Rheumatological side effects were observed in 15 patients (14.7%): inflammatory arthralgia (8), arthritis (6), myositis (2) and aortitis (1). The time of appearance of the rheumatological irAEs was of 6.36 months (± 5.81). From the 7 patients with previous diagnosis of an immune-mediated disease, only 1 patient with psoriasis suffered a worsening of skin symptoms and another one with psoriatic arthritis had a monoarthritis episode.

Among the 14 patients who suffered from arthralgia/arthritis, the most frequent pattern was oligoarticular (40%), followed by poly (30%) and monoarticular (30%). The most affected joint was the knee ($n=4$), followed by wrist (3), hands (3), ankle (2), shoulder (2) and foot (1).

Apart from the articular disease, 2 patients suffered from myositis in lower limbs, with weakness of legs and arthralgia in their knees. A patient was diagnosed of aortitis through a PET, although the patient did not have any symptom.

Immunology tests with RF, ACCP and ANA were done in 8 out of the 15 patients with musculoskeletal irAEs, being negative in all cases.

Interviewing 40 of the patients who received ICBT, 7 of them (17.5%) referred xerophthalmia. 6 of them (15%) had a positive Schirmer test, and 3 (7.5%) had a reduced tear break-up time. None patient met Sjögren criteria.

In 6 out of the 15 patients with rheumatological irAEs (all of them with arthralgia/arthritis) the ICBT was removed, temporary in all cases, and could be reintroduced after a mean of 33 ± 18.9 days. 6 patients were treated with NSAIDs and 5 with oral prednisone (2 of them also required intra-articular corticoids).

Conclusion: Rheumatological irAEs are common in clinical practice. Arthralgia-Arthritis and dry-syndrome are the most frequent adverse events.

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LYMPHADENOPATHY IN IGG4-RELATED DISEASE IS ASSOCIATED WITH DISEASE ACTIVITY AND EOSINOPHILIA

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Background: IgG4-related disease (IgG4-RD) is an immune-mediated systemic disease characterized by the enlargement of multiple organs. IgG4-RD frequently involves the pancreas, lacrimal glands, salivary glands, bile duct, kidneys, lungs, and retroperitoneum, but can potentially affect any organs. Lymphadenopathy is often seen in patients with IgG4-RD along with other organ involvements, but the significance of the exist of lymphadenopathy in IgG4-RD is unclear.

Objectives: The aim of this study is to clarify the characteristics of patients with IgG4-RD who have lymphadenopathy.

Methods: We conducted cluster analysis with clinical data in consecutive 85 patients with untreated active IgG4-RD in our institute. We divided the patients into two groups who had lymphadenopathy or not (lymph group and non-lymph group), and compared clinical characteristics between the two groups. We counted the areas of swollen lymph nodes and performed correlation analysis with clinical characteristics. We also comprehensively screened serum protein concentrations in IgG4-RD patients ($n=15$) and healthy controls ($n=5$) by using high-throughput proteomic analysis, and differentially expressed proteins were extracted. We compared the concentrations of serum proteins which were extracted between lymph group and non-lymph group.

Results: Among 85 patients with IgG4-RD, 44 (52%) were male. Median age was 63 years old. Forty-seven (55%) patients were lymph group. Cluster analysis revealed that lymphadenopathy was in a cluster with serum IgG4, IgG4/IgG, IgG4-RD responder index (IgG4-RD RI), soluble interleukin-2 receptor (sIL-2R), eosinophilia, erythrocyte sedimentation rate (ESR), lung involvement, hemoglobin, albumin, C3 and C4. In lymph group, serum IgG4, IgG4/IgG, IgG4-RD RI, eosinophilia, sIL-2R and ESR were significantly higher than non-lymph group (Figure). Spearman's correlation analysis revealed that areas of the swollen lymph nodes were positively correlated with serum IgG4, IgG4/IgG, sIL-2R, eosinophilia. Proteomics analysis revealed that 68 proteins were significantly higher in IgG4-RD ($p<0.05$). Among them, proteins related to adhesion of immune cells, fibrosis, formation of lymph node and eosinophil activation such as integrin $\alpha 1\beta 1$, CCL18, CCL19 and eotaxin3 were higher in lymph group ($p=0.03$, $p=0.03$, $p=0.06$ and $p=0.06$, respectively).

Conclusion: Lymphadenopathy in IgG4-RD is associated with disease activity and eosinophilia. Proteomics analysis can provide some clues for elucidating the significance of lymphadenopathy in IgG4-RD.

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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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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)