

Results: We studied 100 patients/180 affected eyes (54M/46W), mean age 40.7±10.1. The ocular pattern was panuveitis (n=62), posterior (27) and anterior uveitis (11). Before IFX they received IV MP (28), cyclosporine (75), azathioprine (56), methotrexate (43) and others (33). IFX dose ranged between 3–5mg/kg/4 or 8 weeks. In patients in remission IFX was optimized (n=28) or stopped (n=20).

Table

	Baseline	1 st week	1 st month	3 rd month	6 th month	1 st year	2 nd year	3 rd year	4 th year
VA (mean±SD)	0.5±0.35 n=193	0.5±0.3* n=189	0.6±0.3* n=187	0.6±0.3* n=183	0.6±0.3* n=176	0.6±0.3* n=159	0.6±0.3* n=130	0.6±0.3* n=88	0.7±0.3* n=60
Cells in the anterior chamber (median [IQR])	1 [0-2] n=190	0 [0-1]* n=187	0 [0-0]* n=184	0 [0-0]* n=180	0 [0-0]* n=171	0 [0-0]* n=153	0 [0-0]* n=126	0 [0-0]* n=82	0 [0-0]* n=58
Vitritis (median [IQR])	1 [0-2] n=188	0.5 [0-2]* n=182	0 [0-0]* n=182	0 [0-0]* n=177	0 [0-0]* n=168	0 [0-0]* n=149	0 [0-0]* n=122	0 [0-0]* n=84	0 [0-0]* n=58
Retinal vasculitis (% affected eyes)	53.4% n=188	43.1% n=182	22.2% n=182	10.9% n=177	7.4% n=171	2.4% n=153	1.9% n=124	0.4% n=84	0.4% n=54
OCT (μ) (mean±SD)	331.5 ± 136.1 n=59	324 ± 123.8* n=53	298.7 ± 101.8* n=53	283.2 ± 83.6* n=49	265.8 ± 65.3* n=42	264.1 ± 61.3* n=46	263.6 ± 65.2* n=27	239.8 ± 26.8* n=14	231.2 ± 33.7* n=8

*p<0.05. VA=visual acuity. Data are of affected eyes.

Conclusions: IFX is an effective long term-treatment in refractory Uveitis of BD. Optimization and even discontinuation of IFX after remission is possible.

Disclosure of Interest: None declared

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FRI0608 STUDY OF ARTICULAR SARCOIDOSIS IN A TERTIARY CARE HOSPITAL

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Background: Sarcoidosis is a systemic granulomatous disease, being the joint involvement a poorly studied manifestation.

Objectives: To describe the clinical and demographic characteristics of patients with sarcoidosis, paying particular attention to the joint involvement and its possible relationship with other extra-articular manifestations, as well as the treatment received.

Methods: A retrospective, observational study that included 104 patients who were admitted to the Hospital of León between January 2011 and December 2015 with main or secondary diagnosis of sarcoidosis; according to clinical onset, imaging tests and/or anatomopathological study. The variables studied were: age at the time of diagnosis, sex, type of joint involvement, forms of extra-articular involvement, serologic parameters and drugs received. Statistical analysis was performed using SPSSv22.0, p<0.005.

Results: 57.7% of the patients included in the study were women with a mean age of 53.42±18.4 years. At the systemic level, 35% of them presented fever, 66.3% lymphadenopathy and 4.8% splenomegaly. 97.1% of the patients presented pulmonary involvement, with stage II being the most common (46.2%). Only 8 patients had cardiac abnormalities. Ocular involvement was observed in 10.6%, predominating uveitis. The most common renal manifestation was hypercalciuria in 6.7%. The presence of neurological involvement was exceptional, detecting 3 cases of neurosarcoidosis. In the cutaneous involvement (2.9%), erythema nodosum predominated (17.3%). The CRP levels were normal in 44.3% (<5mg/dl) and high levels of CRP were found in 25.3% of the sample (>30mg/dl). 62.6% presented pathological figures of ACE. The joint involvement was present in 38.5% of the patients (14.4% in the form of arthralgia, 2.9% as periartthritis, 13.5% as acute arthritis, 4.8% as chronic arthritis and 2.9% as sacroiliitis). The 19.23% debuted as Löfgren syndrome, being seen an association between the presence of acute arthritis and erythema nodosum (p0.000). The mean age observed in patients with acute arthritis was lower than the one of the other patients with other joint manifestations (p0.044) and a statistically significant relationship was observed between the absence of joint and ocular involvement (p0.011). Regarding the treatment of joint manifestations, 30.77% of the cases were resolved with NSAIDs, 46.15% with corticosteroids in a monotherapy and 15.38% required an immunosuppressant. In our study, most patients treated with corticosteroids did not show articular nor ocular involvement (p0.018), although when analysing the ocular involvement separately, many received corticoid treatment (42.86%). Only one patient required Adalimumab for refractory uveitis and another presented anti-TNFα-induced sarcoidosis (Infliximab).

Conclusions: The pulmonary involvement is the predominant one in patients with sarcoidosis. The acute arthritis occurs in younger patients and is associated with the onset of erythema nodosum. The joint involvement is usually not severe and, although it has not been demonstrated, it seems that the use of corticosteroids

predominates in the extra-articular manifestations. Only 1 in 4 patients showed an increase of acute phase reactants.

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FRI0609 USEFULNESS OF FDG-PET/CT IMAGING AND SEROLOGICAL BIOMARKERS TO PREDICT RELAPSE IN IGG4-RELATED DISEASE

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Background: IgG4-RD shows relapses frequently. It is important to search to the factors to predict relapse. Recent research has shown the usefulness of FDG-PET/CT for IgG4-RD because it is more sensitive than conventional imaging to detect organ involvement of the disease. It has been suggested that FDG-PET/CT is also useful for monitoring therapeutic response of IgG4-RD.

Objectives: We investigate the usefulness of FDG-PET/CT imaging and serological biomarkers to predict relapse in IgG4-RD.

Methods: We analyzed 24 patients with IgG4-RD treated for more than 1 year between 2008 and 2016 in our facility. The diagnosis for IgG4-RD was based on comprehensive diagnostic criteria for IgG4-RD. All cases underwent FDG-PET/CT at least once, and laboratory data were collected from their medical records retrospectively. Levels of serum C-reactive protein (CRP), eosinophil/leukocyte ratio, serum IgG, IgG4, IgA, IgM, IgE, sIL-2R and serum complement were investigated.

Results: The patients had a mean age of 67.9 years (range: 50–87 years). In the cases with high FDG uptake on FDG-PET/CT, they had a greater number of organ involvements, higher serum IgG and sIL-2R levels. Eight patients experienced relapses following treatment. Higher serum IgG predicted relapses of IgG4-RD. FDG-PET/CT findings at baseline were not associated with relapse. FDG-PET/CT was performed in 13 patients after initiation of treatment and 4 patients had a relapse. There were no significant reduction of abnormal FDG uptake in 6 patients, and 4 of 6 patients relapsed.

Conclusions: In this study, we examined the factors to predict relapse in IgG4-RD. Patients with higher serum IgG were regarded as a risk of relapse, but FDG-PET/CT findings at baseline were not associated with relapse. FDG-PET/CT reexamined after initiation of treatment is useful to predict relapse of IgG4-RD.

References:

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FRI0610 PROBLEMS IN THE DIAGNOSIS OF FAMILIAL MEDITERRANEAN FEVER IN TURKEY

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Background: The diagnosis of FMF (Familial Mediterranean Fever) can be missed or delayed even in a country like Turkey, one of the most prevalent places for FMF (1).

Objectives: We compared the duration of delay in diagnosis before and after year 2000, and assessed the problems related with the diagnosis of FMF.

Methods: We studied 143 (102 F, 41 M) consecutive patients with FMF seen at the rheumatology outpatient clinic, between November 2016 and January 2017. Patients completed a self-administered questionnaire that assesses initial symptoms, previous diagnosis and treatments received before the formal diagnosis of FMF. Patients were divided in 2: as Group 1, which includes 70 patients seen for the first time by a physician before 2000 and Group 2, which includes 73 patients seen after 2000.

Results: The median age of the patients was 34 years [IQR:27–45]. The F/M ratio was 2.4. The initial symptom was abdominal pain in the majority (n=134, 89%), followed by fever (n=114, 75%), arthritis (n=66, 44%), pleuritic pain (n=21, 14%), arthralgia (n=11, 7%) and other complaints (n=14, 9%).

The median age at initial symptom was 10 years (IQR:6–23). The median delay in diagnosis was 8 years [IQR:2–15]. This was significantly shorter in Group 2 (median: 4 years [IQR:0–11]) than that observed in Group 1 (median: 10 years [IQR:6–17]), as shown in Table.

A total of 110 patients (73%) were diagnosed with one or more diseases or syndromes other than FMF. These were appendicitis (n=50, 45%), gastrointestinal diseases (n=44, 40%), acute rheumatic fever (n=36, 33%), inflammatory

arthritis (n=18, 16%), gynecological diseases (n=11, 10%) kidney stones (n=9, 8%) and others (n=34, 31%). As shown in Table, the frequency of patients with misdiagnosis, was significantly lower in Group 2 (66%) compared to Group 1 (84%). A total of 59 patients (39%) received other long-term treatments, mainly monthly penicillin (n=28), prior to colchicine. There were 41 surgical interventions in 36 patients (24%), before the diagnosis of FMF, the most common being appendectomy in 31, gynecological operations in 5, cholecystectomy in 3 and others in 2 occasions. It was noted that, the frequency of surgical operations was significantly decreased in Group 2 (12%) compared to Group 1 (27%) (Table). The presence or absence of MEFV mutations was assessed in 69 patients (46%) before the diagnosis or after to reinforce the diagnosis. As expected, this was significantly more frequent in Group 2 (59%) compared to Group 1 (33%) (Table). Seventy patients (46%) were diagnosed as FMF only after someone else in the family (n=44) or a friend (n=26) had a similar diagnosis. The frequency of these patients was similar when Group 1 and 2 were compared.

Table: Demographic and clinical characteristics of Group 1 (patients seen before 2000) and Group 2 (patients seen after 2000)

	Group 1, (n=70)	Group 2, (n=73)	P
Male/Female, n	22/48	19/54	Non significant
Current age, med [IQR] years	40[31-51.5]	32[24.5-38]	<0.001
Delay in diagnosis, med [IQR] years	10 [6-18]	4 [0-11]	<0.001
Misdiagnosed patients, n (%)	59 (84)	48 (66)	0.011
Surgery before diagnosis, n (%)	19(27)	9(12)	0.026
Assessment of MEFV mutations, n (%)	23 (33)	43 (59)	0.005
Diagnosed as FMF after someone else, n (%)	27(39)	38(52)	0.1

Conclusions: Although there is considerable decrease in delayed diagnosis of FMF, there is still significant amount of misdiagnoses after the year 2000, even in a geography where FMF is highly prevalent.

Table: Demographic and clinical characteristics of Group 1 (patients seen before 2000) and Group 2 (patients seen after 2000)

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[1] Tunca M et al; Turkish FMF Study Group. Familial Mediterranean fever (FMF) in Turkey: results of a nationwide multicenter study. *Medicine* (Baltimore). 2005 Jan;84(1):1–11.

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FRI0611 ASSESSMENT OF PANNICULITIS CLINICAL OUTCOMES: RISK FACTORS FOR RECURRENCE AND PREDICTORS OF SLOW REGRESSION OF INDURATIONS

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Background: Currently there's no clear understanding of the clinical course and outcomes of panniculitis with predominant involvement of subcutaneous adipose tissue (SAT), which is often associated with the involvement of locomotor system and viscera. Consistent elaboration of both is of paramount importance, as it may give a deeper insight into clinical and curative factors which may have impact on the disease prognosis.

Objectives: To assess clinical outcomes of panniculitis (risk factors for recurrence and predictors of slow regression of indurations).

Methods: 186 pts (172 females, 14 males) aged 43.5±14.5 years with different types of Pn, who were at the record of V. A. Nasonova Research Institute of Rheumatology during 2009–2015 yy. Disease duration varied from 1 week to 20 years. General clinical examination and lab tests (serum levels of α1-antitrypsin, amylase, lipase, ferritin, creatine phosphokinase (CPK), rheumatoid factor) (RF), ANF, anti-DNA, ANCA), bacteriological and serological tests, radiological examination (chest CT scan), ultrasound examinations of skin and subcutaneous fat (SF) around the nodules, tuberculin skin test and histopathology of nodular skin samples were performed at baseline. Patients were re-tested during the follow up (FUP) when necessary.

Results: Female middle-aged patients prevailed in the study group with the disease duration from 1 week to 25 years. *Saucer-like* depressions as a Pn - specific phenomenon was documented in 28.5% cases. The estimated risk of *saucer-like* depressions was higher in patients who manifested Pn at the age of >40 years (OR 4.9, 95% CI 2.3–10.4; *p*<0.0001); this subgroup also showed lesion proneness to confluence and forming of irregular shape conglomerates (OR 2.9, 95% CI 1.45–5.7; *p*=0.002), tendency to forming large size >3 cm lesions (OR 4.96, 95% CI 2.2–10.97; *p*<0.0001), and the disease lasting over 3 months (OR 24.9, 95% CI 9.2–67.4; *p*<0.0001). All pts were re-examined within 1–6 years. Median time to onset of nodules regression was 2 [1;6] months. Recurrences were documented in 66 (35.5%) pts during the follow up. Logistic regression was used for multivariate statistical analysis in order to identify the potential predictors of delayed (>3 months) nodules regression and recurrence risk factors. Used model identified the following predictors of delayed nodules regression (i.e., disease regression): age >40 years (OR 2.58; CI 1.02–6.5; *p*=0.04), and presence of saucer-like depressions (OR 5.05; CI 1.2–21.7; *p*=0.03). The sensitivity of used

statistical model was 70%, specificity – 71%, positive predictive value - 74%. Disease duration >3 months (OR 4.7; CI 2.0–10.6; *p*=0.0002) was identified by our model as the predictor of recurrences with 60% sensitivity, 78% specificity, and 59% positive predictive value.

Conclusions: Pn tends to regress more slowly in pts aged over 40 and in pts having saucer-like depressions. Trend to recurrences is not so evident in Pn, although probability of recurrence increases in pts with longer disease duration at the time of initiation of therapy.

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FRI0612 IGG4-RELATED DISEASE IN EASTERN MEDITERRANEAN: CLINICAL FEATURES AND OUTCOMES OF A LARGE COHORT

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Background: Since majority of IgG4-related disease (IgG4-RD) patients in the literature are from Far East and United States, there is a lack of large series from other parts of the world.

Objectives: We aimed to identify the clinical characteristics and outcome of Turkish IgG4-RD patients from a tertiary center.

Methods: In step 1, clinical and histopathological features of patients having any disease under the clinical spectrum of IgG4-RD were retrospectively reviewed. For step 2, patients prospectively diagnosed are enrolled into the study. Patients fulfilling the "definite" diagnosis according to comprehensive diagnostic criteria were recruited (enrolled (n=52), excluded (n=47)).

Results: Median age was 51.1 years and no gender predominance was observed (Male/female: 26/26). Retroperitoneal fibrosis is the most frequent presentation;

Table 1. Distribution of clinical findings and organ involvement (n=52)

Constitutional symptoms, n (%)	
Fatigue	31 (59.6)
Tiredness	16 (30.7)
Night sweats	15 (28.8)
Weight loss	14 (26.9)
Fever	13 (25)
Retroperitoneal fibrosis, n (%)	23 (44.2)
Lymphadenopathy, n (%)	20 (39.2)
Any cardiovascular involvement, n (%)	15 (28.8)
Periaortitis	12 (23.1)
Pericardium	5 (9.6)
Coronary periarthritis	4 (7.7)
Abdominal aort aneurysm	1 (1.9)
Orbital pseudotumor, n (%)	12 (23.1)
Orbital mass/proptosis	6 (11.5)
Extraocular muscles	6 (11.5)
Pancreas, n (%)	12 (23.1)
Major salivary glands, n (%)	11 (21.2)
Lacrimal glands, n (%)	9 (17.3)
Mediastinal fibrosis, n (%)	6 (11.5)
Ear, nose, sinuses, n (%)	5 (9.6)
Lung fibrosis, n (%)	5 (9.6)
Skin, n (%)	4 (7.7)
Pleura, n (%)	4 (7.7)
Gall bladder and Biliary ducts, n (%)	4 (7.7)
Thyroid, n (%)	3 (5.8)
Liver, n (%)	3 (5.8)
Kidney (mass), n (%)	3 (5.8)
Pachymeningitis, n (%)	2 (3.8)

Breast involvement: (n=1), tubulointerstitial nephritis (n=1).

