showed that age, dry cough, arthritis, dyspnea on exertion, anti-Jo-1 antibody, and elevated GLOB were independent risk factors for ILD among patients with PM/DM.

Table 1. Bivariate logistic regression analysis on risk factors associated with ILD in patients with PM/DM

Variables	B	SE	p value	OR	95% CI for OR	
					Lower limit	Upper limit
Age	.053	.018	.003	1.054	1.018	1.092
Dry cough	.997	.449	.026	2.71	1.125	6.592
Arthritis	1.089	.481	.024	2.971	1.157	7.63
Dyspnea on exertion	1.192	.483	.014	3.295	1.279	8.493
Anti-Jo-1 (+)	1.517	.706	.032	4.558	1.141	18.201
GLOB	.093	.029	.001	1.097	1.036	1.162

Conclusion: The morbidity associated with PM/DM-ILD is high. Older age, dry cough, arthritis, dyspnea on exertion, anti-Jo-1 antibody positivity, and increased GLOB increase the risk of developing ILD. This information could be used to carefully monitor changing lung function in these patients.

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THU0587

COMPARING OF TWO DISTINCT SEVERITY SCORES FOR EVALUATING DISEASE SEVERITY IN FAMILIAL MEDITERRANEAN FEVER

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Background: Familial Mediterranean Fever (FMF) is a chronic hereditary disease which is characterized by recurrent attacks of fever accompanied by serositis. Severity of disease is related with morbidities such as amyloidosis and chronic renal failure.

Objectives: The aim of this study was to evaluate disease severity with two disease severity scores in FMF patients, and compare them.

Methods: The study included 160 patients with FMF. Demographic and clinical data were recorded in the patients' form. The patients' disease severity were evaluated with International Severity Scoring System for FMF (ISSF) and The Second Set of FMF severity score (F-SS-2).

Results: A total of 160 patients, comprising of 106 males (66.3%) and 54 (33.8%) females with a mean age of 33.5 ± 12.2 years were evaluated. Mean disease duration was 16.6 ± 10.3 years. Other demographic and clinical characteristics of the patients were shown in Table 1. According to ISSF, 67 (41.9%) patients were classified as having mild disease, 80 (50.0%) patients as intermediate disease and 13 (8.1%) patients as severe disease. On the other hand, 59 (36.9%) patients were classified as having mild disease, 41 (25.6%) patients as intermediate disease and 60 (37.5%) patients as severe disease according to F-SS-2 score. Higher rates of patients were found having severe disease according to F-SS-2 compared with ISSF ($p<0.001$). Also, lower rates of patients were scored as having intermediate disease with F-SS-2 compared with ISSF ($p<0.001$) (Table 2).

Table 1. Sociodemographic and clinical characteristics of patients (n=160)

Characteristics	
Gender, n (%)	
	Male 106 (66.3)
	Female 54 (33.8)
Age (years), mean $\pm$ SD	33.5 $\pm$ 12.2
Disease duration (years), mean $\pm$ SD	16.6 $\pm$ 10.3
Age of diagnosis (years), mean $\pm$ SD	23.1 $\pm$ 12.7
Delay of diagnosis (years), mean $\pm$ SD	6.2 $\pm$ 7.9
Age of symptom onset (years), mean $\pm$ SD	16.9 $\pm$ 11.8
Dose of colchicine, (mg/day), mean $\pm$ SD	
ISSF Score (n, %)	
Mild	67 (41.9)
Intermediate	80 (50.0)
Severe	13 (8.1)
F-SS-2 Score (n, %)	
Mild	59 (36.9)
Intermediate	41 (25.6)
Severe	60 (37.5)

Table 2. Comparison between F-SS-2 and ISSF Score

		ISSF Score (n=160)				p ^a
		Mild	Intermediate	Severe	Total	
F-SS-2 Score (n=160)	Mild	37 (62.7)	20 (33.9)	2 (3.4)	59 (100.0)	<0.001
	Intermediate	22 (53.7)	17 (41.5)	2 (4.9)	41 (100.0)	
	Severe	8 (13.3)	43 (71.7)	9 (15.0)	60 (100.0)	
	Total	67 (100.0)	80 (100.0)	13 (100.0)	160 (100.0)	

F-SS-2: The second set of Familial Mediterranean Fever severity score, ISSF: International Severity Scoring System for Familial Mediterranean Fever, ^aPearson Chi-Square test

Conclusion: Disease severity scores should be used for treatment choice, evaluation of disease prognosis and classify patients according to disease severity in clinical trials. In the current study, we found a difference between the classifications of disease severity, which could be a result of each scoring system uses different parameters of the disease.

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THU0588 CLINICAL PHENOTYPES OF IGG4-RELATED DISEASE IN SPAIN

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Background: Recently, several clinical phenotypes in IgG4-related disease have been described in a multinational and ethnically variate cohort¹.

Objectives: To assess the clinical presentation of IgG4-related disease (IgG4-RD) in Spanish patients and assess the distribution among different clinical phenotypes.

Methods: Clinical data were obtained from the Spanish IgG4-RD registry (REERIGG4) from October 2013 to December 2018, including 9 centers. We reviewed demographic data and organ involvement. The assignment of clinical phenotypes was done by 2 experts, based on organ involvement and clinical manifestations, following Wallace et al.¹ subsets. The phenotypes were: pancreato-hepato-biliary (HBP), retroperitoneum and aorta (RA), head and neck limited (HNL) and Mikulicz and systemic (MS). A fifth group designated not defined (ND), included the patients that did not fit in the previous phenotypes.

Results: One-hundred patients were included. Thirty-four (34%) were females, median age at diagnosis was 54.8 years (IQR 20.7). The ethnicity of the participants was: Caucasian 83%, Hispanic 12% and North-African/Middle-East 5%. Ninety-two percent were diagnosed with a biopsy. Regarding the diagnostic criteria, 85% met consensus pathology criteria and 94% comprehensive criteria.

Fifty-one patients (51%) had systemic IgG4-RD involving >1 tissue. The most commonly involved tissues were: retroperitoneum (35%), lymph nodes (19%), orbit pseudotumor (18%), salivary glands (16%) and pancreas (14%). Forty-two patients (42%) had elevated serum IgG4.

The representation of each clinical phenotype was: HBP 14%, RA 25%, HNL 26%, MS 20%, ND 15%. Patients were equally distributed from the perspective of ethnicity. Men were predominant in all groups (71, 84, 70, 73%) except in HNL (61% for women). Systemic disease was present in all the MS patients, but in 1/3 of the other groups. Serum IgG4 was elevated in 86% of the cases in HBP, 28% in RA, 19% in HNL, 60% in MS and 40% in ND.

Conclusion: The Spanish IgG4-RD population was mainly ethnically Caucasian. Few patients had serum IgG4 elevation. The most frequent phenotype was HNL, followed by RA. The HBP phenotype was less frequent than in previous reports. The influence of race could modify the clinical expression of IgG4-RD. Knowing the regional phenotypes of IgG4-RD may help clinicians improve disease management.

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THU0589 FREQUENCY, CHARACTERISTICS AND CLINICAL DETERMINANTS OF "PRODROME" IN FAMILIAL MEDITERRANEAN FEVER PATIENTS

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Background: Prodrome was defined by the presence of manifestations that precede an FMF attack and predict its emergence. This period might be turned to a window of the opportunity for the prompt treatment of impending attacks with fast-acting IL-1 inhibitors.

Objectives: We aimed to determine the frequency, characteristics and clinical determinants of prodrome in patients with FMF

Methods: 401 FMF patients were enrolled in this cross-sectional study. We applied a questionnaire to the FMF patients about attack types, pre-attack manifestations and latent time. Four hours was accepted as the cutoff point for prodrome as based on the literature, which is compatible with the pharmacokinetics of anakinra.

Results: The mean age was 37.7 (±11.0) years, and the disease duration was 20 years (3-58). 248 of the patients (61.8%) were female. MEFV gene mutations were achieved in 364 (90.8%) patients, of these 121 (30.2%) patients harbored homozygous exon 10 mutations. Patients with a latent time over four hours considered as prodrome positive (PP) patients (n=141). Male gender, homozygous MEFV mutations, peritonitis, pleuritis, arthritis and ELE were more pronounced in PP group. 25.9% of patients with peritonitis, 14.7% of patients with pleuritis, 13.2% with arthritis and 5.5% with ELE had prodrome with variable latent time durations (median, 6.7(4-72), 6 (4-48), 6 (4-72), and 7.7 (4-30) hours, respectively). Prodrome was found to be more common in those with peritonitis. Male gender, having peritonitis or arthritis were found to be the independent clinical determinants of having prodrome (RR 1.72 (1.07-2.76), P=0.02, 4.27 (1.80-10.1), P=0.001, 1.77 (1.04-3.04), P=0.04, respectively). Age, MEFV mutations, pleuritis and ELE were not found as clinical determinants of prodrome.

Conclusion: Especially male patients with peritonitis or arthritis tend to have prodrome more than other patients, and should be questioned. Prodrome positive patients are candidates for prevention of the impending attacks with on-demand treatments.

Table 1. Patients demographics and disease characteristics

	Prodrome (+) (n=141)	Prodrome (-) (n=260)	P
Age, years	37.7 (10.5)	37.8 (12.8)	0.92
Female	78 (55.3%)	170 (65%)	0.05
Age at FMF diagnosis, years	24(3-62)	25(2-59)	0.52
Disease duration, years	21 (4-58)	19 (3-58)	0.08
Fever	0	9 (3.5%)	0.03
Peritonitis	133 (94.3%)	197 (75.8%)	<0.001
Pleuritis	92 (65.2%)	123 (47.3%)	0.01
Arthritis	103 (72.9%)	129 (49.8%)	<0.001
Myalgia	0	6 (2.3%)	0.09
Erysipelas like erythema	46 (32.6%)	58 (22.3%)	0.02
Homozygous*	57 (43.2%)	64 (27.6%)	0.02
M694V/M694V	50(35.5%)	51(19.6%)	0.01
M694V/-	19 (13.5%)	46(17.7%)	0.19
M694V/M680I	13 (9.2%)	23 (8.8%)	0.98
M694V/V726A	12 (8.5%)	20 (7.7%)	0.88
M694V/E148Q	6 (4.3%)	15 (5.8%)	0.45
M680I/M680I	6 (4.3%)	9(3.5%)	0.76

* 364 patients MEFV mutations were achieved. 9.2% of patients MEFV mutations is not known, 7.7% of patients is gen negative FMF.

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