SAT0236

CLINICAL AND LABORATORY FEATURES OF PATIENTS WITH SJOGREN'S SYNDROME COMPLICATED WITH MALT LYMPHOMA

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Background: The risk of developing lymphomas in Sjogren's syndrome (SS) is more than 10-fold higher than in general population. Available publications describe a number of indicators considered as predictors for SS development.

Objectives: To describe clinical and laboratory characteristics of patients with SS and MALT lymphoma and compare them with idem parameters in the control group. To identify factors associated with development of lymphoma.

Methods: The study included 87 SS patients with MALT lymphoma. In all cases lymphoma involved the parotid salivary glands. In all cases MALT lymphoma was diagnosed simultaneously with SS. At the time of inclusion none of the patients was on immunosuppressive therapy. Fifty five SS patients without lymphoproliferative pathology composed the control group. All cases were newly diagnosed and treatment-naïve.

SS was diagnosed based on the ACR-EULAR criteria. The histologic and immunohistochemical diagnosis of lymphoma was performed with B-cell clonality determination in salivary gland tissue.

The following clinical and laboratory parameters were monitored in both groups: rates of stage 3 xerostomia (<0.5 ml/5 min), grade 3 hypolacrimation (<5 mm/5 min), lymphadenopathy, hemorrhagic rashes, decreased C3, C4 complement components, increased RF, high anti-Ro and anti-La antibodies positivity, hematological changes, serum levels of secretory monoclonal antibodies, cryoglobulinemia, etc. Pearson's χ^2 criterion (analysis of contingency tables) was used for statistical analysis. The differences were considered statistically significant when p values were <0.05. Statistica 10 for Windows (StatSoft Inc., USA) package was used for statistical data processing.

Results: In the study group 84 patients were females, their mean age at the onset of lymphoma was 53±10 years. The disease duration was 7 years (3-12) before the diagnosis was established. All patients in the control group were females, the mean age at diagnosis was 50.2 ±13 years. Patients' age at diagnosis did not differ significantly between the groups. Enlarged salivary or lacrimal glands were found in all SS-MALT patients and in 18% of patients in the control group.

The rates of such systemic manifestations as polyneuropathy, kidney and joint damage were low and did not differ between groups.

Increased levels of anti-Ro antibodies was documented in the majority of patients in both groups (87 % and 81%, p=0.4), while aLa antibodies were significantly more common in MALT lymphoma patients (60% and 40.3%, p=0.045).

Similar rates of increased IgG and IgM levels were found in both groups, while increased IgA levels were 6-fold more common in the lymphoma group (p<0.00001). Anemia and leukopenia were documented in approximately 25%, and thrombocytopenia - in 2% of patients in both groups. Cryoglobulinemia (36% vs. 24%) and circulation of secretory monoclonal immunoglobulins (32% vs. 18%) were more common in the lymphoma group, but the difference was insignificant (p=0.2). The incidence of other clinical and laboratory abnormalities in SS and SS-MALT patients is presented on the graph Forest plot with OR and CI indication.

Graphic 1. Clinical and laboratory features of SS patients with MALT-lymphoma compared with SS patients without lymphoma.

Conclusion: Therefore, universally recognized predictors of lymphoma development, such as cryoglobulinemia and hypocomplementemia did not show reliable association with lymphoma. In analyzed cohort development of MALT lymphoma was statistically significantly associated with recurrent parotitis in past medical history, presence of lymphadenopathy at diagnostic examination, increased levels of anti-La antibodies and IgA, and hypergammaglobulinemia. Probably we should more actively treat patients with these clinical and laboratory features in order to prevent the development of lymphoma in them.

Disclosure of Interests: None declared

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SAT0237

THE SYSTEMIC LUPUS INTERNATIONAL COLLABORATING CLINICS (SLICC) FRAILTY INDEX (SLICC-FI) PREDICTS DAMAGE ACCRUAL IN SYSTEMIC LUPUS ERYTHEMATOSUS (SLE) PATIENTS. DATA FROM A LATIN AMERICAN MESTIZO COHORT

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Background: The Systemic Lupus International Collaborating Clinics (SLICC) Frailty Index (SLICC-FI) has been developed as a predictor of outcomes in SLE patients¹⁻³. However, it needs to be validated in several populations.

Objectives: To evaluate the SLICC-FI as a predictor of future damage accrual in systemic lupus erythematosus (SLE) patients.

Methods: Patients from a single-center prevalent cohort were included. Damage accrual was defined as the increase in SLICC/American College of Rheumatology (ACR) damage index (SDI) scores between the baseline and last visits. The SLICC-FI was measured at baseline. Univariable and multivariable negative binomial regression were performed to determine the association between the baseline SLICC-FI (per 0.05 increase) and damage accrual during follow-up, adjusted for sex, age at diagnosis, socioeconomic status, disease duration, SLE Disease Activity Index 2000 (SLEDAI-2K), SDI, prednisone daily dose, antimalarial and immunosuppressive drug use at baseline, and duration of follow-up.

Results: Of the 265 patients included, 248 (93.6%) were female with mean (SD) age 35.1 (13.6) years at diagnosis. At baseline, mean (SD) SLE disease duration was 7.3 (6.5) years, SDI was 1.1 (1.3) and SLEDAI-2K was 5.3 (4.6). The mean (SD) baseline SLICC-FI was 0.22 (0.05). After a mean (SD) of 5.2 (2.2) years of follow-up, the SDI increased in 126 (47.5%) patients, and the final mean (SD) SDI score was 1.7 (1.7). Higher SLICC-FI scores at baseline predicted greater damage accrual in the univariable analysis [Incidence Rate Ratio (IRR)=1.283, (CI95% 1.072-1.536); p=0.007]. The SLICC-FI remained associated with damage accrual in the multivariable model, after adjustment for possible confounders [IRR= 1.224 (CI95% 1.007-1.488); p=0.042].

Conclusion: The SLICC-FI predicts damage accrual in prevalent SLE, supporting the relevance of this index in the evaluation of SLE patients. This is the first study validating the SLICC-FI in South American population

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SAT0238 VALIDATION OF THE ADJUSTED GLOBAL ANTIPHOSPHOLIPID SYNDROME SCORE AND CORRELATION WITH EXTRA-CRITERIA MANIFESTATIONS

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Background: Adjusted global antiphospholipid syndrome score (aGAPSS) is the simplified version GAPSS that was recently developed to assess thrombotic risk by the consideration of antiphospholipid antibody (aPL) profile and conventional cardiovascular risk factors.

Objectives: The aim of this study was to evaluate the validity of the aGAPSS in predicting thrombosis and extra-criteria manifestations in our antiphospholipid syndrome (APS) cohort.

Methods: Ninety-eight patients with APS were classified according to clinical manifestations as vascular thrombosis (VT), pregnancy morbidity (PM) or both (VT+PM). The aGAPSS was calculated as defined before. Arterial hypertension and hyperlipidemia definitions were made according to the ESC/ESH ve NCEP/ATP III guidelines, respectively.

Results: Demographic, laboratory and clinical characteristics of patients are summarized in table-1. Mean aGAPSS was calculated as 10.2 ± 3.8. Significantly higher aGAPSS values were seen in VT (n=58) and VT+PM (n=29) compared to PM (n=11) (mean aGAPSS 10.6 ± 3.7 vs 7.3 ± 2.9, P=0.005; 10.5 ± 4 vs 7.3

Table-1. Demographic, laboratory and clinical characteristics of patients.

	PAPS (n=42) n (%)	SLE/APS (n=56) n (%)	P
Female	36 (85.7)	47 (83.9)	0.52
Age, years (mean±SD)	44.6 (11.6)	40.8 (10.1)	0.42
Disease duration, years (mean±SD)	10 (8.8)	9.7 (7.1)	0.16
Thrombosis	35 (83.3)	52 (92.9)	0.12
•Arterial	24 (68.6)	34 (65.4)	0.47
•Venous	19 (54.3)	26 (50)	0.43
•Recurrent	15 (42.9)	22 (42.3)	0.56
Pregnancy morbidity	20 (47.6)	20 (35.7)	0.16
•<10 weeks, ≥ 3 abortions	5 (25)	4 (20)	0.5
•≥ 10 weeks, ≥ 1 abortion	14 (70)	15 (75)	0.5
•Pre-eclampsia/eclampsia	3 (15)	5 (25)	0.34
•<34 weeks, ≥ 1 premature birth	1 (5)	5 (25)	0.09
Conventional risk factors			
•Arterial hypertension	17 (40.5)	35 (62.5)	0.02
•Hyperlipidemia	21 (50)	26 (46.4)	0.41
•Diabetes mellitus	3 (7.1)	3 (5.4)	0.51
•Obesity	19 (45.2)	16 (28.6)	0.07
•Smoking	12 (28.6)	18 (32.1)	0.43
aPL profile			
•LA	29 (69)	48 (85.7)	0.04
•aCL IgG/IgM	31 (73.8)	28 (52.8)	0.03
•aβ2GPI IgG/IgM	22 (56.4)	27 (52.9)	0.45
•Triple positive	14 (33.3)	17 (30.4)	0.46

Figure 1. ROC curve according to cut-off aGAPSS value: 10 (AUC: 0.71, sensitivity: 0.52, specificity: 0.91, positive predictive value: 0.98, negative predictive value: 0.19, p-value: 0.01).

± 2.9, P=0.01, respectively). AUC demonstrated that aGAPSS values ≥ 10 had the best diagnostic accuracy for thrombosis (figure-1). Higher aGAPSS values were also associated with recurrent thrombosis (mean aGAPSS 11.5 ± 3.7 vs 9.9 ± 3.6, P=0.04). Regarding extra-criteria manifestations, patients with livedo reticularis (n=11) and APS nephropathy (n=9) had significantly higher aGAPSS values (mean aGAPSS 12.9 ± 3.4 vs 9.9 ± 3.7, P=0.02; 12.4 ± 2.9 vs 10 ± 3.8, P=0.04, respectively).

Conclusion: Our results suggest that patients with higher aGAPSS values are at higher risk for developing vascular thrombosis (either single or recurrent) and extra-criteria manifestations, especially livedo reticularis and APS nephropathy.

Disclosure of Interests: None declared

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SAT0239 LONG TERM OUTCOMES IN PATIENTS WITH ANTIPHOSPHOLIPID SYNDROME AND RENAL ARTERY STENOSIS

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Background: Antiphospholipid syndrome (APS) is characterized by thrombosis and obstetric morbidity in the context of positive antiphospholipid antibody markers. More than a quarter of hypertensive APS patients (26%) have renal artery stenosis (RAS) (1). Treatment includes anticoagulation, blood pressure control and management of cardiovascular risks. In some cases, the severity of the renal vascular lesion requires surgical intervention.

Objectives: To evaluate long term outcomes in APS patients with RAS.

Methods: Retrospective database study. All records of APS patients with RAS were analyzed and demographic variables, comorbidities, treatment received, renal outcome and mortality were recorded.

Uni- and multivariate analyses were performed by logistic regression with death and chronic kidney disease (CKD) as dependent variables. In the multivariate analysis, the covariates considered were age at diagnosis of stenosis, diabetes, smoking, dyslipidemia, unilateral or bilateral stenosis, stenosis greater than 50%, surgery and the use of immunosuppressants, anticoagulation and statins.

Research and development office has approved this study.

Results: 33 RAS patients were analyzed. Diagnosis of RAS was made by MRI renal angiography and in some cases by CT angiogram and intra-arterial digital subtraction angiography. Patient characteristics are detailed in Table 1. The median duration of follow-up was 152 months (IQR 65).

Table 1. Patient Characteristics.

N° Patients	33
Age median	48 (IQR 16)
Smokers%	18.1(6)
Comorbidities %	
Diabetes mellitus type 2	6 (2)
Hypertension	100(33)
High LDL	36.3(12)
Primary APS %	39.4 (13)
Secondary APS %	60.6 (20)
Unilateral RAS%	75.8 (25)
Bilateral RAS%	24.2 (8)
Degree of stenosis	
< 30%	15.1 (5)
30-50%	30.3 (10)
50-80%	33.3 (11)
>80%	21.2 (7)
Treatment%	
Medical treatment	75.7 (25)
Surgical treatment	30.3 (10)

Renal biopsy was performed in 3 patients: crescentic glomerulonephritis was found in 2 patients and one had a thrombotic microangiopathy.

Treatment: 25 patients (75.7%) were anticoagulated with vitamin K antagonists, 19 (57.6%) received immunosuppressive therapies, 18 (54.5%) were on statins. Ten patients (30.3%) were managed surgically with balloon angioplasty. Restenosis occurred in 4/10 patients (40%) and percutaneous renal artery stenting was performed successfully in all four. Ten patients died (30.3%). Renal outcomes are shown in Table 2.

Table 2. Outcome in APS with RAS Patients

N %	
Died	10 30.3
Transplanted	1 3
Renal Dysfunction	17 51.5
CKD Stage III	10 30.3
CKD Stage IV	4 12.1
CKD Stage V	3 9