POS1381

BIOLOGIC TREATMENT OF ADULT-ONSET STILL'S DISEASE: A SINGLE CENTER EXPERIENCE

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Background: Even though corticosteroids and cDMARDs are effective for inducing remission in patients with Adult-Onset Still Disease (AOSD), relapse is common. Hence, maintaining the clinical stability is challenging. Almost all of the patients face side effects because of high dose steroid treatment. Biological DMARDs have been reported to be effective in refractory patients.

Objectives: We aimed to evaluate the patients' outcomes who were diagnosed with AOSD and treated with at least one bDMARDs in our tertiary center.

Methods: Patients with AOSD who were followed in our clinic between 2007 and 2020 were screened retrospectively. For the diagnosis of AOSD, all of the patients fulfilled Yamaguchi criteria. The demographic characteristics, baseline and post-treatment clinical findings and outcomes were reported.

Results: Twenty-eight patients (21 F, 7 M) were screened (Figure 1). The mean disease duration of the first bDMARD was 21,76 ± 28,05 months (mean ± SD). The mean duration of bDMARD treatment was 37,04 ± 30,75 months. The reasons for starting a bDMARD were systemic symptoms (%80) and chronic arthritis (%20). All of the patients used methotrexate (MTX) except one. This patient had macrophage activation syndrome during diagnosis and was treated with cyclosporine. All of the patients were treated with corticosteroids (34,28 ± 26,70mg/d) and a cDMARD at initiation (22 of them MTX, 1 of them azathioprine, 2 of them cyclosporine and 1 of them IVIg). Anakinra was the most preferred biologic as a first-line treatment modality (TNF inhibitors=5, tocilizumab=4). The main reason for switching was loss of efficacy (8/22). Tocilizumab was the most used agent in 2nd line and canakinumab was in 3rd line. Twenty-two patients were in remission at last visit. Also, 15 patients were steroid-free, 14 patients were MTX-free. Patient global visual analogue scale, acute phase reactants and daily steroid dose were reduced significantly at last visit compared to the initial visit (Table 1).

Figure 1. Presenting signs and the symptoms of the patients

Table 1. Comparison of important laboratory findings and the mean steroid dose

Clinical finding	At initiation of bDMARD mean ± SD	At the last visit mean ± SD
PG-VAS	9.8 ± 0.8	2.3 ± 2.3
ESR (mm/h)	34,28 ± 33,95	18.82 ± 11.60
CRP (mg/l)	70,76 ± 67,80	13.44 ± 27.33
Ferritin (ng/mL)	1662 ± 1239	275.7 ± 381.4
Daily steroid dose (prednisolone, mg/d)	34.28 ± 26.70	5.60 ± 8.60

bDMARD treatment was terminated in 5 patients due to complete remission (n=2) and side effects (1 of them pneumonia, 1 of them EBER (+) Hodgkin Lymphoma and 1 of them tuberculosis). Six patients experienced local injection site reaction, none of them stopped treatment. Also, one patient died while she was in remission under anakinra treatment with an unknown cause.

Conclusion: The most common presenting symptoms in our cohort were fever and salmon-colored rash. Tocilizumab is an alternative treatment modality in cases with chronic arthritis and IL-1 inhibitors are an alternative for systemic course of disease. bDMARDs, especially IL-1 inhibitors are highly effective for refractory patients with AOSD.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.3968

POS1382

RITUXIMAB FOR INDUCTION OF REMISSION IN TYPE 1 AUTOIMMUNE PANCREATITIS: A SYSTEMATIC REVIEW AND META-ANALYSIS

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Background: Type 1 autoimmune pancreatitis (AIP) is a rare form of pancreatitis that belongs to the IgG4-related disease (IgG4-RD) spectrum (1). Autoimmune pancreatitis responds brilliantly to glucocorticoids, but iatrogenic toxicity associated with prolonged steroid therapy represents a relevant clinical concern in the long-term (2). B cell depletion therapy proved to be effective in inducing IgG4-RD remission, yet most evidence are derived from small and heterogenous cohorts.

Objectives: We performed a systematic review and meta-analysis to estimate the remission rate following rituximab-based induction treatment.

Methods: Two reviewers searched MEDLINE, SCOPUS, and EMBASE until December 2020 to identify studies on remission rate and relapse rate of Type 1 AIP after induction of remission with rituximab. Studies involving three or more patients were included, including the most recent or most numerous cohorts in case of duplicates. This study was conducted in accordance with Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines. Pooled effects were calculated using a random-effects model and expressed in terms of pooled remission, relapse and adverse effects (AEs) rates.

Results: Seven cohort studies met inclusion criteria for the meta-analysis for a total of 101 patients. Reasons for rituximab administration were relapsing disease (63.5%), new diagnosis (18.5%) or glucocorticoids intolerance (17.9%). The median follow-up time was 18.5 months. The pooled rate of complete response at 6 months from induction of remission was 88.9% (95% CI 80.5%-93.9%) without heterogeneity ($I^2=0\%$) (Figure 1). The median time to relapse was 9.75 months. The pooled estimate of relapse rate was 21% (95% CI 10.5%-40.3%) with moderate heterogeneity ($I^2=51\%$), being 35.9% (95% CI 17.3%-60.1%) in the 4 studies with rate of other organs involvement (OOI)>80% and as low as 10.1% (4.8%-20.3%) in the others. The pooled estimate of rituximab-related AEs was 25% (95% CI 8.8%-53%) with substantial heterogeneity ($I^2=73.6\%$). There was no publication bias.

Conclusion: Treatment of type 1 AIP with rituximab is associated with high remission rate, a higher relapse rate in presence of OOI and limited AEs. Randomized controlled trials with adequate power are needed to confirm these findings.

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Fig.1 Pooled rate of complete response

Meta Analysis

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.3993

POS1383

THE ASSOCIATION BETWEEN BARIATRIC SURGERY AND DUPUYTREN DISEASE: A COHORT STUDY FROM SWEDISH NATIONWIDE HEALTHCARE REGISTRIES

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Background: Dupuytren disease (DD) is multifactorial, with several genetic and environmental risk factors contributing to disease susceptibility. High body mass index, however, was suggested to be protective of DD.¹ The impact of weight loss among obese patients on DD has not been assessed to date.

Objectives: To assess the association between bariatric surgery and DD in a secondary care setting.

Methods: We performed a propensity score (PS)-matched cohort study using data from Swedish nationwide healthcare registries (patient registry [secondary care], causes of death registry, prescribed drug registry). Patients aged 30-79 years who underwent bariatric surgery between 2006 and 2019 were matched to up to 2 obese bariatric surgery-free patients (called unexposed patients) based on their PS. PS-matching was carried out in risk set sampling to reduce selection bias, within 4 sequential cohort entry blocks to account for time trend biases. The outcome DD was defined as a diagnosis of DD in secondary care or partial or total fasciotomy of wrist or hand. After a 1-year run-in period, patients were followed in an "as-treated" approach. We applied Cox proportional hazard regression to calculate hazard ratios (HR) with 95% confidence intervals (CIs) of incident DD among bariatric surgery patients when compared to obese unexposed patients overall, and in subgroups of age, sex, bariatric surgery type, and by duration of follow-up.

Results: A total of 34 959 bariatric surgery patients were PS-matched to 54 769 obese unexposed patients. A total of 71.6% of bariatric surgery patients were women. Bariatric surgery patients had a mean age of 45.5 years and a mean follow-up of 6.9 years. All patient characteristics in obese unexposed patients were highly similar. We observed 126 and 136 severe DD cases among bariatric surgery and obese unexposed patients, respectively. The risk of DD was significantly increased in bariatric surgery patients compared to obese unexposed patients (HR = 1.30, 95% CI 1.02-1.65). The risk of DD was higher in women (HR = 1.36, 95% CI 1.00-1.84) than in men (HR = 1.05, 95% CI 0.70-1.58). Age did not modify the risk of DD among bariatric surgery patients compared to obese unexposed patients. Malabsorptive bariatric surgery yielded an increased risk of DD when compared to obese unexposed patients (HR = 1.33, 95% CI 1.04-1.71), while restrictive bariatric surgery yielded a null result. The risk of DD increased with duration of follow-up (>5 years of follow-up: HR = 1.63, 95% CI 1.14-2.34, null result in earlier follow-up).

Conclusion: Our results suggest that substantial weight loss is associated with a latent increased risk of severe DD in an obese population. This observation further strengthens current evidence that high body mass index is protective against DD. The latency of risk increase of DD after bariatric surgery may suggest that slowly adapting metabolic changes may be part of the mechanism of DD emergence.

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Acknowledgements: We thank Prof. Dr. Jesper Lagergren (Karolinska Institutet, Stockholm, Sweden) for hosting Dr. Theresa Burkard for a research stay at the Upper Gastrointestinal Surgery Group and making the data available for use. Furthermore, we thank Dr. Giola Santoni (Karolinska Institutet, Stockholm, Sweden) for her technical support.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.4067

POS1384

DIFFERENT CLINICAL CHARACTERISTICS OF IGG4-RRD PATIENTS AND NON-IGG4-RRD PATIENTS: A LARGE CHINESE COHORT STUDY

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Background: IgG4-related disease (IgG4-RD) is an autoimmune disorder and frequently involve multiple organs. The respiratory tract is one of the most frequently involved sites.

Objectives: This study aimed to compare the demographic and clinical characteristics of IgG4-related respiratory disease (IgG4-RRD) and non-IgG4-RRD patients in a large cohort.

Methods: We carried out a retrospective study of 452 cases of IgG4-RD (104 IgG4-RRD patients and 348 non-IgG4-RRD patients) diagnosed at Peking University People's Hospital between 2003 and 2020.

Results: IgG4-RRD patients had an elder age of disease onset and diagnosis. Multiorgan involvement and hypocomplementemia were more common in IgG4-RRD. Besides, the level of ESR, eosinophilia, IgG and IgG4 were higher in IgG4-RRD patients. In IgG4-RRD group, salivary gland, lacrimal gland, lymph nodes, biliary system and kidney were more commonly involved than those in the non-IgG4-RRD group. Also, more numbers of organ involvement and biliary

involvement were independent risk factors for the development of respiratory involvement in IgG4-RD patients.

Conclusion: Our study revealed demographic, clinical, laboratory and imaging features of IgG4-RRD patients and the underlying differences in pathogenesis between the two phenotypes, which have important implications for the diagnosis and treatment of the disease.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.4277

POS1385

AUTOIMMUNE DISEASES ASSOCIATED WITH CHRONIC LIVER DISEASES

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Background: Chronic liver diseases whatever their etiologies could be associated with immunological disturbances.

Objectives: Our aim was to evaluate the prevalence and the characteristics of autoimmune diseases associated with chronic liver diseases.

Methods: We performed a retrospective analysis of data from consecutive patients followed in our department for chronic liver diseases recruited from January 2010 to December 2019. Demographic, clinical, and paraclinical data were collected.

Results: A total of 224 patients were included. The mean age was 61.02 ± 13.2 years and the sex-ratio was 1.6. The main etiology of chronic liver diseases was viral infection C (32.1%) followed by viral infection B (22.8%) and non-alcoholic steatohepatitis (21.4%). The prevalence of autoimmune chronic liver diseases was 7.58%: autoimmune hepatitis (AIH) in four cases, primary biliary cholangitis (PBC) in huit cases, primary sclerosing cholangitis (PSC) in three cases and overlap syndrome (AIH-PBC) in two cases. Autoimmune diseases were noted in 31 cases (13.9%): autoimmune hemolytic anemia in 15 cases, autoimmune thyroiditis in 12 cases, one case of psoriasis, one case of CREST syndrome, one case of Sjögren's syndrome and one case of autoimmune thrombocytopenia. Autoimmune pathologies were more associated with autoimmune chronic liver disease than other causes of chronic liver disease (47% vs 11.1%, p < 0.001). Autoimmune pathologies were not statistically associated with female gender (p = 0.085) or young age (p = 0.483).

Conclusion: In our study, the prevalence of autoimmune diseases during chronic liver diseases was 13.9%. This prevalence was higher in the case of autoimmune chronic liver diseases (47%), which would underline the importance of systematic screening for clinical and biological immune manifestations in those patients.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.4288