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THU0570 EFFICACY AND SAFETY OF ANAKINRA IN THE TREATMENT OF AUTOIMMUNE MYOCARDITIS

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Background: Virus-negative or "autoimmune" myocarditis(VNM) is a severe, inflammatory heart disease with a poor prognosis, and is a leading cause of inflammatory dilated cardiomyopathy(i-DCM). Therapies are limited. Preliminary data indicate that interleukin-1(IL-1) plays a key role in the initiation and maintenance of the inflammatory heart response, sustaining an auto-inflammatory cycle^{1,2}.

Objectives: to evaluate the efficacy and safety of anakinra(ANK) in improving Left Ventricular Ejection Fraction(LVEF) on transthoracic echocardiography(TTE) in patients with VNM.

Methods: Biopsy-proven VNM patients were enrolled and treated with ANK 100 mg daily subcutaneously. All patients received treatment with the maximum tolerated dose of any beta blockers and ACE-inhibitors, according to current guidelines. At baseline and 8±4 weeks after ANK therapy, all patients underwent a full evaluation with assessment of functional status(New York Heart Association[NYHA]), measurement of high-sensitive troponin T(hs-TnT) and NT-proBNP serum levels, electrocardiography(ECG), 24h-ECG-Holter, TTE and cardiac magnetic resonance(CMR). Any myocarditis-related complication, cardiovascular deaths and adverse events(AEs) was recorded during follow-up. Continuous variables were assessed with the Wilcoxon signed-rank test for non-parametric tests and a p value<0.05 was considered statistically significant.

Results: Eleven patients(F/M=5/6, mean age 46,2±12,2 years) diagnosed with EBM-proven VNM were enrolled. Nine patients received ANK as first line therapy, and in 5 cases ANK was used as monotherapy; ANK was combined with prednisone(mean dose 31,7±16,7 mg daily) in 6 patients, 5 of them were concomitantly treated with azathioprine. On EMB, 6 patients were classified as i-DCM, 3 with acute VNM e1 with active and chronic VNM³⁻⁴. Clinical onset was characterized by congestive heart failure in the most cases(72.7%). The majority of patients(72.7%) was in NYHA class III-IV. Mean LV-EF on TTE at baseline was 38,7%±19,6, with comparable findings on CMR(36,45%±18,0), and 8 patients(72.7%) had a depressed LV-EF(<55%). At baseline, mean levels of hs-TnT and NT-proBNP were 150,0±153,9 ng/L and 6968,8±10788,4 pg/ml, respectively. Hs-TnT and NT-proBNP levels were elevated in 10(90.9%) and 9 patients(81.8%) respectively. At 8 weeks, LV-EF improved in 10 patients(90.9%). The LV-EF increase was>10% in 5 patients(45.5%) and between 5-10% in 5 cases(45.5%); only 1 patient showed a <10% LV-EF decrease. Mean LV-EF at the end of follow-up improved to 49,4%±10,8(p=0.059). When evaluating the 8 patients with baseline reduced LV-EF, the LV-EF improvement was statistically significant (baseline 29,2%±12,9; after ANK 45,2%±9,2,p=0.025). The LV-EF amelioration was paralleled by clinical improvements in all patients, since the majority of them(90.9%) were in NYHA class I-II at the end of follow-up. Consistently, hs-TnT declined after 8 weeks(64,6±100,7 ng/L,p=0,028), and a similar trend was observed for NT-proBNP, even though not statistical significant(2582,6±5048,1 pg/ml,p=0,06). We did not observe any myocarditis-related death or complications, nor any ANK-related AEs.

Conclusion: Our pilot study supports the efficacy and safety of ANK in the treatment of inflammatory heart failure in VNM and provides the first

clinical evidence to support the therapeutic blockade of IL-1 in myocarditis.

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THU0571 THE IMPACT OF AGING ON FAMILIAL MEDITERRANEAN FEVER PATIENTS

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Background: Familial Mediterranean Fever (FMF) is a monogenic autoinflammatory disorder with innate immune activation with an onset before age 20 in approximately 90% of the patients. There is scarce data on the effect of aging on FMF patients over 40 years of age

Objectives: This study aims to collect data on FMF patients who have survived over 40 years of age. Here we report our preliminary data on disease course and treatment status and comorbidities of our patients with FMF.

Methods: Among the FMF patients who have been followed in our FMF outpatient clinic with a pool of approximately 5000 patients, those who have aged 40 and over are being included to the study. As by today 180 patients are considered for evaluation. The files of patients were reviewed and a standard questionnaire was used to interview the patients. Here we report the results of 100 of these patients (56%) who were contacted for this purpose. These patients were questioned on their demographic characteristics, comorbid conditions, colchicine treatment details, and attack information. In order to see the trend of the change in the parameters assessed, the patients were divided into two groups based on their present age (Group 1: 40-50 years, Group 2: ≥50 years).

Results: A total of 100 (78 F, 22M) patients were evaluated. There were 61(46F, 15M) patients aged between 40-50 years and 39 (32F, 7M) over 50. The demographic characteristics and clinical features of these patients are given in Table 1. Besides 3, all patients were still on colchicine regularly. Ninety-six percent of the patients declared overall benefit from colchicine therapy; however 38% experienced a side effect related to this treatment. Over 88% of the patients reported decrease in severity and frequency of FMF attacks. The mean daily colchicine dose was lower in the age 50 and over group (1.7±0,77 mg versus 1,35 ±0,38 mg). There were no patients with AA amyloidosis in neither age group. The mean duration from the last attack increased from 15.3 ±19.7 months to 35.6±52 months in the older patients. One or more additional disease was present in 75% of this patient group. Among the comorbidities hypertension was the most frequent, diagnosed in 25% of the patients, followed by hypothyroidism (16%), diabetes mellitus (10%) and cardiac disease (5%). Sixty-five of the patients were receiving other medications in addition to colchicine.

Table 1. Clinical course and co-morbidities in two age groups over 40 years

n	Group 1* (n=61)	Group 2** (n=39)	p
Sex (F:M); current age (mean±SD) (yr)	(46 :15);45.5 ± 2.29	(32:7); 57.05 ± 6.81	0,43; <0.001
Mean duration since the last episode, (mean±SD, mo)	15.3 ± 19.7 (1-60)	35.67 ± 52.05 (1-276)	0,012
Number of patients on colchicine therapy, n (%)	59 (96.7)	38 (97.4)	NS
Mean colchicine dose, mg/day (current)	1.7±0.76	1.41±0.45	0.03
Number of patients with decrease in attack severity, n (%)	54(88,5)	35 (89,7)	NS
Number of patients with decrease in attack frequency, n (%)	57(93,4)	37 (94,8)	NS

Co-morbidities, n (%)			
Hypertension, n (%)	12(19.6)	13 (33.33)	NS
Hypothyroidism, n (%)	12(19.6)	4(10.2)	NS
Type 2 Diabetes Mellitus, n (%)	5(8.2)	5(12.82)	NS
Rheumatological diseases, n (%)	5(8.2)	3(7.7)	NS
Cardiac disease, n (%)	3(4.9)	2(5.1)	NS
Malignancies, n (%)	2(3.2)	2(5.13)	NS
Additional medications (number of patients, %)	36 (61)	29 (74.3)	NS

Conclusion: According to our preliminary data the majority of the patients continue to take colchicine after age of 40. However the frequency of FMF attacks as well as daily colchicine dose decrease as the patients get older. With well designed trials stopping colchicine treatment may be considered in a subgroup of patients after 50 years of age. Approximately $\frac{3}{4}$ of the FMF population over 40 years of age has a comorbidity that necessitates additional medications which underlines the need for special attention.

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THU0572 RHEUMATOLOGICAL ASPECTS OF FORMS AND VARIANTS OF PANNICULITIS

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Background: Panniculitis (Pn) is a set of heterogeneous inflammatory diseases characterized by lesions of subcutaneous fat (SCF) and often accompanied with the defeat of the musculoskeletal system and internal organs. Depending on the etiology and histomorphological picture, Pn is divided into two forms: septal (SPn) and lobular panniculitis (LPn) with and without signs of vasculitis, which is reflected in the clinical picture of the disease. Often Pn is one of the symptoms of rheumatic diseases (Rd), it can lead to late diagnosis of the underlying disease.

Objectives: to study the frequency of occurrence of forms and variants of Pn in rheumatological practice on the basis of long-term prospective observation

Methods: We examined 687 patients with Pn (613 women and 74 men, mean age 39.7±11.31 and 41.2±12.57 years, respectively) with the referral diagnosis of "Erythema Nodosum" (EN) or "Panniculitis", who were observed in the rheumatologic clinic in 2007-2017. In addition to general clinical examination, we determined serum concentrations of α -1 antitrypsin, amylase, lipase, ferritin, creatine phosphokinase, conducted a CT scan of the chest organs, immunological, TB tests and pathological study of skin biopsy from the node.

Results: As a result of application of the developed diagnostic algorithm, 430 patients (62.6%) were diagnosed with SPn and 249 patients (36.2%) - with LPn. In 9 cases (1%) (3 patients with SPn and 6 — with LPn), it was not possible to confirm the variant of Pn, despite the comprehensive examination. In 8 patients (1.1%) no Pn data were revealed, the following diagnoses were verified: 2 patients (25%) — discoid lupus and lipodystrophy, 1 (12.5%) — erysipelas, lichen, fixed erythema and keratoacanthoma. SPn in 94% of cases (400 patients) was presented by EN, in 4% (18) — by superficial migrating thrombophlebitis, in single cases (1-0%) — by eosinophilic fasciitis, skin nodular polyarteritis and scleroderma-Pn (2, 6 and 1 patients, respectively). In 28% of cases LPn was associated with systemic connective tissue lesions, in 27% with idiopathic lobular Pn (Weber — Christian panniculitis) ($p < 0.002$), in 32.92% with lipodermatosclerosis and in 14.28% with subcutaneous sarcoid. In a few cases LPn was caused by lymphoproliferative (2.88%) and cancer (2.49%) diseases, calciphylaxis (2.05%), erythema Bazin (1.64%), cold Pn (0.82%) and alpha 1-antitrypsin failure (0.41%)

Conclusion: the results confirm that patients with SPn were found twice as often as with LPn. Pn was mainly associated with sarcoidosis, EN and Rd, which confirms the relevance of further research of Pn.

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THU0573

COMPLICATIONS AND ACTIVITY OF UVEITIS IN A MULTIDISCIPLINARY REFERENCE UNIT IN THE NORTH OF SPAIN

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Background: Uveitis, an intraocular inflammatory disease, is a significant cause of visual impairment. It is not known how many patients with uveitis will retain visual acuity and how many develop visual impairment or even blindness. The activity of uveitis, the use of topic and oral steroids could produce ocular complications.

Objectives: To describe the complications and the activity of the uveitis in a cohort of 500 patients diagnosed in one unit of reference in the North of Spain.

Methods: A prospective study of complications and the activity of the 500 adults with uveitis were evaluated in the multidisciplinary Uveitis Unit of Complejo Hospitalario de Navarra from January of 2010 to March of 2015. Uveitis activity and the type of complication to the year of follow-up were analysed. The variables were collected according to the terminology of the activity of the SUN (inactive, worsening, improvement and remission). To facilitate statistical analysis variables inactive and remission were grouped in a same term. A study of characterization of the evolution and worsening of the activity to the year of follow-up with age, sex, foreign, type of uveitis, etiology, laterality, treatments and complications. The characterization was carried out through the SPAD 8 program and was carried out by contrast Chi square.

Results: We collected a total of 500 patients, 54.4% of patients remained inactive or in remission after 1 year of follow-up, 13.2% presented worsening of their activity and 32.45% had improved. A bivariate analysis was performed between the variable evolution per year and other variables. We didn't observe relationship between the evolution per year and age, sex, foreign, type of uveitis, etiology, laterality, topical etreatment, intravitreal treatment, biological treatment or surgical treatment. Relationship was found statistically significant between the evolution with the existence of complications ($p = 0, 0$) and oral immunosuppressive treatment ($p = 0, 008$). There was statistically significant relationship between the variable worsening with the existence of complications ($p = 0, 0$), topical eye treatment ($p = 0, 035$), oral immunosuppressive treatment ($p = 0, 011$), and not relationship with age was found ($p = 0, 8$), sex ($p = 0,35$), foreign ($p = 0, 27$), type of uveitis ($p = 0, 85$), laterality ($p = 0, 6$) etiology ($p = 0, 1$), periocular treatment ($p = 0, 50$), intravitreal (0.56) and biologic treatment ($p = 0,182$). 65% of the patients did not show any complication a year of follow-up and the most frequent complications were cataract (10%), synechiae (8%), macular edema (5%), glaucoma (3%), detachment of retina (3%), epiretinal membrane (2%) and other (5%).

Conclusion: 87% of the patients were in remission or improvement to the year of follow-up. The worsening of the uveitis is related with the existence of complications and oral immunosuppressive treatment.

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