

variant of unknown significance, but it may have contributed to the DADA2 associated increased risk of amyloidosis. A better response of proteinuria to adalimumab treatment indicates superiority of anti-TNFs in DADA2 patients compared to anti-IL-1 drugs.

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AB1301

DETERMINING THE RELATIONSHIP BETWEEN SERUM INTERLEUKIN 33 LEVELS AND CLINICAL FEATURES OF THE DISEASE IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER

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Background: Familial Mediterranean Fever (FMF) is an autoinflammatory disease characterized by recurrent fever, serositis, arthritis and erysipelas-like erythema caused by mutations over activating caspase-1. As Interleukin (IL)-1 beta, IL-33 is a nuclear cytokine from IL-1 family which is activated by caspase-1. IL-33 is known to take part in pathogenesis of several rheumatic diseases.

Objectives: The aim of this research is determining the relationship between serum IL-33 levels and clinical features of the disease in patients with FMF disease.

Methods: The research involved 54 FMF patients and 29 healthy volunteers. Serum IL-33 levels were evaluated in both patients and healthy individuals, and its relationship between clinical and laboratory features of FMF.

Results: 28 out of 54 patients (%51.8) had favorable response to colchicine while 26 patients (%48.2) had colchicine resistant disease. FMF patients had lower IL-33 levels compared to healthy control group ($p = 0.06$). There were no difference between colchicine responsive and resistant patients ($p = 0.12$) and no association was found between clinical features and serum IL-33 levels. Additionally, IL-33 did not correlated with C-reactive protein and disease activity assessed by autoinflammatory disease activity index.

Conclusion: No association was found between serum IL-33 levels and FMF disease features and laboratory findings. This may be due to the small size of our patient group, the involvement of IL-33 in tissue homeostasis as well as inflammation, and the use of higher doses of colchicine in the resistant disease group than in the remission group. Additional research is needed to determine IL-33's role in FMF pathogenesis and its relationship with clinical and laboratory features.

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AB1302

EVALUATING THE CLINICAL UTILITY OF PATIENT ACCEPTABLE SYMPTOM STATE IN PATIENTS WITH FAMILIAL MEDITERRANEAN FEVER

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Background: Familial Mediterranean Fever (FMF) is an autoinflammatory disease characterized by recurrent attacks of fever, serositis, arthritis and erysipelas-like erythema. Patient acceptable symptom state (PASS) is a disease evaluation method to assess disease activity with a simple question especially in rheumatic diseases.

Objectives: We aimed to investigate clinical utility of PASS in FMF patients.

Methods: The research involved 54 FMF patients. Patient acceptable symptom state was applied to all patients in the study. The answers to PASS were compared with the patients clinical and laboratory features.

Results: 28 out of 54 patients (51.8%) were colchicine responsive whereas, 26 patients (48.2%) had colchicine resistant disease. The number of patients who answered yes to PASS (I'm happy with my current disease condition) was 32 (59%), while answered no (I need further treatment options) was 22 (41%). Considering the disease severity assessed with International severity scoring FMF (ISSF) of those who answered yes, 22 (68%) patients had mild disease, 10 patients had moderate (32%) disease, and there was no patient with severe disease in this group. Among those who answered no, 3 (14%) had mild disease, 14

(63%) had moderate disease, and 5 (23%) had severe disease ($p < 0.001$). When the CRP levels of the patients were compared, the median CRP value of those who answered yes was found to be 4.45 mg/L, and the median value of CRP for those who answered no was 11.25 mg/L ($p = 0.04$).

Sensitivity and specificity of PASS for detecting patients in remission was 78% and 61% respectively. Moreover, PASS had a positive and negative predictive value of %68 and %72 respectively, for determining patients in remission. If cut off level of CRP was chosen as 6.5 mg/L for answering "yes" to PASS, sensitivity of test has been found to be 62.5% while the specificity is 59.1%. On the other hand, if cut off level of CRP is selected as 9.35 mg/L; sensitivity and specificity of the test was found as 75% and 72.7% respectively ($p = 0.045$).

Conclusion: Patient acceptable symptom state is found beneficial in evaluation these patients simply and swiftly especially in terms of distinguishing severe FMF disease. In FMF, laboratory remission is as important as clinical remission, therefore, PASS by alone, is not sufficient for making treatment decisions and should be supported by inflammatory markers.

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AB1303

VACCINATION PRACTICES OF ADULT FAMILIAL MEDITERRANEAN FEVER PATIENTS IN TURKEY.

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Background: Vaccines are the safest and most effective method to prevent invasive and life-threatening infections. Vaccines against influenza, pneumococcal disease, herpes zoster, and human papillomavirus are the main recommended vaccines for adults. In addition, rheumatology patients are advised to receive adult vaccinations according to the vaccines available in their country and local guidelines. In Turkey, both influenza and pneumococcal disease vaccines are commercially available. In addition, these vaccines are strongly recommended for rheumatology patients in local guidelines. Although familial Mediterranean fever (FMF) is one of the most common rheumatological diseases in Turkey, it is often neglected in vaccination recommendations.

Objectives: In this study, we surveyed the vaccination practice against influenza or pneumococcal diseases of adult FMF patients in our cohort. In addition, we evaluated the factors related to favorable vaccination practice.

Methods: We included 360 FMF patients over 18 years of age. All patients fulfilled the Tel-Hashomer criteria for FMF. We asked them if they had ever been vaccinated against pneumococcal or influenza, and how often they received them. In addition, we dichotomised patients in terms of vaccinated against at least one of influenza or pneumococcal diseases. We then compared the groups for demographic (age, gender and comorbidities) and disease related characteristics (disease duration, disease activity calculated by ISSF and colchicine dose). We used chi-square test to compare categorical variables and Mann-Whitney U test to compare continuous variables. $P < 0.05$ was accepted as significant.

Results: Of 360 FMF patients, 238 (66.1%) were female. The mean age of the patients was 34.5 ± 10.7 years. Disease duration of the patients was 9.38 ± 0.7 years. In addition, the mean ISSF score of the patients was 1.83 ± 1.5. The mean dose of colchicine received by the patient was 1.23 ± 0.47 mg. Only 54 (15.0%) of the patients had at least one comorbidity. In our cohort, 22 (6.1%) patients were vaccinated against influenza or pneumococcal disease. Only 18 (5.0%) of the patients have been vaccinated against influenza at least once so far. Half of these patients (9/18) were vaccinated against influenza each year. In addition, 8/360 (2.2%) patients were fully vaccinated against pneumococcal diseases. Here, six of them received the pneumococcal vaccine after the start of the COVID-19 outbreak. There was no statistically significant difference between the groups in terms of demographic and disease related characteristics.

Conclusion: We found that vaccination practice of FMF patients in our cohort was unsatisfactory. Few patients follow adult vaccination recommendations. In addition, clinicians should be concerned about the importance of vaccination and guide their patients to get the adult vaccines available in their country.

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AB1304

COMPARISON OF CHARACTERISTICS OF PATIENTS WITH IDIOPATHIC GRANULOMATOUS MASTITIS IN REMISSION AND NON-REMISSION: RESULTS FROM A TERTIARY REFERRAL CENTRE

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Background: Idiopathic granulomatous mastitis (IGM) is an uncommon, benign, chronic inflammatory disease of the breast of unknown etiology. Although an ideal treatment regimen is controversial in IGM, the ultimate goal is remission.

Objectives: To compare the clinical, radiological, laboratory and treatment features of patients in remission and non-remission, and assess predictor factors of remission.

Methods: The data of patients who were histopathological diagnosed with IGM between 2010 and 2020 were evaluated retrospectively. The patients were divided into 2 groups as those in complete remission and non-remission. Complete improvement in clinical and physical examination with medical and/or surgical intervention findings at 2 consecutive visits was defined as complete remission. Patients who were not in complete remission including partial remission and refractory to treatment, were defined as non-remission group.

Results: In this study, there were a total of 103 patients with a diagnosis of IGM followed-up in the rheumatology clinic. Of these, 39 (38%) were in remission and 64 (62%) were non-remission. Age, age at symptom onset, comorbidity, pregnancy (ever), disease localization, clinical signs and symptoms, imaging classification, and baseline acute phase reactants and autoimmune markers were similar in the remission and non-remission groups. The use of antibiotics and azathioprine was significantly higher in the remission group than in the non-remission group. However, there was no difference between the groups in terms of breast surgery, use of corticosteroids and methotrexate (Table 1). In the

univariable analysis, antibiotic use [OR: 2.55 (95% CI 1.07-6.11)] was found to be a predictor factor of remission.

Conclusion: Patients with in remission did not have the distinctive baseline demographic, clinical, laboratory and radiological features that distinguish them from patients with in non-remission. Patients in remission used more antibiotics and azathioprine. Antibiotic use seems to be associated with remission in univariable analysis. However, further studies are needed to evaluate the factors associated with remission in IGM.

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AB1305

A SYSTEMATIC REVIEW OF AA AMYLOIDOSIS AMONG PATIENTS WITH BEHÇET'S SYNDROME

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Background: Data on patients with Behçet's syndrome (BS) complicated with AA amyloidosis is limited to case reports or case series with a small number of patients.

Objectives: In this study, we aimed to perform a systematic review (SR) of published reports on BS patients with AA amyloidosis.

Methods: PubMed and EMBASE were searched with the keywords "Behçet AND amyloidosis", without date and language restriction, until May 2020. Two independent reviewers (SNE, GK) performed title/abstract and full text screening and data extraction. A third reviewer (GH) made the final decision in case of disagreement between the two reviewers. Studies that reported patients who were reported by authors as having BS and AA amyloidosis were included. The risk of bias assessment was done using the Grading of Recommendations Assessment, Development and Evaluation (GRADE) tool.

Results: The systematic literature search yielded 760 articles of which 703 were excluded after title and abstract review. After full-text review, we further excluded 15 duplicate articles and 1 article was added after handsearching the reference lists of the full texts. Finally, we included 43 articles reporting 96 cases. Among these articles, 38 were case reports and 5 were case series reporting between 6 and 14 patients. All patients but 8 were reported from Mediterranean countries.

Table 1. Characteristics of patients in remission and non-remission

	All patients n= 103	Patients in remission n = 39	Patients not in remission n= 64	p value
Age, years	38.1 ± 7.1	39.6 ± 7.8	37.2 ± 6.5	0.07
Age at symptom onset, years	33.5 ± 6.6	34.4 ± 7.3	33 ± 6.1	0.30
Comorbidity, n (%)				
Thyroid disorders	4 (3.9)	1 (2.6)	3 (4.7)	1.00
Hypertension	2 (2)	2 (5)	0	0.14
Diabetes mellitus	1 (1)	1 (2.6)	0	0.38
Pregnancy (ever), positive/total (%)	75/77 (97.4)	31/32 (97)	44/45 (98)	1.00
Breast localization, n (%)				
Left	47 (45.6)	19 (48.7)	28 (43.8)	0.88
Right	36 (35)	13 (33.3)	23 (35.9)	
Bilateral	20 (19.4)	7 (17.9)	13 (20.3)	
Clinical signs and symptoms, n (%)				
Palpable mass	94 (91.3)	35 (89.7)	59 (92.2)	0.72
Fistula formation	37 (38.1)	16 (42.1)	21 (35.6)	0.52
Erythema	61 (65.6)	23 (65.7)	38 (65.5)	0.98
Nipple inversion	11 (12)	2 (5.4)	9 (16.4)	0.19
Axillary lymph node	45 (48)	17 (47.2)	28 (48.3)	1.00
Erythema nodosum	10 (9.9)	4 (10.3)	6 (9.7)	1.00
Arthralgia	13 (12.6)	3 (7.7)	10 (15.6)	0.36
BI-RADS classification*, n (%)				
2	13 (25)	7 (35)	6 (18.8)	0.12
3	31 (59.6)	9 (45)	22 (68.8)	
4	6 (11.5)	2 (10)	4 (12.5)	
5	2 (3.8)	2 (10)	0	
Baseline laboratory				
-CRP (mg/dL)	0.75 (1.4)	0.75 (1.4)	0.73 (1.4)	0.77
- ESR (mm/h)	23 (28)	21.5 (28)	25.5 (28)	0.39
-ANA (more than or equal 1/100), n ^a (%)	37/69 (53.6)	12/25 (48)	25/44 (56.8)	0.48
-RF, n ^a (%)	12/30 (40)	5/16 (45.5)	7/19 (36.8)	0.71
-anti-CCP, n ^a (%)	3/12 (25)	2/5 (40)	1/7 (14)	0.52
Treatment, n (%)				
Surgical intervention	38 (36.9)	16 (41)	22 (34.4)	0.49
Antibiotics	63 (61.2)	29 (74.4)	34 (53.1)	0.03
Corticosteroids	93 (90.3)	34 (87.2)	59 (92.2)	0.49
Methotrexate	66 (64.1)	26 (66.7)	40 (62.5)	0.67
Azathioprine	19 (18.4)	11 (28.2)	8 (12.5)	0.04

* 52 patients with data were evaluated. ^a positive/total

Results are given as mean ± SD or median (IQR).

ANA: Antinuclear antibody; anti-CCP: Anti-cyclic citrullinated peptides; BI-RADS: Breast imaging-reporting and data system; RF: Rheumatoid factor