THU0548 PSORIASIS IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER

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Background: Familial Mediterranean fever (FMF) is a periodic fever syndrome caused by *MEFV* mutations. FMF may be associated with psoriasis in some cases. Previous study has shown that psoriasis was more common in the relatives of FMF patients [1].
Objectives: We aimed to investigate the prevalence of psoriasis among FMF patients and their relatives.
Methods: FMF patients followed at Hacettepe University Adult and Pediatric Rheumatology Departments between January and August 2016 were consecutively enrolled to this study. Demographic data, clinical manifestations, laboratory data and *MEFV* variant analysis were documented by medical file screening and face-to-face interview. The presence of psoriasis and psoriatic arthritis in patients and their relatives (first [Mother, father, children]-second [Brothers, grandchildren, grandfather and grandmother]-third degree [Nephew, uncle, maternal uncle, aunt, paternal aunt] relatives) and drug use history were also questioned. The patients were accepted to have psoriasis if the diagnosis was made by a dermatologist.
Results: 351 FMF patients (177 adults; 174 children) were included in this study (Table). 70.1% of adult patients were female, 29.9% were male. 53.4% of pediatric patients were female, 46.6% were male. The median age (min-max) of the adult patients was 35 (19–63), while the median age of the pediatric patient group was 10 (2–18). The onset age of symptom was 12 (0–39) in the adult group and 3 (1–14) in the pediatric group. The median age at diagnosis was 25 (2–52) in the adult group and 5 (1–18) in the pediatric group. Thirteen (3.7%) patients had psoriasis. Psoriasis was more common in adult patients than pediatric patients (p=0.02). Psoriasis was present in 22 (12.4%) of adult patients' and 9 (5.2%) of pediatric patients' relatives (p=0.023). The frequency of psoriasis in one or more relatives of all FMF patients was found to be 8.8%.

Table. Demographic and clinical characteristics of 177 adult and 174 pediatric patients with familial Mediterranean fever (FMF)

Characteristics	Adult patients (n=177)	Pediatric patients (n=174)	p value
Gender, female, n (%)	124 (70.1)	93 (53.4)	**0.001**
Abdominal pain, n (%)	159 (89.8)	166 (95.4)	**0.046**
Fever, n (%)	152 (85.9)	170 (97.7)	**<0.0001**
Arthralgia, n (%)	147 (83.1)	76 (43.7)	**<0.0001**
Arthritis, n (%)	93 (52.5)	27 (15.5)	**<0.0001**
Pleuritic chest pain, n (%)	104 (58.8)	14 (8)	**<0.0001**
Erysipelas-like erythema, n (%)	57 (32.2)	6 (3.4)	**<0.0001**
Pericarditis, n (%)	7 (4)	0 (0)	**0.007**
Amyloidosis, n (%)	5 (2.8)	1 (0.6)	0.215
Family history of FMF[a], n (%)	99 (55.9)	94 (54)	0.719
Parental consanguinity, n (%)	44 (25)	25 (14.4)	0.012
Hemodialysis history in family associated with FMF, n (%)	20 (11.3)	11 (6.3)	0.132
Psoriasis, n (%)	11 (6.2)	2 (1.1)	**0.020**
Psoriatic arthritis, n (%)	3 (1.7)	1 (0.6)	0.623
Psoriasis in any degree relatives, n (%)	22 (12.4)	9 (5.2)	**0.023**
Psoriasis in 1° relatives, n (%)	5 (2.8)	2 (1.1)	0.44
Psoriasis in 2° relatives, n (%)	5 (2.8)	6 (3.4)	0.73
Psoriasis in 3° relatives, n (%)	15 (8.5)	1 (0.6)	**<0.0001**

Conclusions: IL-1 has an essential role for signaling early T helper 17 (Th17) differentiation and Ashida et al have shown the presence of Th17 cells in the upper dermis of psoriasis-like lesions in a patient with FMF [2]. We may speculate that high IL-1 in FMF may cause Th17 activation and stimulation of keratinocytes; and this may be the reason for higher frequency of psoriasis in FMF patients. Thirteen (3.7%) patients had psoriasis; more common than the normal population (0.40%) (p<0.0001). FMF increases the likelihood of psoriasis in relatives of FMF patient. Thus, FMF patients should be questioned and carefully examined for psoriasis lesions and psoriasis family history.
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THU0549 SYSTEMIC TREATMENT FOR ACUTE ANTERIOR UVEITIS (SYNTHETIC AND BIOLOGIC DISEASE-MODIFYING ANTIRHEUMATIC DRUGS): A SYSTEMATIC REVIEW

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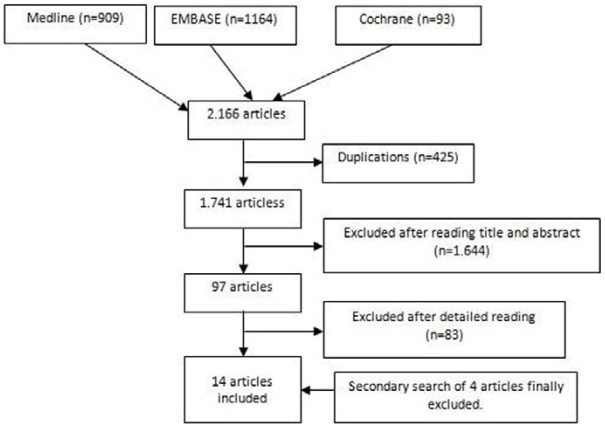
Background: Acute anterior uveitis (AAU) is the most common form of uveitis. Most of them are idiopathic, followed by those related to rheumatic conditions. One third of AAU patients may present recurrences, some requiring systemic disease-modifying antirheumatic drugs (DMARDs). The use of DMARDs in AAU is heterogeneous.
Objectives: To perform a systematic and critical review of the literature about the use of synthetic and biologic DMARDs in adult patients with AAU.
Methods: *Selection criteria:* Articles including adult patients with non-infectious AU treated with synthetic or biologic DMARDs including efficacy, and/or safety or cost-effectivity data were selected. Only meta-analysis, systematic reviews, clinical trials and observational studies (OS) were included.
Search strategies for Medline, Embase and Cochrane Library databases up to 3–2016 were designed.
Article selection: 2 independent reviewers. Selected articles were analyzed in detail.
Quality asessment of the studies: Oxford scale and Jadad scale were used.
Analysis and data presentation: evidence and results tables.
Results: 14 articles included, 2 RCTs and 12 OSs, with low or moderate quality. The mean duration/follow-up, number (n) and patients characteristics were highly variable. The definition of the anatomic classification of AUs was generally not clear. Systemic DMARDs were used, including Methotrexate (MTX), Azathioprine (AZA), Cyclosporine A (CsA) and anti-TNFα (Adalimumab (ADA), Golimumab (GLM)), at usual dosage prescription. Number of flares, disease activity and corticoid sparing (CS) effect were the most common outcomes, with big differences between studies in variables included and their definitions.
MTX showed efficacy in disease remission, n of flares, time between flares, lower activity and CS effect. *SSZ* showed lower n of flares and improvement in visual acuity (VA) in AS-associated AAU patients. *AZA* (low quality RCT) showed no differences in VA, Tyndall, flares or IOP. A prospective OS showed lower activity and CS effect. *CsA* (moderate quality OS) showed efficacy improving activity and as CS agent (mid/long term). *Anti-TNFα: ADA*, (2 OSs) with SpA-associated AU patients lowered n of flares (mid/long term), can improve VA, Tyndall and be used as CS agent. *GLM* in AU patients refractory to DMARDs (some to other biologics), showed CS effect in 2 studies. One showed improvement in VA and Tyndall, but not in OCT or n of flares. Adverse events recorded were those usually registered for all these drugs.

Figure 1. Article selection strategy.



Conclusions:
1. Evidence quality is low.
2. Great variability.
3. MTX showed efficacy in idiopathic and systemic disease-associated (SDA) AU.(EL 2c; RG B).
4. SSZ showed efficacy in idiopathic and SDA AU.(EL 3a; RG B-C).
5. AZA seems to be effective in naïve and DMARDs-refractory AU (EL 3a; RG C).
6. CsA showed efficacy in idiopathic and SDA AU (EL 2c; RG B-C).
7. ADA showed efficacy in idiopathic and SDA AU, naïve or DMARDs-refractory AU (EL 2c; RG B).

8. GLM showed efficacy in DMARD-refractory AU (2nd line and further) and other biologic therapies (EL 3a; RG B-C).
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THU0550 TREATMENT AND OUTCOMES IN SPANISH PATIENTS WITH IGG4-RELATED DISEASE

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Background: There is a lack of strong evidence on IgG4-related disease (IgG4-RD) treatment. There is only one clinical trial published, supporting the use of rituximab in American patients.
Objectives: To describe the treatments used in a series of patients diagnosed with IgG4-RD in Spain and to review the outcomes.
Methods: Clinical data were obtained from the Spanish Registry of IgG4-RD from October 2013 to January 2016, including 14 centers. Outcomes were assessed by a self-made response scale and the IgG4 responder index (RI). We categorized the outcomes as a total response (disappearance of the pseudotumoral lesions and absence of symptoms), partial response (<50% regression of the tumefactive lesions or persistence of inflammation without symptoms) and no response if no changes were noticed. Treatment failure was considered if an increase of the activity, mass size or reappearance of symptoms were noticed among patients under treatment.
Results: Sixty-eight patients were included. Twenty-six (38%) were females, mean age 53.4 years. Thirty-six patients (52.9%) had systemic IgG4-RD involving >1 tissue. The most commonly involved tissues were: retroperitoneum (33%), orbital pseudotumor (28%), and maxillary and paranasal sinuses (24%). The main treatments used were: steroids (90%), surgery (45%) and azathioprine (19%).
All treatments were successful in achieving complete or partial response. The mean pre and post-treatment RI values were 6.7 (SD 4.6) and 1.9 (SD 2.6) respectively. There were no differences between systemic and non-systemic disease regarding the chosen treatments and the outcomes. The combination azathioprine-steroids was used in 12 patients. Fourteen percent of them relapsed (considering relapse as an increase of the inflammation, mass size or reappearance of symptoms, since the first month after the treatment withdrawal). The treatment failed in 28.6% of them. The combination steroids-rituximab was indicated in 6 patients, showing no relapses and 1 treatment failure. The majority of patients treated with azathioprine or rituximab combined with steroids had a systemic disease (6.6 and 80%, respectively). Nearby all of them had already failed other previous treatments.
Conclusions: In our series, IgG4-RD has been treated with a myriad of drugs and procedures. The outcomes have been acceptable but the disease tended to relapse (21%) and the treatment failures were common (27%), probably due to the lack of well-defined treatment schemes supported by solid studies. Steroids were still the cornerstone of the treatment. Rituximab results were promising in our study but the number of patients was limited. Azathioprine, in combination with steroids, may be an accessible alternative treatment for IgG4-RD that should be explored. The RI correlated with the treatment outcomes and will have an important role monitoring future studies on IgG4-RD therapies.
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THU0551 ARTHRITIS IN SARCOIDOSIS - A MULTI-CENTRE STUDY FROM INDIA

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Background: 10–15% of sarcoid patients have associated arthropathy. Chronic arthritis is less common varying from 1–2% [1].Data on articular manifestations of the disease from India is sparse.[2]
Objectives: To study the clinical manifestations of sarcoid arthritis patients from India.
Methods: Case records of patients presenting to ten rheumatology centres from 2005 to 2016 with sarcoidosis were retrospectively reviewed. Joint involvement was assessed clinically, classified as acute or chronic depending on duration of symptoms lesser or greater than 6 months respectively.
Results: A total of 103 patients with sarcoid arthritis were reviewed. 58 patients were classified as Lofgren syndrome. Pattern of joint involvement revealed ankle as most commonly affected in both the groups (Table1). Wrist, MTP and PIP involvement was significantly more common in chronic sarcoid arthritis (table 1). Peripheral lymphadenopathy, plaques and uveitis were more frequent ($p<0.05$) in chronic sarcoid arthritis (50%, 20.8%, 25% respectively) compared to those with acute sarcoid arthritis (16.5%,6.3%,6.4% respectively). 45 of 62 patients with acute arthritis with follow up details had achieved complete remission.15/24 patients with chronic sarcoid arthritis patients with a median follow up of 2 years had achieved complete remission with 14, 11 and 5 patients on steroids, methotrexate and hydroxychloroquine respectively. One patient with concomitant interstitial lung disease had died due to lung infection.

Table 1. Clinical characteristics of patients with sarcoid arthritis from India

	Acute, n=79	Chronic, n=24
Age (years)	40† (33.5 TO 51)**	42.5† (35–55)**
Male : Female	0.9:1	1.7:1
Duration of Symptoms (years)	0.25† (0.08 to 1.5)**	0.72† (0.5 to 4)**
Oligoarthritis (≤4 joints)	58 (73.4)*#	12 (50)*
Polyarthritis (≥5 joints)	19 (24.1)*	12 (50)*
Ankle	71 (89.9)*	21 (87.5)*
Wrist	22 (27.8)*	16 (66.7)*‡
MTP	5 (6.3)*	5 (20.8)*‡
PIP	12 (15.2)*	9 (37.5)*‡
Hilar/Mediastinal Lymphadenopathy	66 (83.5)*	21 (87.5)*
ILD	23 (29.1)*	12 (50)*

*Percentage; †Median; **IQR. ‡$p<0.05$. #One patient each had dactylitis and enthesitis as the only musculoskeletal manifestation.

Conclusions: Acute oligoarthritis was the commonest presentation with ankle most commonly affected joint. Wrist, PIP, MTP involvement were more common in chronic sarcoidosis. One of the limitation was retrospective analysis.
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THU0552 COMPLIANCE TO COLCHICINE TREATMENT IN FAMILIAL MEDITTERRANEAN FEVER RELATED AMYLOIDOSIS

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Objectives: To assess the colchicine compliance in patients diagnosed with Familial Mediterrenean Fever (FMF) related amyloidosis in our center.
Methods: Fourty one patients (18 male/23 female) were questioned with regards to colchicine compliance by using retrospective scanning of patient data.
Results: The mean age at the symptomatic onset of FMF was 7.13±5.24 years. The mean age at the time of the FMF diagnosis was 21.21±14.85 years. The mean age at the initiation of colchicine treatment was 21.42±14.75 years and the mean age at time of the diagnosis of amyloidosis was found 29.57±12.14 years. Mean duration of the disease was 31.702±11.84 years and the duration of delayed diagnosis was 14.35±13.84 years. Maximum dose of colchicine was 2,103±0,673 mg/day. Compliance of colchicine treatment was poor in FMF related amyloidosis during their follow-up (11/25, %44), rates of skipped doses were also high (17/25, %68). Compliance rates were high in patients in whom FMF and amyloidosis were diagnosed simultaneously (12/13, %93), rates of skipped doses were also low (2/13, %14). One of the patients diagnosed with FMF after the diagnosis of amyloidosis was compliant, two of them were non-compliant; with regards to skipping doses, two patients were found to be compliant and therefore never skipped doses while one was skipping doses. The compliance to colchicine was high in all FMF patients once amyloidosis was evident (31/41, %75), and rates of skipped doses were also low (12/41, %30). In five FMF patients, amyloidosis was observed despite their compliance to treatment.
Conclusions: The overall delay of diagnosis in FMF patients with amyloidosis was found to be high. Particularly the FMF patients who were diagnosed with amyloidosis during their follow up were found to have lower rates of compliance. There were also a group of patients who were diagnosed with amyloidosis despite adequate and convenient colchicine treatment. It was emphasized that not only the early diagnosis and adequate treatment, but also the close follow up is important in managing FMF patients.
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