THU0590

COMPARISON OF FMF PATIENTS WITH AGE OF ONSET BEFORE 20 VERSUS 40 YEARS AND OVER

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Background: Familial Mediterranean fever (FMF) is a disease with an onset before 20 years of age in 90% of the patients. However late onset FMF defined as age of onset over 40 years is being recognised more frequently.

Objectives: To better define patients with FMF who had their first attack before age 40 and compare them with early onset patient group in Turkish population

Methods: The files of 2180 FMF patients followed in a single center between 2008-2017 who have fulfilled Tel-Hashomer criteria, were reviewed with regard to age of onset 40 years and over (index patients, Group 1). For control purposes files before and after the index patients were browsed and first patients with an onset before age 20 years (Group 2) were included. The demographic, clinical and genetic characteristics are compared between these 2 subgroups.

Results: Patients with an onset after 40 years consisted 2.7% of our FMF population. 50 of the 59 patients with an onset 40 years or over were re-evaluated and compared with early onset group consisting of 100 patients (Table 1). The delay in diagnosis, and disease duration were significantly longer and number of patients with M694V homozygosity and M694V allele frequency were significantly more frequent among group 2. In general, phenotypes of both onset groups were similar, the only significant differences being the frequency of fever and myositis which were less common among group 1. Also response to colchicine was more pronounced in group 1. One other interesting observation was the low incidence of amyloidosis in a group with such a significant delay in diagnosis and thus treatment.

Table 1. Demographic, clinical and genetic features of the study groups

	≥40 years n=50	≤20 years n= 100	p
Sex (F:M); present age (mean±SD) (yr)	32:18; 57.2±7.9	62:38; 31.8±9.1	NS;
Mean age at onset,(mean±sd) (yr)	45.6±5.2	8.7±4.8	<0.001
Mean age at diagnosis (mean ±sd) (yr)	50.4±7.3	19.1±11.2	<0.001
Delay in diagnosis (mean ±sd) (yr)	4.8±5.5	10.4±11.8	<0.001
Mean disease duration (mean ±sd) (yr)	11.5±6.4	23.1±10.8	<0.001
Abdominal pain, n (%)	44(88)	89(89.0)	NS
Chest pain, n (%)	7(14.0)	27 (27.0)	NS
Fever, n (%)	30(60.0)	81(81.0)	0.005
Arthritis, n (%)	12(24.0)	33(33.0)	0.25
Myalgia, n (%)	1(2.0)	12(12.0)	0.04
Amyloidosis, n (%)	1(2.0)	3(3.0)	NS
Positive family history, n (%)	33(66.7)	62 (65.2)	NS
Response to colchicine, n (%)	37(82.2)	93 (94.8)	0.014
M694VHomozygous, n (%)	2(4.5)	23(25.8)	0.003
N of M694V alleles	24(48)	82 (82)	0.014
No mutation, n (%)	3(6.8)	2(2.2)	NS

Conclusion: FMF should be included among the differential diagnosis of patients over 40 years of age with recurrent autoinflammatory manifestations. Less than 3% of FMF patients experience their first attacks after 40 years of age. The frequency of M694V is significantly less in the late onset group, pointing out a milder disease.

Disclosure of Interests: None declared

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THU0591

SALIVARY GLAND INVOLVEMENT DISPARITIES IN CLINICAL AND LABORATORY CHARACTERISTICS OF IGG4-RELATED DISEASE: A RETROSPECTIVE STUDY OF 428 PATIENTS

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Background: IgG4 related disease (IgG4-RD) is recently recognized as a fibro-inflammatory condition featured by tumefactive lesions in multiple organs, among which salivary gland is one of the most commonly involved sites. Some previous studies have suggested that salivary gland involved IgG4-RD (IgG4-RD SG+) patients may present different features compared to IgG4-RD patients without salivary gland lesions (IgG4-RD SG-), however, only dozens of patients were examined and the comparison has not been studied in detail.

Objectives: We undertook this study to compare detailed demographic, clinical and laboratory characteristics of IgG4-RD patients with salivary gland lesion (IgG4-RD SG+) and salivary gland free IgG4-RD (IgG4-RD SG-) in a large cohort.

Methods: We carried out a retrospective review of the medical records of 428 cases of IgG4-RD diagnosed at Peking University People's hospital between March 2006 and May 2018.

Results: Among 428 patients, 249 had salivary gland affected. IgG4-RD SG+ patients showed younger age at disease onset and diagnosis, and a longer interval between symptom onset and diagnosis. IgG4-RD SG+ group involved more female patients, and allergic diseases were more common in this group. In terms of organ involvement, IgG4-RD SG+ group were more frequently presented with lacrimal gland involvement, while lymph node, retroperitoneal fibrosis, pancreas, biliary system, kidney, and aorta were more prominent in IgG4-RD SG- group. In addition, the serum IgG4 levels, IgG4/IgG ratio and IgE level were significantly higher in IgG4-RD SG+ patients. Patients with eosinophilia were more common in IgG4-RD SG+ group, while elevated ESR, CRP, and positive ANA were more common in IgG4-RD SG- group.

Conclusion: We have revealed demographic, clinical and laboratory differences between IgG4-RD SG+ and SG- patients, which indicated potential differences in pathogenesis and important implications for the diagnosis and management of these two phenotypes.

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PREDICTORS OF RELAPSE OF IGG4-RELATED DISEASE AFTER INDUCTION THERAPY: A RETROSPECTIVE STUDY

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Background: IgG4-related disease (IgG4-RD) is a systemic fibroinflammatory condition characterized by tumefactive lesions and, often but not always, elevated serum IgG4 levels [1]. Glucocorticoids (GC) are usually used as induction therapy for IgG4-RD. However, relapse often occurs following GC tapering, and predictors of relapse remain unclear.