

diagnosis between 13 and 60 years, 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 BDCAF was strong $r=0.862$ with $p<0.001$. The outcome analysis after remission was calculated and rank correlation coefficient between VDI, 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 corticosteroid 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 Behçet's Disease Current Activity Form 2006 (BDCAF) are reliable tools for evaluating disease activity in patients with Behçet'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 absolute eosinophil count (348 vs. $137.5/\text{mm}^3$, $p=0.05$), 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 BEHÇET'S DISEASE PATIENTS WITHOUT VASCULAR INVOLVEMENT

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Background: Vascular involvement is seen in up to 40% of the patients with Behçet'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 Behçet's Clinics, 14 healthy male controls and 14 male patients with Ankylosing Spondylitis (AS) were included in 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.

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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 $0.3\text{mg}/\text{kg}/\text{j}$ during the two first weeks, then, the dose was slowly decreased