diagnosis between 13and 60years, 14 (70%) under the age of 40,with a male predominance 60% (12 patients). All patients presented active disease at the time of the diagnosis. In the clinical case series, Spearman's rank correlation coefficient between BVASv3 and BDCAFwas strong r=0,862with p<0,001.The outcome analysis after remission was calculated and rank correlation coefficient betweenVDI, and both BVASv3 and BDCAF was moderate (VDI-BVASv3 r=0,747, p<0,001, VDI- BDCAF r=0,795, p<0,001).As for immunosuppression induction decision and activity scores,the correlation coefficient was moderate (r=0,734 for BVASv3,r=0,647 for BDCAF)with p<0,001. There was a moderate correlation between immunosuppressive treatment and VDI (r=0,700, p<0,001). Since the cause of damage (vasculitis vs. treatment)is not taken into consideration when we calculate VDI, we tried to observe if there are any connections between this and immunosuppression duration. There was a mild correlation and no statistical impact between cyclophosphamide treatment duration and damage calculated as VDI (r=0,474, p=0,36).In contrast, when rank correlation coefficient between cortisone therapy and VDI was calculated, a moderate statistical impact was observed (r=0,609, p<0,001).

Conclusions: Birmingham Vasculitis Activity score (BVAS) v3 and Behcet's Disease Current Activity Form2006 (BDCAF) are reliable tools for evaluating disease activity in patients with Behcet's Disease. They are able to anticipate the need for immunosuppressive therapy and the damage progression, as calculated with Vasculitis Damage Index (VDI).

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THU0556 SALIVARY GLAND ENLARGEMENT IN IGG4-RELATED DISEASE IS ASSOCIATED WITH MULTIORGAN INVOLVEMENT AND HIGHER BASAL DISEASE ACTIVITY

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Background: IgG4-related disease (IgG4-RD) is an immune-mediated condition which clinical spectrum encompasses single or multiple organ involvement. Enlargement of major and minor salivary glands is one of the main disease features. Whether salivary gland enlargement is associated with systemic involvement has not been previously evaluated.

Objectives: To elucidate if salivary gland enlargement is associated with systemic disease.

Methods: We included patients with an established diagnosis (definitive: organ involvement, biopsy proven and high IgG4 levels, probable: organ involvement, biopsy proven without high IgG4 levels, possible: organ involvement, high IgG4 levels without histology) of IgG4-RD according to the Comprehensive Diagnostic Criteria, who regularly attend a tertiary referral center in Mexico City (2000–2017). We retrospectively collected demographics, clinical (organ involvement, disease activity and damage assessed by the IgG4-RD Responder Index [IgG4-RD RI] at basal and at 6 months of follow-up, number of relapses, remission and treatment), basal laboratory (C3, C4, ESR, PCR, total eosinophil count, IgG4 levels) as well as imaging and histologic data.

Results: We included 32 patients, 17 (53.1%) men, mean age 50.2±14.1 years and median disease duration 20.5 months. Seven (21.9%) have a definitive diagnosis, 12 (37.5%) probable and 13 (40.6%) possible. Overall we identified 21 anatomic sites affected, mainly pancreas 56.2%, lymph nodes 56.2%, lacrimal glands 37.5% and bile duct 34.3%. Salivary gland involvement was present in 12 (37.5%) patients (2 parotid, 3 minor salivary gland and 7 both). Among these patients, only 5 (41.6%) referred dry mouth and in 7 patients (58.3%) glandular enlargement was the onset disease feature. Salivary glandular enlargement was identified only radiologically in 5 patients (41.6%) and both clinical and radiologically in 7 (58.3%) patients. When we compared patients with (n=12) vs. without (n=20) salivary gland enlargement, the first group had a higher number of affected organs (6.5 vs. 2, p=0.0001) and absolut eosinophil's count (348 vs. 137.5/mm^3, p=0.05), a higher prevalence of lacrimal glands (75% vs. 15%, p=0.002), lymph nodes (91.7% vs. 35%, p=0.002) and lung involvement (33.3% vs. 0%, p=0.01), azathioprine use (83.3% vs. 30%, p=0.003), as well as a higher basal IgG4-RD RI (12 vs. 6, p=0.001) and a longer delay in diagnosis (64 month vs. 6.5 months, p=0.001). We did not find differences regarding gender, age, IgG4 serum levels, C3, C4, ESR, PCR, antinuclear antibodies, rheumatoid factor, anti-Ro/SSA and anti-La/SSB antibodies (negative in all patients), number of relapses, remission at 6 months and damage. We performed a logistic regression analysis (only including the number of organs, the basal IgG4-RD RI and time of follow-up) and found an association of salivary glandular enlargement with the basal IgG4-RD RI (OR 1.63, 95% CI 1.12–2.35, p=0.009).

Conclusions: Our study highlights the systemic nature of IgG4-RD. Patients with salivary gland enlargement should be routinely screened for systemic involvement.

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THU0557 VENOUS VESSEL WALL THICKNESS IN LOWER EXTREMITY IS INCREASED IN MALE BEHCET'S DISEASE PATIENTS WITHOUT VASCULAR INVOLVEMENT

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Background: Vascular involvement is seen in up to 40% of the patients with Behcet's Disease (BD), especially in young males and is one of the major causes of mortality and morbidity. Lower extremity vein thrombosis due to vascular inflammation is the most frequent form of vascular involvement in BD. Recently, assessment of vessel wall thickness (VWT) and venous dilatation by US is suggested to be valuable in patients with vascular inflammation.

Objectives: In this study, we investigated whether vessel wall thickness or dilatation is present in young male BD patients prone to venous vascular disease.

Methods: Fifteen male patients with BD without major organ involvement followed in Marmara University Behcet's Clinics, 14 healthy male controls and 14 male patients with Ankylosing Spondylitis (AS) were included the study. Bilateral lower extremity venous doppler ultrasonography (US) was performed by an experienced radiologist blinded to cases. No patient was under immunosuppressive treatment. Bilateral common femoral vein (CFV) wall thickness and great/small saphenous vein dilatations were examined. Behçet Syndrome Activity Score (BSAS) was used for the general assessment of disease activity.

Results: The mean disease duration was 9.1±6.3 years in patients with BD. BSAS score was 28.9±19. Bilateral CFV wall thickness was significantly higher in BD patients compared to healthy controls and AS (p=0.001, p=0.002, respectively for right CFV; p=0.001, p<0.001, respectively for left CFV) (Table 1). The width of great and small saphenous veins were also higher in patients with BD, but without reaching statistical significance. There were no correlations between BSAS and wall thickness of any vessel.

Table 1. Venous wall measurements of lower extremity in study groups

	Behçet's Disease (n=15)	Healthy Controls (n=14)	Ankylosing Spondylitis (n=14)	P value
Age, years	30.2±4.5	30±5.9	30.8±4.2	0.891
Body Mass Index (kg/m^2)	23.5±3.5	23.8±2.8	26.3±3.8	0.080
Right Common femoral VWT (mm)	0.69±0.4	0.26±0.08	0.28±0.27	<0.001
Left Common femoral VWT (mm)	0.74±0.4	0.31±0.13	0.23±0.13	<0.001
Right Great saphenous width (mm)	2.94±2.6	2.1±0.71	2.5±0.73	0.436
Left Great saphenous width (mm)	3.1±2.2	2.5±0.65	2.4±1.1	0.512
Right Small saphenous width (mm)	2.4±1.8	1.4±0.3	1.7±0.5	0.126
Left Small saphenous width (mm)	2.1±1.5	1.5±0.8	1.8±0.6	0.315

VWT: Venous wall thickness.

Conclusions: In preliminary results of our study, an increased venous vessel wall thickness in lower extremity was shown in male BD patients without vascular involvement. As a similar change was not observed in controls, we think, increased VWT might be an early sign of venous inflammation in patients with BD rather than a result of non-specific systemic inflammation. Further studies with a larger group of patients is planned.

Disclosure of Interest: None declared

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THU0558 THERAPEUTIC RESPONSE TO PREDNISONE ACCORDING TO THE AGE IN POLYMYALGIA RHEUMATICA: A CONTROLLED STUDY

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Background: Polymyalgia rheumatica (PMR) is an inflammatory rheumatic disorder which usually affect patients over 65 years old. Different poor prognostic factors are involved in prednisone response including rapid decrease of prednisone dose or female sex. To date, there is no data relating the impact of the age on therapeutic response in PMR.

Objectives: The aim of this study was to compare, in case of PMR, the response to prednisone in patients younger than 60 to patients over 65 years old.

Methods: This was a retrospective, monocentric study. We included patients suffering from PMR, meeting ACR 2012 criteria. Patients were classified into two groups, one group with patients less than 60 years, and one group with patients over 65 years. We registered demographic, clinical, biological, and imaging data as well as therapeutic response profile. The local inflammation was evaluated with PET scan, by studying each anatomical site usually affected by PMR. Then, the rate of inflammation was scored from 0 to 3, according to the intensity of uptake compared to liver. The treatment was standardized. The initial dose of prednisone was of 0.3mg/kg/j during the two first weeks, then, the dose was slowly decreased

by 10% each month. The main endpoint was a steroid dependence defined by the recurrence of PMR symptoms and/or the increase of CRP at two times during the decrease of prednisone.
Results: We included 14 patients younger than 60 years old (average age 54+/- 0.8 years) and 28 patients older than 65 years old (average age 75.8 +/- 1.5 years). The population younger than 60 years was mainly male (60% VS 27%, $p<0.05$). Both groups were similar in terms of morning stiffness (2,1±0,4 VS 1,9±0,3 hours; $p>0.05$), disease duration (4,2±0,8 VS 4,1±0,6 months; $p>0.05$), leukocytes rate (8,3±1,37 VS 8±0,7 G/L; $p>0.05$) and percentage of antinuclear antibodies rate over 1/320 (20% VS 10%; $p>0.05$). However, regarding to local inflammation, the intensity of FDG uptake highlighted by the Pet scan was lower among young patients (score of 16,9±1,7 VS 26,5±3,0; $p<0.05$). Furthermore, we observed a significant difference concerning therapeutic response according to the age: 60% of the young patients developed a steroid dependence compared to20% in group of old patients ($p<0.05$). Moreover, the introduction of methotrexate was necessary for 35% of the young patients against 6.5% ($p<0.05$).
Conclusions: Our study is the first to highlight the age as a bad prognosis factor in case of PMR. This difference is independent to the systemic inflammation and surprisingly, local inflammation (assessed by the TEP score) is more important in elderly people. Young patients suffering from PMR are mostly men and are more dependent on steroids. Thus, methotrexate could be straightaway proposed, particularly in patient younger than 60 years old.
Disclosure of Interest: None declared
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THU0559 PERSISTENT PRURITIC SKIN LESIONS WITH DYSKERATOTIC CELLS IN UPPER LAYER OF EPIDERMIS ARE SPECIFIC AND ASSOCIATED WITH HIGH LEVELS OF SERUM IL-18 IN ADULT-ONSET STILL'S DISEASE

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Background: Adult-onset Still's disease (AOSD) is an acute and systemic inflammatory disorder that is characterized by high spiking fever, evanescent rash, arthralgia/arthritis and hyperferritinemia. However, recent reports showed that not only typical evanescent salmon-colored rash but also atypical skin lesions, persistent pruritic papules and plaques, could be associated with AOSD.
Objectives: The aim of this study is to assess the clinical significance of dyskeratotic cells in skin lesions of Japanese patients with AOSD.
Methods: We retrospectively assessed clinical and histological findings of skin lesions including persistent pruritic skin lesions in Japanese patients with AOSD (n=12). Moreover, we compared serological and histological finding of AOSD with that of dermatomyositis (DM) (n=6), drug eruptions (DE) (n=6), and Graft versus Host disease (GVHD) (n=6).
Results: AOSD with persistent pruritic skin lesions (n=7) histologically showed dyskeratotic cells only in upper layer of epidermis and horny layer without intraepidermal infiltrations of inflammatory cells. These dyskeratotic cells were positive by TUNEL and single stranded DNA (ssDNA) stainings, suggesting apoptotic cells. AOSD with evanescent rash (n=5) histologically showed no dyskeratosis. On the other side, the pathological findings of DM (n=6), DE (n=6) and GVHD (n=6) had dyskeratotic cells in all layers of epidermis with inflammatory cells infiltrations. Notably, AOSD with dyskeratosis (n=7) had significant higher levels of serum IL-18 (74,300∼307,000 pg/ml) than AOSD without dyskeratosis (n=5).
Conclusions: AOSD with persistent pruritic skin lesions is characterized and specific by prominent epidermal apoptosis, especially involving the upper layers. Therefore, it could play a pivotal role to recognize the atypical skin lesions of AOSD for correct early diagnosis. Finally, the high levels of serum IL-18 might be related with epidermal apoptosis of keratinocyte in AOSD.
Disclosure of Interest: None declared
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THU0560 DEVELOPMENT OF SYMPTOMS IN VERY EARLY PHASE IN PATIENT WITH ADULT ONSET STILL DISEASE

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Background: Adult onset Still Disease (AOSD) is a systemic inflammatory disease presenting various non-specific symptoms like fever, arthralgia and pharyngalgia. Those symptoms are mimicking common cold, which could lead to the delayed diagnosis. However, little is known about the profiles and development of symptoms in very early phase before diagnosis.
Objectives: To clarify the clinical course of AOSD symptom development in very early phase and its effect on the diagnosis delay.
Methods: Consecutive patients with AOSD with enough information in our hospital were enrolled. Initial symptoms before and at diagnosis were investigated in detail. The gradual course from the initial symptoms to the fulfilment of Yamaguchi criteria for AOSD was examined.
Results: A total of 51 patients were enrolled. The mean age at diagnosis was 45.0±18.9 years old and 41 (80%) were female. All patients were met with the Yamaguchi criteria. The mean duration from the first symptom to diagnosis was 50.4 days. The duration from the first symptom to the first visit to the medical facility including a general physician was 19.1 days, and that from the first medical facility visit to the first blood test was 8.0 days. While the first symptom was arthralgia in 29 (34.1%), fever in 20 (23.5%), and eruption in 20 (23.5%), pharyngalgia in 13 (15.3%), at diagnosis, fever was found in all patients, eruption in 47 (92.2%), arthralgia in 45 (88.2%), pharyngalgia in 34 (66.7%), lymphadenopathy and/or splenomegaly in 35 (68.6%), increased white blood cell (WBC) count in 44 (86.3%), ferritin elevation in 42 (82.4%), liver enzyme elevation in 41 (80.4%), negative rheumatoid factor (RF) and anti-nucleolar antibody (ANA) in 30 (58.8%). Arthralgia developed 41.9 days prior to the diagnosis, fever 40.9 days, eruption 29.5 days, pharyngalgia 24.5 days and lymphadenopathy and/or splenomegaly 14.8 days. WBC increase was detected 17.8 days prior to the diagnosis, negative RF and ANA 15.1 days, liver enzyme elevation 14.6 days, and ferritin elevation 10.9 days. At 14 days from the first symptom, 32 (62.7%) met 3 of Yamaguchi criteria, 23 (45.1%) met 4, and 19 (37.3%) met 5. At 28 days, 40 (78.4%) met 3 of Yamaguchi criteria, 32 (62.7%) met 4, and 27 (52.9%) met 5. The duration to diagnosis was significantly shorter in the patients with eruption developing within 7 days since the first symptoms than those without (35.2 vs 74.3 days, p=0.03)
Conclusions: The mean duration from the first symptom to the diagnosis of AOSD was approximately 50 days, and only a half of the patients met Yamaguchi criteria at 28 days after first symptom. The time of eruption emergence affected the early diagnosis.
Disclosure of Interest: None declared
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THU0561 CLINICAL PRACTICE GUIDELINE FOR DIAGNOSIS AND MANAGEMENT OF CATASTROPHIC ANTIPHOSPHOLIPID SYNDROME

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Background: Catastrophic antiphospholipid syndrome (CAPS) is characterized by the rapid onset of widespread or multifocal large and/or small vessel thrombosis associated with multi-organ failure in patients meeting the serological criteria for antiphospholipid syndrome [1]. Mortality in CAPS approaches 50% [2].
Objectives: The RARE-BestPractices project group identified CAPS as a rare disease condition of interest in which to develop a clinical practice guideline. The project was run in partnership with McMaster University, and used the GIN-McMaster Guideline Development checklist and Grading of Recommendations Assessment, Development and Evaluation (GRADE) methodology to develop guidelines on rare diseases [3].
Methods: The CAPS guideline was coordinated by a steering committee including representatives from RARE-BP and methodologists from McMaster University. The CAPS guideline panel consisted of 19 international members, including patient representation. The panel used the GradePro software to brainstorm and prioritize potential questions and outcomes. Systematic reviews were performed for each question. To supplement the published evidence, we compiled raw data for mortality from the CAPS Registry, and systematically elicited expert opinion from the panel members using a systematic observation form. For each question an evidence profile and evidence to decision table was generated and shared.
Results: The question prioritization step generated 47 questions, which were ranked to identify the top priorities. The top 10 questions were chosen for guideline development, yielding 7 therapy and 3 diagnostic questions. The outcome generation step yielded 7 outcomes.
The questions were addressed during an in-person panel meeting, held on April 27, 2016 in Barcelona, Spain, with follow-up via webinar on June 3, 2016, and with web-based voting completed July 31, 2016.
Recommendations were developed for all questions and will be discussed in detail.
Conclusions: Ten recommendations were issued by the CAPS Guideline Panel to assist clinicians in diagnosis and management of suspected CAPS patients. Future research is needed to improve evidence quality in rare diseases such as CAPS. The GIN-McMaster Guideline Development Checklist and the GRADE methodology were effective in producing a rigorous guideline in this rare disease.
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