($p<0.0001$). No AEs occurred with CAN, only in one case (1/4, 25%) treatment was stopped for loss of efficacy.
Conclusions: To our knowledge this is the largest retrospective observational study evaluating the efficacy and safety of ANK and CAN in AOSD. A prompt response was demonstrated already at 3 months of follow-up both in ANK and CAN groups. ANK proved to be effective in both patterns of disease, appearing also safe on the infectious risk. Nonetheless, skin reaction may represent a not negligible AEs during ANK treatment.
Disclosure of Interest: None declared
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THU0542 ANTI-INTERLEUKIN 1 THERAPY IN FMF AMYLOIDOSIS: A SINGLE CENTER EXPERIENCE (CASE SERIES)

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Background: Recently there is increasing number of reports investigating the efficacy of anti-interleukin-1 (anti-IL1) therapy in AA-amyloidosis.
Objectives: Here we report our experience in IL-1 blockade in AA amyloidosis secondary to FMF.
Methods: Twenty nine FMF patients with secondary AA-amyloidosis with insufficient response to colchicine were treated with anti-IL-1 agents (canakinumab and anakinra). Creatinine (Cre), 24-hour urine protein (UP), erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were measured before and throughout the treatment to evaluate the response and side effects.
Results: Twenty nine (16 female, 13 male) patients with FMF-related amyloidosis were administered anti-IL1 agents (12 on canakinumab, 17 on anakinra) in addition to colchicine in 27 patients, with a mean dose of 1.4±0.6 mg/day. The mean age was 40.13±11.76, while the mean duration of FMF was 28.79±10.51 years. The mean follow-up was 13.92±11.31 months for anakinra and 11.82±9.92 months for canakinumab.
Initial Cre levels were less than 1.5 mg/dl in 13 patients (range 0.37-.5). In this subgroup proteinuria decreased significantly from 3739.87±4860.41 to 1321.45±2015.62 mg/day while Cre was stable (0.91±0.30 mg/dl to 1.04±0.39 mg/dl) and acute phase response was normalized (CRP from 6.16±7.86 mg/l to 5.20±9.64 mg/l, ESR from 27.3±18.63 to 14±8.04).In the second group there were 11 patients with initial Cre levels higher than 1.5mg/dl (range 1.73–3.76). Proteinuria decreased from 6321.66±5936.43 to 4827.55±6264.43 mg/day, Cre increased from 2.53±0.76 to 3.07±1.70mg/dl, while there was a decrease in the APR in this subgorup (CRP from 40±64.65 mg/l to 26.71±26.76 mg/l, ESR from 57.37±37.65 to 39.11±27.72).
There are two patients on hemodialysis whereas two underwent renal transplantation. Global patient assessment score of the whole group was decreased from 7.44±2.54 to 3.89±3.53 with anti-IL-1.
Anakinra was stopped in 11 and canakinumab in 3 patients, due to irresponsiveness in 8 and 2 patients respectively. Among patients in whom anakinra was terminated 9 were later treated with canakinumab. Twenty four patients are still recieving anti-IL-1treatment (16 on anakinra, 8 on canakinumab).
Conclusions: Anti IL-1 treatments seems to be effective and safe in the treatment of AA amyloidosis secondary to FMF. The beneficial effect of this approach is more pronounced in patients with creatinine levels of less that 1.5 mg/dl.
Disclosure of Interest: None declared
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THU0543 A RETROSPECTIVE OVERVIEW OF THE TREATMENT MODALITY, OUTCOME AND RELAPSE RISK OF IGG4 RELATED DISEASE IN HONG KONG: A DATASET OF 108 PATIENTS FROM FOUR REGIONAL HOSPITALS

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Background: Currently most consensus suggests treating IgG4 related disease (IgG4RD) with prednisolone 0.6mg/kg/day tapering over 6 months while some experts prefer continuation of steroid up to 3 years (1–3).
Objectives: In this retrospective study, the treatment modality, response and relapse risk of IgG4RD patients over the past ten years from four regional hospitals in Hong Kong were analyzed.
Methods: Four regional hospitals participated with study period from 1/1/2006 to 30/6/2016. Patients were diagnosed IgG4RD according to the Japanese Comprehensive Diagnostic Criteria for IgG4RD. Treatment response at 6 months was recorded as complete, partial or non-remission based on the patient's and physician's perspective. Relapse was defined as disease progression either clinically or radiologically.
Statistical analysis: The association between treatment response and steroid dosage was analyzed by multivariate logistic regression. Risk factors for relapse were analyzed by multivariate cox regression and the hazard ratio was reported.
Results: 37 cases had surgical excision and disease recurred in 5 cases (relapse rate =13.5%). 87 patients (81%) received steroid treatment. The mean starting prednisolone was 33.5mg daily, with a mean duration of 95.2 weeks. At 6 months, 5 patients (6%) had no response, 34 patients (41%) had partial remission and 44 patients (53%) had complete remission.
29 patients also received other immunosuppressants, including azathioprine (n=25), mycophenolate mofetil (MMF) (n=6), cyclosporine A (n=4), mercaptopurine (n=2) and rituximab (n=1). The overall response rate for azathioprine and MMF was 64% and 67% respectively.
In the final multiple logistic regression, an initial prednisolone 30mg daily or above was associated with a higher complete remission at 6 months (OR=3.4, p=0.079) and the effect was more seen in patients with salivary and orbital involvement (OR=6.8, p=0.10)
18 patients relapsed after steroid was stopped and 6 patients relapsed while on prednisolone 2.5–7.5mg daily. The one year, two year and three year relapse rate were 13.3%, 24.1% and 26.5% respectively.
In the final multivariate cox regression, the presence of maintenance steroid was associated with a lower relapse risk (Hazard ratio=0.121, p=0.000) while serum IgG4 level above twice upper limit of normal (Hazard ratio=5.283, p=0.029) and hepatobiliary involvement (Hazard ratio=2.164, p=0.095) were associated with a higher relapse risk.
Conclusions: Most patients (94%) had good response to steroid. Patients with hepatobiliary involvement and serum IgG4 level above twice upper limit of normal were at higher relapse risk and a low dose, maintenance prednisolone for longer period is recommended.
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THU0544 THE IMPORTANCE OF R202Q POLYMORPHISM IN CLINICAL EXPRESSION OF FMF: A SINGLE CENTER CROSS-SECTIONAL STUDY

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Background: Familial Mediterranean fever (FMF) is an autosomal recessive inherited disease with recurrent fever and inflammatory episodes of serous membranes. The MEFV (Mediterranean fever) gene in the short arm of chromosome 16 is affected in the FMF. This gene encodes for a protein called 'Pyrin'. The erroneously synthesized Pyrin protein due to MEFV mutations is unable to control the post inflammatory process. Although there have been many efforts to find out genotype-phenotype association in FMF, no clear relationship has been clarified.
Objectives: In this study, the relationship between FMF clinical symptoms and MEFV gene mutations and polymorphism was investigated.
Methods: Total of 158 patients with FMF was included to the study that was conducted in a tertiary rheumatology outpatient clinic. The demographic and clinical features, as well as MEFV gene mutations were recorded in a "Patient Assessment Form". The clinical status of the disease was evaluated with FMF-severity score-2 (F-SS-2). The associations between clinical features and genetic alterations were calculated with Pearson Chi-square test.
Results: The mean age of patients was 24.3±5.1, mean delay in diagnosis was 5.6±6.3 years, and 155 of the patients (%98.1) were male. The percentage of patients stating they use colchicine regularly was 136 (86.1%), and mean dose was 1.4±0.3 mg/day. The most frequent mutation was M694V (76.6%), and R202Q, M680I and E148Q were found in a descending order (60.8%, 19.0% and 13.9%, respectively). The FMF severity score was found to be higher in patients carrying M694V mutation (p=0.01). In comparison with negative family history for FMF, M694V was more prevalent in patients with positive family history (82.0% vs 67.2%, p=0.035). Besides, M694V mutation found to be associated with arthritis (p=0.045). E148Q mutation was associated with the history of orchitis (p=0.029). The most significant relation between E148Q and clinical feature was family history of hemodialysis (p=0.005). The prevalence of non-periodic myalgia was %34.2, and this symptom was not present in patients carrying V726A mutation (p=0.005). M694V/R202Q was the most prevalent compound heterozygosis and found in 16 patients (10.1%). This mutation (M694V/R202Q) was associated with fewer frequencies of myalgia and peritonitis, and with good response to colchicine.
Conclusions: The presence of R202Q polymorphism is associated with FMF, and should be considered in the routine genetic analysis of the disease. In our patients, its co-existence with M694V seems to be associated with good response to colchicine, and to alleviate the severity of the disease expression of M694V, which is known to be associated with severe course.
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