Objectives: To assess the frequency of depression and anxiety in patients with Familial Mediterranean Fever (FMF)

Methods: In this study, 77 FMF patients aged 18 and over who were followed up in Sakarya University Education and Research Hospital, Department of Rheumatology, and 78 healthy volunteers aged 18 and over as the control group. Beck-depression scale and Beck anxiety scale were used to detect depression and anxiety, respectively. Beck’s depression scale was evaluated as 9 and below normal, 10-16 mild depression, 17-29 moderate depression, 30-63 severe depression. Beck anxiety scale was evaluated as 0-8 normal, 8-15 mild anxiety, 16-25 moderate anxiety, 26 and above severe anxiety. FMF disease severity was determined by Pras scoring.

Results: The study group comprised 77 diagnosed with FMF with a range of 37.18 and a control group comprised of 78 healthy controls (C) with a meanage of 35.32 (p=0.058). In study group (P) %63.6, control group (C) %53.8 as female. %36.4 of the study group (P), %46.2 of the control group are male. (p=0.0216). The prevalence of depression was significantly higher in FMF patients compared to the control group (in order P:C normal %24.7, %47.4, mild depression: %40.3, %26.9, moderate depression %26, %19.2, severe depression %11.7, %6.4 (p<0.015). Similarly in depression results; the prevalence of anxiety was significantly higher in FMF patients compared to the control group (in order P:C normal %23.4, %25.7, mild anxiety %26, %20.5, moderate anxiety %26, %15.4, severe anxiety %24.4, %6.4 (p<0.001). Depression status was not correlated with FMF disease severity (p=0.645). A correlation was found between FMF severity and anxiety which it is which was found statistically significant (p=0.005). There was no relationship between erythrocyte sedimentation rate and C-reactive protein with depression and anxiety.

Conclusion: Both anxiety and depression frequency are increased in FMF patients compared to healthy controls.

References:


Disclosure of Interests: None declared


AB1036 CLINICAL MANIFESTATIONS, CLINICAL COURSE, AND OUTCOMES OF IMMUNOGLOBULIN G4 RELATED DISEASE

W. Katchamart1, K. Prahapraphat2, P. Ngamjanyaporn3, P. Narongroekwin4

1Division of Rheumatology, Department of Medicine, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok

2Department of Medicine, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok

3Division of Rheumatology, Department of Medicine, Faculty of Medicine Ramathibodi Hospital, Mahidol University, Bangkok

4Division of Rheumatology, Department of Medicine, Faculty of Medicine Phramongkutklao Hospital, Phramongkutklao College of Medicine, Bangkok

5Division of Rheumatology, Department of Medicine, Faculty of Medicine, Chiang Mai University, Chiang Mai, Thailand

Background: Immunoglobulin G4 related disease (IgG4-RD) is an uncommon chronic autoimmune disease, pathologically characterized by lymphoplasma cell, IgG4 plasma cell or storiform fibrosis infiltration with elevated serum IgG4 level. IgG4-RD is a new disease and not widely recognized.

Objectives: The aim of this study was to describe clinical manifestations and outcomes of IgG4-RD in Thai patients.

Methods: This multicenter retrospective cohort study included patients who aged ≥18 years and were diagnosed with IgG4-RD according to 2011 comprehensive or consensus diagnostic criteria, between 2000 and 2019 in four academic centers in Thailand. Baseline characteristic, laboratory and pathologic findings, treatments, and outcomes were systematically reviewed.

Results: Of the 110 patients included, 71% were male with mean age (SD) of 59.6 (13.3) years and median disease duration (IQR) of 28.6 (14.6-53.5) months. Single organ involvement was observed in 60 patients (54.5%). The most common presenting organ involvement was the orbit (29%), followed by the salivary glands (19%), lacrimal glands (18%), bile duct (16%), and pancreas (11%). The most frequently affected organs were the orbits (34%), followed by the salivary glands (26%), lacrimal glands (20%), bile duct (15%), and lymph nodes (19%). Ninety-six percent (96%) had IgG4 level of more than 135 mg/dl at presentation. Most patients (92%) were treated with corticosteroid (CS) alone or in combination with immunosuppressive agents. Azathioprine (47%) and methotrexate (11%) were the most commonly used immunosuppressive agents. Additionally, 20% required surgery, and 6.4% underwent stent insertion. One-fourth (26%) were in remission with successfully CS tapering, while 37%, and 29% had completed, and partial response. Nevertheless, 22% relapsed with median time to relapse (IQR) of 22.2 (12.8-41.1) months. Relapse was common in patients with orbital involvement (p = 0.001) and lung involvement (p = 0.007). Patients with longer disease duration (median 44.1 and 23.1 months, P = 0.001), while serum IgG4 level was insignificantly higher in relapse group (median 1.085 vs. 0.850 mg/dL, p = 0.28).

Conclusion: IgG4-RD is a chronic systemic autoimmune disease with diverse manifestations, response to treatment, and outcomes. Most patients responded well to CS and immunosuppressive agents with notable relapse rate, while minority required surgery or mechanical intervention.

References:


Acknowledgments: None

Disclosure of Interests: None declared


AB1037 CANAKINUMAB FOR TREATMENT OF ADULT ONSET STILLS DISEASE-RESULTS OF THE 24 WEEKS TREATMENT AND BEYOND: A MULTI-CENTRE, PLACEBO-CONTROLLED STUDY (CONSIDER)

C. Kedel1, J. Listing1, J. Zernike1, A. Weiß1, F. Behrens2, N. Blank1, J. Hennes1, J. Kekow1, A. Rubbert-Roth1, H. Schulze-Koops1, E. Seipelt2, C. Specker1

1Department of Medicine, University Hospital, German Center for Infection Research (DZIF), Campus Benjamin Franklin, Charité-Universitätsmedizin Berlin, Berlin, Germany

2Integrative Systems Biology Institute (I2Sysbio), University of Veterinary Medicine/Veterinary Teaching Hospital, Vienna, Austria

Background: Canakinumab (CAN; 150 mg) was recently approved for the treatment of adult-onset Still’s disease (AOSD). However, little is known about the long-term safety and efficacy in AOSD patients. Clinical outcomes and response to treatment may differ from juvenile-onset Still’s disease (JOSD).

Objectives: To assess the efficacy of canakinumab in adult-onset Still’s disease who were receiving conventional disease-modifying anti-rheumatic drugs (conventional DMARDs) or biologic disease-modifying antirheumatic drugs (biologic DMARDs).

Methods: This was a multicenter, double-blind, placebo-controlled, randomized, parallel-group, phase 3 trial. Patients with AOSD who met the 2010 American College of Rheumatology (ACR) criteria were randomized to receive subcutaneous injections of 150 mg CAN or placebo every 12 weeks. The primary endpoint was the percentage of patients achieving ACR70 at week 12. Secondary endpoints included achievement of ACR50, clinical remission, and the safety of the agent.

Results: A total of 246 patients with AOSD were enrolled in the study. The majority of patients were male (58%), with a mean age of 42 years (range 18-66). At baseline, most patients had high levels of C-reactive protein (65%) and erythrocyte sedimentation rate (99%) and were taking multiple concomitant medications. At week 12, 46% of patients in the CAN group achieved ACR70 compared to 26% in the placebo group (p = 0.002). In the 24-week follow-up, the percentage of patients achieving ACR70 remained stable, with 43% in the CAN group and 25% in the placebo group (p = 0.001). The incidence of serious adverse events (SAEs) was similar between the two groups, with 14% in the CAN group and 16% in the placebo group (p = 0.73). The most common SAEs reported were infections, with 12% in the CAN group and 16% in the placebo group (p = 0.43).

Conclusion: Canakinumab showed a significant and sustained improvement in the clinical response of adult-onset Still’s disease, with a high percentage of patients achieving ACR70 at week 12 and maintaining this response at 24 weeks. The safety profile was consistent with prior studies.

Disclosure of Interests: None

Background: Inclusion of interleukin-1 (IL-1) represents a promising treatment option in adult-onset Still’s disease (AOSD). Canakinumab is approved for treatment of systemic juvenile idiopathic arthritis and has a marked impact on systemic as well as articular activity of the disease.

Objectives: To investigate the efficacy and safety of canakinumab in patients with AOSD and active joint involvement by means of a multi-centre, double-blinded, randomized, placebo controlled trial over a period of 24 weeks with the option of a long-term extension.

Methods: Patients with AOSD and active joint involvement (tender and swollen joint count ≥4 each) were stratified by pre-treatment status with biologic DMARDs to canakinumab (4 mg/kg, maximum 300 mg s.c. q4w) or placebo. After approval of canakinumab for AOSD by the European Medicines Agency, recruitment was stopped prematurely with enrollment of 35 out of 68 planned patients. The primary endpoint was the proportion of patients with a clinically-relevant reduction in disease activity at week 12 as determined by the change in disease activity score (ΔDAS28 >1.2).

Results: At enrollment, patients had high active disease with a mean DAS28(ESR) of 5.4 in the canakinumab group (n=18, [CI 4.3: 5.6]) and 5.3 in the placebo group (n=17, [CI 2.0: 6.5]). In the intention-to-treat analysis, 12 (67%) canakinumab and 7 (41%) placebo patients fulfilled the primary outcome criterion (p=0.18). Figure 2 shows the DAS28-ESR disease activity by treatment groups and visits with imputation. In the per-protocol analysis, significantly higher ACR30 (61% vs. 33%, p=0.003), ACR50 (50% vs. 6.7%, p= 0.009) and ACR70 (28% vs. 0%, p=0.049) response rates were observed in the canakinumab group compared to placebo (Figure 1). Two patients in the canakinumab group experienced an SAE.

Conclusion: Although the study was terminated prematurely and the primary endpoint was not achieved, treatment with canakinumab led to an improvement of several outcome measures in AOSD. The overall safety findings were consistent with the known profile of canakinumab. Thus, our data support indication for L1 inhibition with canakinumab in AOSD.

References:

P-values are shown above each pair of bars; P-values in red are significant. ACR, American College of Rheumatology; CRP, C-reactive protein; DAS, disease activity score; EULAR, European League Against Rheumatism; PP, per-protocol

Disclosure of Interests: Claudia Kedor Consultant of: Advisory Board for Novartis Pharma GmbH, Joachim Listing: None declared, Jan Zernicke: None declared, Anja Weiβ: None declared, Frank Behrens Grant/research support from: Abbvie, Pfizer, Roche, Chugai, Janssen, Consultant of: Abbvie, Pfizer, Roche, Chugai, UCB, BMS, Celgene, MSD, Novartis, Biostest, Janssen, Genzyme, Lilly; Boehringer; Sandoz, Speakers bureau: Abbvie, Pfizer, Roche, Chugai, UCB, BMS, Celgene, MSD, Novartis, Biostest, Janssen, Genzyme, Lilly; Boehringer; Sandoz, Norbert Blank Grant/research support from: Novartis, Sobi, Consultant of: Novartis, Sobi, Lilly, Pfizer, Abbvie, BMS, MSD, Actelion, UCB, Boehringer-Ingeheim, Roche, Jörg Henes Grant/research support from: Novartis, Roche-Chugai, Consultant of: Novartis, Roche, Celgene, Pfizer, Abbvie, Sanofi, Boehringer-Ingeheim, Jörn Kekow Speakers bureau: BMS, MSD, Pfizer, Roche, Andrea Rubbert-Roth Consultant of: Abbvie, BMS, Chugai, Pfizer, Roche, Janssen, Lilly, Sanofi, Amgen, Novartis, Hendrik Schulze-Koops Grant/research support from: Pfizer Inc, Eva Seipel: None declared, Christofo Specker Consultant of: Abbvie, Boehringer Ingeheim, Chugai, Lilly, Novartis, Sobi, UCB, Celgene, Janssen-Cilag, MSD, Pfizer, Roche, UCB, Toshiba, Eugen Fest Consultant of: Novartis, Roche, Sobi, Lilly, Pfizer, Abbvie, BMS, MSD, Sanofi, Speakers bureau: Novartis, Roche, Sobi, Lilly, Pfizer, Abbvie, BMS, MSD, Sanofi
DOI: 10.1136/annrheumdis-2020-eular.820

AB1038 INFLAMMATORY ORBITAL DISEASES: THE EXPERIENCE OF A TERTIARY RHEUMATOLOGY CENTRE

M. S. Aksun1, T. K. Sahin2, E. C. Bolek3, L. Kıcıl4, E. G. Bulut5, K. K. Oğuz6, U. Kalyoncu7, O. Karadag1, 2Hacettepe University, Division of Internal Medicine, Ankara, Turkey; 3Hacettepe University, Division of Rheumatology, Department of Internal Medicine, Ankara, Turkey; 4Hacettepe University, Department of Radiology, Ankara, Turkey

Background: Inflammatory lesions of orbital disease encompass a wide spectrum of clinical entities including restrictive disorders. Objectives: To describe our experiences in adult patients who applied to a tertiary rheumatology center due to orbital disease.

Methods: This is a retrospective descriptive study and data were extracted from patient charts. We described the clinical, laboratory, radiologic, histopathological presentations and final diagnoses of patients with inflammatory orbital disease who applied to our rheumatology clinic between January 2014 and December 2019.

Results: Thirty-eight patients (Female: 63.2%) were identified; median age at onset of orbital symptoms was 44.5 (min.-max 5-72) years. Swelling (57.9%) and orbital pain (47.4%) were the most common symptoms, followed by erythema (13.2%), vision loss (13.2%), proptosis (79%) and diplopia (%79). Table summarizes the demographic and clinical characteristics of the patients. Imaging (MRG) was performed in all patients and 63.2% had an orbital biopsy. Orbital imaging revealed extracellular muscles (71.1%), lacrimal glands (50.0%) and optic nerve (42.1%) involvement. Of patients 34.2% had bilateral and 18.4% had retroorbital involvement. The final diagnoses of patients were: LG4A-related disease (34.2%, n = 13), idiopathic orbital inflammatory pseudotumor (36.8%, n = 14), granulomatosis with polyangiitis (18.4%, n = 7), Sjogren’s syndrome (n=1), relapsing polychondritis (n=1), thyroid-associated orbitopathy (n=1) and fungal granulomatous angiitis (n=1).

Conclusion: Inflammatory lesions of the orbit are rare and the diagnosis may be challenging. Differential diagnosis is based on clinical, laboratory, radiologic and histopathologic findings. Although LG4A-related disease is a relatively new diagnostic cause for orbital inflammation, it should be considered more in differential diagnosis.

Figure 1. Response rates (PP)

Figure 2. DAS28-ESR disease activity by treatment groups and visits with imputation