SAT0589

MUSCULOSKELETAL MANIFESTATIONS OCCUR PREDOMINANTLY IN PATIENTS WITH OLDER ONSET FAMILIAL MEDITERRANEAN FEVER

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Background: Our previous nation-wide survey showed the clinical manifestations and prevalence of Japanese Familial Mediterranean Fever (FMF) patients. However, the clinical differences between young-onset FMF (YOFMF), adult-onset FMF (AOFMF), and late-onset FMF (LOFMF) have not been yet clarified.

Objectives: We sought to compare between the clinical profile of patients with AOFMF, LOFMF and YOFMF and to determine the clinical characteristics of them.

Methods: We enrolled consecutively 395 patients in 2006–2017. Mutation detection in exons 1, 2, 3, and 10 of the *MEFV* gene was performed. We defined YOFMF, AOFMF and LOFMF as the onset of FMF <20, 20–39 and ≥40 years of age, respectively. We compared clinical manifestations and mutations in *MEFV* gene among these three groups.

Results: The median age at the onset of YOFMF, AOFMF and LOFMF were 12.5, 28 and 51 years old respectively. A family history of FMF and a mutation in exon 10 of the *MEFV* gene were significantly more frequent in groups with younger onset ([YOFMF 28%, AOFMF 17%, LOFMF 12%; p<0.01], [YOFMF 51%, AOFMF 33%, LOFMF 19%; p<0.001], respectively). In the accompanying manifestations during the attacks, abdominal pain and chest pain were significantly more frequent in groups with younger onset ([YOFMF 64%, AOFMF 56%, LOFMF 30%; p<0.001], [YOFMF 45%, AOFMF 33%, LOFMF 24%; p<0.01], respectively), whereas arthritis and muscle pain were significantly more frequent in groups with older onset ([YOFMF 32%, AOFMF 48%, LOFMF 62%; p<0.001], [YOFMF 8%, AOFMF 18%, LOFMF 26%; p<0.01], respectively). There was no significant difference in the response to colchicine among the three groups.

	YOFMF (n=182)	AOFMF (n=115)	LOFMF (n=90)	P value		
				YOFMF vs AOFMF vs LOFMF	YOFMF vs Others	Others vs LOFMF
Thoracic pain	82/182(45%)	38/115(33%)	22/90(24%)	0.0025	0.0015	0.0060
Abdominal pain	117/182(64%)	64/115(56%)	27/90(30%)	<0.0001	0.0001	<0.0001
Arthritis	58/182(32%)	55/115(48%)	56/90(62%)	<0.0001	<0.0001	<0.0001
Myalgia	13/161(8%)	16/91(18%)	17/65(26%)	0.0014	0.0012	0.0051
Erysipelas-like erythema	18/182(10%)	16/115(14%)	17/90(19%)	0.11	0.10	0.08
Headache	25/161(16%)	18/91(20%)	10/65(15%)	0.65	0.65	0.85

Conclusions: Our results suggest that older onset FMF had a lower percentage of mutations in exon10 of the *MEFV* gene and predominantly presented arthritis and muscle pain during the attacks. It is thus important to distinguish them from other inflammatory diseases such as gout, adult Still's disease, and infectious arthritis.

Disclosure of Interest: None declared

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SAT0590

LONG-TERM EFFICACY AND SAFETY OF ADALIMUMAB BY AETIOLOGY IN PATIENTS WITH NON-INFECTIOUS UVEITIS IN THE VISUAL III TRIAL

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Objectives: To evaluate the long-term safety and efficacy of adalimumab in patients with non-infectious intermediate, posterior, or panuveitis, across different disease etiologies.

Methods: Adult patients in the Phase III VISUAL I/II trials, who met treatment failure (TF) criteria or who completed the study without TF, were eligible to enter the Phase III open-label extension study, VISUAL III, wherein patients received adalimumab 40 mg every other week. In this exploratory analysis, endpoints were analysed by six uveitis etiologies: i) idiopathic/other (IO), ii) birdshot chorioretinopathy (BCR), iii) multifocal choroiditis and panuveitis (MCP), iv) Vogt-Koyanagi-Harada syndrome (VKH), v) sarcoidosis, vi) Behçet's disease (BD). Endpoints assessed through Week 78 of treatment were proportion of patients with: no active inflammatory lesions in both eyes; anterior chamber (AC) cell grade ≤0.5 + in both eyes; vitreous haze (VH) grade ≤0.5 + in both eyes; and quiescence (defined as no active inflammatory lesions AND AC cell grade ≤0.5 + AND VH grade ≤0.5+). Mean best corrected visual acuity (BCVA) was also assessed. Missing data were reported using non-responder imputation for categorical endpoints and last observation carried forward for continuous variables. Adverse events (AEs) were collected from first adalimumab dose in VISUAL III through the interim cut-off date of Oct 31, 2016.

Results: Of 371 patients included in the intent-to-treat analysis, disease etiology subgroups were (n[%]): IO (155 [41.8%]); BCR (51 [13.7%]); MCP (14 [3.8%]); VKH (72 [19.4%]); Sarcoidosis (52 [14.0%]); BD (27 [7.3%]). The proportion of patients achieving quiescence improved over time in all uveitis etiologic subgroups: IO 28.4%/63.9% (Week 0/Week 78), BCR 43.1%/64.7%, MCP 35.7%/50.0%, VKH 37.5%/63.9%, sarcoidosis 32.7%/69.2%, and BD 44.4%/74.1%. The proportions of patients with no active inflammatory lesions, AC cell grade ≤0.5+, or VH grade ≤0.5+, showed moderate increases or decreases between Weeks 0 and 78 within specific etiology subgroups; percentage ranges across all subgroups at Weeks 0/78, were 60.8–87.5/57.1–78.8% (no active inflammatory lesions), 47.2–92.2/50.0–77.8% (AC cell grade ≤0.5+), and 42.9–72.2/57.1–77.8% (VH grade ≤0.5+), respectively. Improvement or maintenance of mean BCVA was observed over time across the etiologic subgroups. AE rates were consistent with the VISUAL I and II trials.

Conclusions: Adalimumab increased the percentage of patients achieving quiescence and improved or maintained visual acuity at Week 78 of the VISUAL III study, with consistent results across uveitis etiologic subgroups. No new safety signals were detected with long-term adalimumab treatment.

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SAT0591

DEMOGRAPHIC AND CLINICAL FEATURES OF FAMILIAL MEDITERRANEAN FEVER PATIENTS

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Background: Familial Mediterranean Fever (FMF) is an autosomal recessive genetic disorder that causes recurrent episodes of fever, polyserositis, arthritis, skin eruptions.

Objectives: In this study, we aim to present clinical and demographic features of FMF patients followed up in our clinic.

Methods: The clinical, demographic, genetic features and managements of 402 FMF patients (fulfilling Tel-Hashomer Diagnostic Criteria) were analysed.

Results: The mean age was 36.8±11.2 years, mean diagnosis age was 28±11.9 years, and mean disease duration was 189.2±124.5 months, mean duration between onset of disease and onset of treatment was 93.6±104 months. Consanguineous marriage was detected in 7%²⁹ patients. Fever and abdominal pain both were initial symptoms in 72% of the patients, while 7% of them had chest pain, 4% had only fever, 15% had arthritis, 1% had erysipelas-like erythema and 1.5% had inflammatory back pain as the first symptom of FMF (table 1).

Eight patients (2%) were suffered from chronic kidney disease and 2 of them were on dialysis programme. Amyloidosis were identified in 14 patients (3.5%) with biopsy.

At least one mutation of *MEFV* gene was detected in 78% patients There was no mutation in 8% patients. In 15% patients, *MEFV* gene analysis could not be done. The most frequent mutation was M694V mutation and its allele frequency was 54%; the frequency of V726A, M680I, E148Q, R761H and A744S allele mutation

were 11%, 7%, 7%, 2%, 1%, respectively. The frequency of compound mutation was 38%, and the most common compound mutation was M694V+R202Q (11%). There was significantly relationship between M694V mutation and arthritis, erysip-
pel-like erythema, proteinuria, sacroiliitis (table 2). Amyloidosis was more frequent in patients who had M694V homozygous mutation. Mean age of disease onset was lower in patient who had M694V homozygous mutation than M694V heterozygous mutation ($p<0.001$).

Abstract SAT0591 – Table 1. Demographical and Clinical Features of FMF Patients

n=402	n(%)
Gender Female	241 (60)
Male	161 (40)
Fever	299 (75.5)
Abdominal Pain	347 (86)
Erysipel-like erythema	54 ⁽¹³⁾
Chest pain	83 (21)
Positive family history	174 (43)
Appendectomy	96 ⁽²⁴⁾
Amyloidosis	14 (3.5)
Chronic kidney disease	8 (2)
Inflammatory back pain	63 ⁽¹⁶⁾
Arthritis	129 (32)
Hip pain	27 (7)
Heel pain	30 (7.5)
Uveitis	4 (1)
Sacroiliitis	44 ⁽¹¹⁾
Biological treatment	16 ⁽⁴⁾

Abstract SAT0591 – Table 2. Relationship between M694V mutation and clinical findings

N(%)	Negative	Heterozygous	Homozygous	P
Fever	75 (60)	80(62)	67(75)	0,0503
Abdominal pain	107(85,6)	107(82,9)	79(88,8)	0,487
Erysipel-like erythema	5 (4)	13(10,1)	19(21,3)	0,0003
Chest pain	21(16,8)	24(18,6)	20(22,5)	0,0575
Myalgia	2(1,6)	5(3,9)	3(3,4)	0,535
Arthritis	32(25,6)	34(26,4)	41(46,1)	0,002
Sacroiliitis	11(8,8)	12(9,3)	19(21,3)	0,0096
Amyloidosis	4(3,2)	0(0)	10(11,2)	N/A
Proteinuria	7(5,6)	5(3,9)	18(20,2)	0,000

Conclusions: Tight control and sustained management are important in FMF to protection from amyloidosis. Similar to literature, the most frequent mutation was M694V mutation, and there was significantly relationship between M694V mutation and arthritis, erysip-
pel-like erythema⁽¹⁾, proteinuria, sacroiliitis⁽²⁾ in our study.

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SAT0592 HRCT PULMONARY MANIFESTATIONS IN PATIENTS WITH SYNOVITIS, ACNE, PUSTULOSIS, HYPEROSTOSIS, AND OSTEITIS (SAPHO) SYNDROME

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Background: Synovitis, acne, palmoplantar pustulosis, hyperostosis and osteitis syndrome (SAPHO) is a rare syndrome that affects the skin, bones and joints.¹ SAPHO syndrome shares a variety of features with the seronegative spondyloarthropathies (SpAs), such as psoriatic arthritis (PsA).¹ While the extra-articular manifestations of PsA are well defined, the systemic involvements of SAPHO syndrome are only reported occasionally.² Previous cases of accompanied pleural effusion³,⁴ and organising pneumonia⁵ were reported. In clinical work, we observed that pulmonary lesions progressed when diseases aggravated.

However, the relationship between pulmonary manifestations and diseases activity remains unclear.

Objectives: This study is to explore the pulmonary comorbidity with SAPHO syndrome.

Methods: Pulmonary HRCT images were reconstructed from whole-spine CT images in 67 SAPHO patients. HRCT images of 58 healthy controls were obtained and reviewed. Pneumonia and tuberculosis were excluded. We investigated the presence of different HRCT images findings with detailed examinations.

Results: In detailed HRCT evaluations, abnormalities were identified in 45 of all patients. We found stripe in 29 (43.3%) cases, patchy shadows in 22 (32.8%), ground-glass opacity in 11 (16.4%), pleural thickening in 9 (13.4%), solitary nodule in 6 (9%), bronchiectasis in 3 (4.5%), pulmonary bulla in 2 (3%), multiple nodules in 1 (1.5%), and interstitial change in 1 (1.5%). Compared with healthy controls, SAPHO patients have significantly higher rate of patchy shadows while significantly lower percentage of nodules (especially multiple nodules), although the overall rates of abnormal HRCT findings are similar.

Abstract SAT0592 – Figure 1. HRCT images in 3 SAPHO patients. A: Ground glass opacities (arrows); B: Solitary nodule (arrow); C: Interstitial change (arrow).

Conclusions: Our study was the first to study HRCT pulmonary changes in SAPHO patients. SAPHO patients have significantly higher percentage of patchy shadows and significantly lower rate of pulmonary nodules than healthy controls. BASDAI and age are possible good predictors for abnormal HRCT pulmonary findings.

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SAT0593 PROGRESSIVE FIBROSING INTERSTITIAL LUNG DISEASE (PF-ILD) IN PATIENTS WITH AUTOIMMUNE RHEUMATIC DISEASES

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Background: Interstitial lung disease (ILD) associated with autoimmune conditions is among the most challenging aspect of care for patients with rheumatic diseases. While idiopathic pulmonary fibrosis (IPF) is the classic fibrosing ILD, some patients with differing clinical ILD diagnoses including autoimmune associated-ILD can develop a progressive fibrosing phenotype. This phenotype is characterised by progressive pulmonary fibrosis, worsening respiratory symptoms, declining lung function, resistance to immunomodulatory therapies and early mortality. There are limited data available on current practice in diagnosis, management and treatment of PF-ILD in patients with autoimmune rheumatic diseases.

Objectives: To investigate the patient journey in patients with autoimmune rheumatic diseases and PF-ILD.

Methods: Twenty-two ILD experts from Germany, Japan, UK and the US participated in a 1 hour interview. Physicians who spend $\geq 75\%$ of their professional time managing patients and in whose caseload ≥ 10 patients had PF-ILD in the