Vassilopoulos et al reported three adult cases, where two were not treated with immunosuppressive drugs before anakinra and one had recurrence after anakinra was tapered. The Double Blind Placebo Controlled Clinical Trial AIRTRIP, shows efficacy of anakinra in treating 11 patients with recurrent pericarditis over 14 months. It is unclear from this study if anakinra should be tapered or not. We suggest that once steroids and immunosuppressive drugs have been discontinued, anakinra should be gradually tapered over months, to avoid relapse. These experiences warrant further long term controlled trials in order to determine the efficacy and appropriate treatment regimen of anakinra for recurrent pericarditis.
Disclosure of Interest: None declared
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THU0571 THE CLINICAL FEATURES OF 223 BEHCET'S DISEASE PATIENTS IN JAPAN

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Background: Behcet's disease is a systemic vasculitis disease with oral and genital aphthous ulcers, ocular involvements, skin manifestations, arthritis, gastrointestinal manifestations, neurogenic diseases and vascular involvements. Patients with Behcet's disease are known to distribute along the ancient Silk Road, including Japan.
Objectives: We evaluate the clinical features of Behcet's disease in Japan.
Methods: We retrospectively investigated 223 patients (108 males and 115 females) who fulfilled the International Criteria for Behcet's Disease (ICBD) from January, 2006 until May, 2015.We examined sex, onset age, disease type, clinical symptoms, laboratory data and medications.
Results:
Median age at diagnosis was 36.0±12.8 years old. Oral ulcers were the most common manifestation (98.2%), followed by genital ulcers (62.4%), ocular involvements (53.2%), erythema nodosum (53.2%), acneiform lesions (51.8%), arthritis (38.6%), gastrointestinal manifestations (25.1%), neurogenic diseases (9.0%), and vascular involvements (8.1%). The relationship of HLA and disease manifestations was studied in 123 patients (41.5% with HLA-B51 and 24.1% with HLA-A26). The frequency of acneiform lesions, ocular involvements and HLA-B51 was significantly higher in male, while genital ulcers and arthritis were significantly higher in female. Patients with ocular involvements showed a higher assosiation rate with neurogenic diseases and HLA-B51, and lower wih gastrointestinal manifestations. TNFα inhibitor (infliximab or adalimumab) were used for 66 cases (30%), and it could be continued for 1 year in 91%, and for 2 years in 83%.

Table 1

	Total (n=223)	Male (n=108)	Female (n=115)	p value
Oral ulcers	218/222 (98.2%)	105/107 (98.1%)	113/115 (98.3%)	0.94
Skin manifestations	190/220 (86.4%)	90/107 (84.1%)	100/113 (88.5%)	0.34
Erythema nodosum	116/218 (53.2%)	49/105 (46.7%)	67/113 (59.3%)	0.06
Acneiform lesions	113/218 (51.8%)	65/105 (61.9%)	48/113 (42.5%)	0.004
Thrombophlebitis	11/218 (5.0%)	5/105 (4.8%)	6/113 (5.3%)	0.85
Ocular involvements	118/222 (53.2%)	68/108 (63.0%)	50/114 (43.9%)	0.004
Genital ulcers	138/221 (62.4%)	58/107 (54.2%)	80/114 (70.2%)	0.01
Artiritis	86/223 (38.6%)	33/108 (30.6%)	53/115 (46.1%)	0.02
Gastrointestinal manifestations	56/223 (25.1%)	23/108 (21.3%)	33/115 (28.7%)	0.20
Neurogenic diseases	20/223 (9.0%)	12/108 (11.1%)	8/115 (7.0%)	0.28
Vascular involvements	18/223 (8.1%)	6/108 (5.6%)	12/115 (10.4%)	0.18
HLA-B51 positive	51/123 (41.5%)	32/62 (51.6%)	19/61 (31.1%)	0.02
HLA-A26 positive	28/116 (24.1%)	14/56 (25.0%)	14/60 (23.3%)	0.91

Table 2

	Total (n=223)	With ocular involvements (n=118)	Without ocular involvements (n=104)	p value
Gastrointestinal manifestations	56/223 (25.1%)	17/118 (14.4%)	39/104 (37.5%)	<0.001
Neurogenic diseases	20/223 (9.0%)	19/118 (16.1%)	1/104 (1.0%)	<0.001
Vascular involvements	18/223 (8.1%)	7/118 (5.9%)	11/104 (10.6%)	0.21
HLA-B51 positive	51/123 (41.5%)	36/69 (52.2%)	15/54 (27.8%)	0.006
HLA-A26 positive	28/116 (24.1%)	19/65 (29.2%)	9/51 (17.6%)	0.15

Conclusions: A higher incidence of gastrointestinal manifestations was observed in patients with Behcet's disease in Japan. Patients with ocular involvements showed a higher assosiation rate with neurogenic diseases, and lower wih gastrointestinal manifestation. Most patients could continue TNFα inhibitor safety and effectively.
Disclosure of Interest: None declared
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THU0572 ASSOCIATION BETWEEN RETROPERITONEAL FIBROSIS AND MALIGNANCY: A POSSIBLE PARANEOPLASTIC SYNDROME

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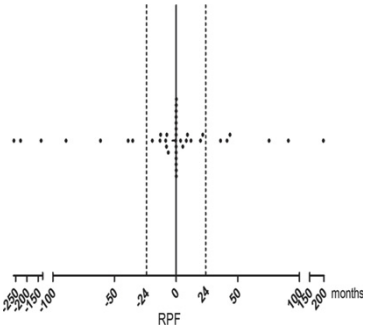
Background: Retroperitoneal fibrosis (RPF) are associated with malignancies. However it is unclear what is the incidence of malignancies and whether particular malignancies are more prevalent in RPF.
Objectives: The objective of this study was to examine standardized incidence ratios (SIRs) of cancers in patients with retroperitoneal fibrosis (RPF) compared with age- and sex-matched general population.
Methods: Medical records of 111 patients diagnosed as having RPF by computed tomography, positron emission tomography and/or histological evaluation were reviewed. Forty one cases of cancers, which were confirmed by biopsies, were identified in 35 patients with RPF. SIRs were calculated for cancers, cancer types, and age at cancer diagnosis and stratified according to RPF-cancer intervals compared with general population in Korea.
Results: The mean ± SD age at RPF diagnosis was 59.1±14.9 years, and 69.4% of the patients were male. The cancer SIR (95% confidence intervals) in patients with RPF relative to age- and sex-matched individuals in the general population was 3.18 (2.23 - 4.41) [2.65 (1.7 - 3.94) in men; 5.34 (2.76 - 9.32) in women]. The most frequent cancer was unspecified urinary organ cancers with SIR of 733.41 (238.14 – 1711.53). SIRs of multiple myeloma [27.58 (3.34 – 99.64)], renal cell cancers [9.53, (1.15 - 34.42)] and unspecified cancers [16.92, (2.05 - 61.12)] were also significantly higher than in general population. Whereas cancers were most frequently developed in the eighth decade of life, the peak SIR was observed in the fifth decade (8.41, 2.29 – 21.53). When stratified by RPF-cancer intervals, SIR was 6.85 (4.55 - 9.90) within 2 years of RPF diagnosis, while no significant increase in SIR was found out of 2 years. Malignancies (n=28) within 2 year of RPF diagnosis included unspecified urinary organ cancer (n=4), stomach cancer (n=4), lung cancer (n=4), colon cancer (n=3), renal cell cancer (n=2), pancreatic cancer (n=2), unspecified cancer (n=2), rectal cancer (n=1), gallbladder cancer (n=1), non-Hodgkin lymphoma (n=1), multiple myeloma (n=1), prostate cancer (n=1), thyroid cancer (n=1) and gastrointestinal stromal tumor (n=1). Predominant origin of these malignancies were epithelial cell types [transitional cell carcinomas (n=4), adenocarcinoma (n=16)].



Conclusions: RPF was strongly associated with cancers, particularly within 2 years of RPF diagnosis. Our results indicate that cancer screening in patients with RPF should may be performed regularly up to 2 years after RPF diagnosis.
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THU0573 CONCOMITANT AUTOIMMUNE DISEASES IN PATIENTS WITH SARCOIDOSIS

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Background: Sarcoidosis is a chronic granulomatous disease characterized by non-caseating granuloma formation. It can mimic many rheumatic diseases and/or may be coexist with them. There are limited data in the literature about the association of sarcoidosis with autoimmune diseases.
Objectives: The purpose of this study is to determine the frequency and characteristics of autoimmune diseases associated with patients with sarcoidosis.
Methods: One hundred and thirty-one sarcoidosis patients folowed-up in single rheumatology center were included in the study. Demographic, clinical, laboratory and radiological data of these cases were evaluated retrospectively. The

characteristics of autoimmune diseases associated with sarcoidosis (sarcoidosis-overlap group) patients and isolated sarcoidosis (isolated sarcoidosis group) were analyzed and compared.

Results: Autoimmune disease was detected in 15 (11.5%) of 131 patients with sarcoidosis (1Sjögren syndrome, 3rheumatoid arthritis, 1Still disease, 1scleroderma, 4ankylosing spondylitis, 1familial Mediterranean fever, 1gut arthritis, 1immune trombocytopenic purpura, 1Hashimoto thyroiditis and 1Graves disease). Most of these diseases occurred before (such as RA, AS, Still, FMF) and others after sarcoidosis diagnosis. Among 15 sarcoidosis patients with autoimmune disease 10 were female and 5 were male, the mean age was 50.8 years and mean disease duration was 3 months (1–30 months). When compared with isolated sarcoidosis patients, more hand finger joint involvement, RF positivity, higher ESR and less NSAIDs usage were found in patients with sarcoidosis-overlap group (p=0.035, p=0.049, p=0.015, p=0.018 respectively). There was no statistically significant differences between the two groups when evaluated for demographic, clinical parameters and other treatment modalities.

Conclusions: Concomitant autoimmune diseases in patients with sarcoidosis may be often seen. This patients are characterized with more hand finger joint involvement, RF positivity, higher ESR and less NSAIDs usage. Therefore, in patients with a diagnosis of sarcoidosis, it is necessary for the physician to be careful and to make a wider differential diagnosis in terms of the presence of another underlying autoimmune disease. Multicenter, prospective studies involving large numbers of patients are needed to understand whether the association of sarcoidosis-autoimmune diseases is based only on coincidence or on a common etiopathogenesis.

Disclosure of Interest: None declared

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THU0574 CLASSICAL IMMUNOSUPPRESSION AND DAMAGE PROGRESSION IN A GROUP OF PATIENTS DIAGNOSED WITH BEHCET'S DISEASE

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Background: Behcet's Disease is a rare type of vasculitis that involves both arterial and venous blood vessels of all sizes. The type of organ involvement and overall disease activity evaluated in the clinical practice determine the course of treatment and the decision to initiate immunosuppression. Activity scores such as Birmingham Vasculitis Activity score (BVASv3), Behcet's Disease Current Activity Form2006 (BDCAF), or damage indices like Vasculitis Damage Index (VDI) have been developed in this respect.

Objectives: To evaluate the ability of classical immunosuppressant therapy to prevent damage progression. To find the correlation between disease activity scores: BVASv3, BDCAF, long term treatment, immunosuppressant use and damage after remission, as calculated by VDI.

Methods: A study on a cohort of patients diagnosed with Behcet's Disease from an Internal Medicine and Rheumatology Clinic was performed. Activity and damage scores,BVASv3,BDCAF and VDI after obtained remission, were calculated. The documented cases were diagnosed according to the International Criteria for Behçet's Disease (ICBD).Windows Excel/SPSS20.0 (Spearman's correlation)were used to analyse the data.

Results: The study included 16 patients treated with long term cortisone and immunosuppressive therapy. The mean age at the time of the diagnosis was 32.3years with a male predominance 62% (10 patients). Severe systemic involvement was present in 10 cases (Ophthalmological involvement-6cases, recurrent venous thrombosis-6cases, pulmonary vasculitis-1 case, severe cardiac involvement-1case, central nervous system involvement-3cases) and all patients received classical immunosuppression (cyclophosphamide, azathioprine).The mean scores for BVASv3 and BDCAF at the time of the diagnosis were 9 and 4.12.A strong correlation was identified between BVASv3 and BDCAF (r=0.830, p<0,001). The use of immunosuppressive therapy due to severe organ involvement and long-term immunosuppression correlated stronger with BVASv3 (r=0.718) than with BDCAF (r=0.533). Vasculitis damage index (VDI) calculated after remission was obtained. There was an important correlation between disease activity scores and damage (BVASv3-VDI r=0,687,p<0,001, BDCAF-VDI r=0,676, p<0,001). Types of treatment were evaluated, a comparison was made between long-term cortisone therapy and immunosuppression.There was a stronger correlation between long term cortisone use and VDI (r=0,600) than between immunosuppression duration and damage (r=0,472).

Conclusions: Damage progression is influenced by disease activity, as calculated by activity scores (BVASv3 and BDCAF). Classical immunosuppression is used for severe organ involvement and for limiting new organ lesions once started. There was a stronger correlation between long-term cortisone use and VDI than between immunosuppression duration and damage. The damage index increased by irreversible organ damage due to disease activity and long term cortisone use, but not due to the immunosuppressive therapy.

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THU0575 EFFICACY OF RITUXIMAB IN RESISTANT PALINDROMIC RHEUMATISM:FIRST REPORT IN LITERATURE

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Background: Palindromic rheumatism (PR) although often considered as a benign disease can be severe and resistant to DMARDs in some patients. In these patients it can result in almost daily attacks, migrating from joint to joint resulting in poor quality of life. Rituximab has been proven to be effective in treatment of seropositive RA.

Objectives: To determine the efficacy and safety of Rituximab in patients with seropositive PR who had an inadequate response to CsDMARDs

Methods: PR was diagnosed based on criteria proposed by Hannonen P et al. Seropositive (ACPA±RF positivity) PR patients who had active disease despite being treated with two Cs DMARDs for >3 months, were treated with Rituximab. Active disease was defined as >4 attacks per month requiring intake of NSAIDS. All the patients were started on 500mg of rituximab after baseline work up. If complete control of palindromic attacks was not achieved and B cells were detectable in the peripheral blood by flow cytometry another 500 mg infusion was given after 2 weeks. Patients were continued on maximum tolerable dose of DMARDS. Patients were given repeat infusion of Rituximab once the patient developed clinical relapses as evidenced by recurrence of palindromic attacks.

Results: Twenty three patients with a mean age of 44.60±13.51 yrs and mean disease duration of 5.47±3.25 yrs were included. All patients were positive for ACPA while 17 patients were positive for RF. These patients were on a background of minimum of 2 DMARDs. Despite the maximum tolerable dose of DMARDs they had mean attack rate of 5.30±2.38 attacks per month. During a mean follow up of 14.17±8.62 months seven patients required two infusions and three patients required three infusions. Of the 33 infusions 500 mg was effective in controlling the attacks majority (88%) of the times. Seven patients required another 500 mg infusion after 2 weeks as initial 500 mg dose failed to achieve complete control of disease and B cell were not depleted in the peripheral blood. At one month follow up all patients achieved complete control of disease. Remission lasted for 10.33±5.75 months. When symptoms recurred patients were treated with rituximab again and all regained complete control of the symptoms. None of the patients evolved into RA during the study period. No serious adverse events were observed. Five patients experienced minor allergic reactions during infusion which were managed according to the standard protocol.

Conclusions: This case series indicates in patients of PR resistant to Cs DMARDs rituximab not only achieves disease control but also prevents progression to RA. To best of our knowledge this is the first report regarding efficacy of rituximab in PR. Although it needs to be proved in a larger blinded RCT this early data indicates that Rituximab may be a therapeutic option to prevent development of RA in seropositive patients.

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THU0576 THERAPEUTIC OPTIONS FOR PATIENTS WITH RARE RHEUMATIC DISEASES: A SYSTEMATIC REVIEW AND META-ANALYSIS

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Background: Rare rheumatic diseases are challenging for both patients and clinicians. This problem is further propelled by the scarce number of approved treatment regimens. Therapy is often limited to immunosuppression with corticosteroids or off-label use of drugs for common rheumatic diseases.

Objectives: We have recently described a set of 82 classified rare diseases in rheumatology [1]. In this systematic review, we analysed the evidence for therapeutic regimens from randomized clinical trials of rare rheumatic diseases.

Methods: For this systematic review and meta-analysis, we searched the Cochrane Central Register of Controlled Trials (CENTRAL), MEDLINE, PUBMED and SCOPUS, up to 20th of October 2016. To validate the search strategy, we selected sentinel references.

We included randomized controlled trials regarding rare rheumatic diseases and put a focus on pharmacological treatment compared with placebo, application of