Results: Two hundred forty-two SLE patients were evaluated; 94.4% of them were female. Mean values were as follow: age at diagnosis 33.29±13.53 years, disease duration 15.82±10.56 years, SLEDAI 5.91±5.06, SLICC score 1.06±1.42, BLyS levels 1.811±1.757 ng/mL. The 22.5% of patients displayed increased BLyS levels. The 29.6% of total patients exhibit SLEDAI values up to 6, and only the 7% of them showed SLEDAI values up to 6 and high BLyS levels simultaneously. Higher BLyS levels were significantly correlated to the ANAs positivity (p=0.0006) and lymphopenia (p=0.01) but showed no correlation with hypocomplementemia neither anti-dsDNA. The statistical analysis did not yield differences in the clinical activity or accumulated damage between patients with lower and higher BLyS levels.
Conclusions: In our series we observed a 22.5% of patients with high levels of BLyS, and the 7% of cases had BLyS high levels and SLEDAI>6. BLyS upregulation is related to ANAs positivity and lymphopenia. We have found no statistical evidences on the relationship of BLyS levels and clinical activity in our series of patients.
Disclosure of Interest: None declared
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AB0494 INCREASED LEVELS OF INTERFERON ALPHA AND INTERLEUKIN-10 AS CLINICAL ACTIVITY BIOMARKERS IN SYSTEMIC LUPUS ERYTHEMATOUS PATIENTS

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Background: Systemic lupus erythematous (SLE) is an autoimmune disease characterized by immune system disruption, including deregulation of cytokine production. Interferon alpha (INF1A) is considered a key molecule in SLE etiopathogenesis, being responsible of the differentiation of dendritic cells from monocytes, and indirectly of interleukin 10 (IL10) upregulation. The B lymphocyte stimulating factor (BLyS) is involved in autoantibodies production and clinical activity, and is regulated by other cytokines as IL10 and INF1A.
Objectives: To analyze the association among INF1A, IL10 and BLyS levels and clinical activity in SLE.
Methods: A cross-sectional, observational study of 142 patients diagnosed of SLE according to SLICC 2012 criteria and 34 healthy controls was performed. A complete blood-test was made, and clinical data by personal interview was collected. We analyzed serum concentration of IL10, BLyS and INF1A by colorimetric methods. Patients were dichotomized as high and low levels for each cytokine based on the cytokine level above 2 SD of the mean in healthy controls. Biostatistical analysis with R (3.3.2.) was performed.
Results: In our SLE patients we observed higher values of IL10, BLyS and INF1A than controls ($P<0.001$, P=0.005 and P=0.043 respectively), showing an average values in patients of 13.39±27.73 pg/mL INF1A, 9.99±15.84 pg/mL IL10 and 1811.31±1757.81 pg/mL BLyS. The mean clinical activity measured by SLEDAI was 5.91±5.06.
Statistical analysis indicate that INF1A levels are correlated to IL10 levels (P=0.001) and BLyS levels (P=0,034). Due to this finding, we categorized SLE patients by low or high level of the three cytokines: 44 INF1A(-)IL10(-)BLyS(-); 61 INF1A(+)IL10(-)BLyS(-); 5 INF1A(+)IL10(-)BLyS(+); 18 INF1A(+)IL10(+)BLyS(-) and 14 INF1A(+)IL10(+)BLyS(+). There is a high association of increased IL10-INF1A levels and the increased of clinical activity by SLEDAI score ($P<0.001$), and to a lesser extent with increased IL10-INF1A-BLyS levels. Patients with high IL10-INF1A and IL10-INF1A-BLyS showed a significant rise in C3-C4 consumption ($P<0.001$ and P=0.001 respectively) and high anti-dsDNA (P=0.001 and P=0.002 respectively). Patients with increased INF1A-BLyS exhibited high anti-dsDNA (P=0.004) and ENA positivity ($P<0.001$). In addition, patients with increased levels of IL10-INF1A-BLyS showed ANAs ($P<0.001$) and antiphospholipid autoantibody positivity (P=0.004).
Conclusions: The 69% of our SLE patients displayed almost one cytokine increased, being the INF1A the cytokine that mainly is increased. However, increased IL10 levels, irrespective of whether there is also increased levels of BLyS and/or INF1A, is the cytokine which best fits to clinical activity in SLE.
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AB0495 ASSESSMENT OF FRACTURE RISK IN A COHORT OF EGYPTIAN FEMALE SYSTEMIC LUPUS ERYTHEMATOSUS PATIENTS

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Background: Survival of systemic lupus erythematosus (SLE) patients has improved dramatically due to improved treatment, and the morbidity pattern has shifted towards long-term complications as osteoporosis. SLE occurs in women during child-bearing years and the disease often persists to the postmenopausal period[1]. Assessment of fracture risk in SLE patients is important as fractures may occur while bone mineral density (BMD) is above the osteoporotic threshold or at the normal range[2]. Osteocalcin measurement helps to assess fracture risk and select patients for treatment.
Objectives: To assess the fracture risk in a cohort of Egyptian female SLE patients by using BMD and osteocalcin level with correlation to disease activity, damage index and drugs in use.
Methods: 70 females with SLE ≥40 years old satisfying the SLICC classification criteria were enrolled with detailed history taking including disease duration, drugs in use, traditional risk factors, regular exercise, history of previous fractures and menstrual history. Assessment of disease activity using Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) and disease damage using the Systemic Lupus International Collaborative Clinics/ American College of Rheumatology Damage Index (SLICC/ACR DI). Serum calcium, phosphorus and alkaline phosphatise were measured. BMD measured by dual energy X-ray absorpiometry (DEXA) scans at lumbar spine (LS) (L2-L4) and femoral neck (FN), serum osteocalcin level and World Health Organization fracture risk assessment tool (FRAX®).
Results: 14//70 (20%) patients had LS osteoporosis, 25//70 (35.7%) had LS osteopenia and 6/70 (8.6%) had FN osteoporosis, 30/70 (42.9%) had FN osteopenia. FRAX-Major ≥20% was observed in 10% of patients, FRAX-HIP ≥3% was seen in 27.1% of patients. Serum osteocalcin level was significantly decreased in SLE patients with lower BMD than those with normal BMD, and significantly decreased in patients with osteoporosis than those with osteopenia. A significant negative correlation was found between osteocalcin level and age of patients, disease duration, SLEDAI and SLICC scores, current, IV pulse and cumulative steroids, immunosuppressants, anticoagulants, but there was a positive correlation with antimalarials and calcium supplements.
Conclusions: SLE patients are at greater risk for developing osteoporosis and osteopenia. Ten-year risk of major and hip fractures was high in SLE patients. Increasing age, disease duration, high anti-DNA titres, SLEDAI and SLICC were associated with a higher 10-year probability of major osteoporotic fracture. FRAX predicted incident hip and major osteoporotic fractures among SLE patients with normal and low bone mass not just those with frank osteoporosis. Physicians should be alerted to the higher risk of future fractures in SLE patients for periodic monitoring.
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AB0496 VENOUS THROMBOSIS IS MORE PREVALENT IN PATIENTS WITH ANTIPHOSPHOLIPID SYNDROME (APS) ACCOMPANYING SYSTEMIC LUPUS ERYTHEMATOSUS, WHILE LIVEDO RETICULARIS IN PRIMARY APS

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Background:
APS more often overlaps with other systemic autoimmune diseases like systemic lupus erythematosus (SLE) or rheumatoid arthritis (RA) than occur as a distinct disease. Our purpose was to evaluate what are the differences between patients with primary APS and APS accompanying SLE.
Objectives: The objective of this study was to compare patients with primary APS and APS/SLE group whther we can find find any clinical or laboratory parameters that can distinguish them from each other.
Methods: 112 patients with APS were included to the study, 57 of them with primary APS and 55 with coexisting SLE. These patients were followed at the Department of Connective Tissue Diseases, NIGRiR, Warsaw, Poland. At inclusion a full medical history and physical examination data were recorded.
Results: Both groups were similar in age, gender and duration of disease. Among all the clinical manifestations of APS, venous thrombosis was more frequent in patients with concomitant SLE. Skin involvement was significantly more prevalent in primary APS and it was caused mainly by livedo reticularis presence. From neurological manifestations, the occurrence of epilepsy was comparable, when EEG changes were more frequent in APS/SLE group. From laboratory measures, leucopenia, low complement concentrations and proteinuria were more prevalent in APS/SLE group. No differences were observed in thrombocytopenia and elongation of APTT as well as antiphospholipid antibody profile.
Conclusions: In conclusion, arterial thrombosis is more characteristic for primary APS. We found that livedo reticularis is very characteristic feature for primary APS. Although epilepsy occurrence is comparable in APS and APS/SLE, higher frequency of EEG changes in APS/SLE group suggests that the mechanisms

Table 1. Main differences in both groups

	Total N=112	APS N=57 (50,9%)	TRU+APS N=55 (49,1%)	P
Age	48,3±13,7	46,9±14,3	49,7±13,2	0,2187
Gender	96 (85,7%)	45 (78,9%)	51 (92,7%)	0,0698
APS duration (months)	72 [24–144]	48 [13–120]	80 [31–156]	0,2329
Venous thrombosis	61 (54,5%)	25 (43,9%)	36 (65,4%)	**0,0218**
Arterial thrombosis	63 (56,2%)	30 (52,6%)	33 (60,0%)	0,4320
Thrombosis all	97 (86,6%)	47 (82,5%)	50 (90,9%)	0,1891
livedo reticularis	53 (47,3%)	41 (71,9%)	12 (21,8%)	**<0,0001**
EEG changes	18 (16,1%)	5 (8,8%)	13 (23,6%)	0,0323
Epilepsy	20 (17,9%)	8 (14,0%)	12 (21,8%)	0,2823

leading to this feature can be distinct between this two conditions. Additionally, it appears to be more reasonable to add thrombocytopenia to clinical criteria of APS, because its frequency does not depend on SLE presence.

Disclosure of Interest: None declared

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AB0497 SIX CASES OF MACROPHAGE ACTIVATION SYNDROME AS PRESENTING MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: Macrophage Activation Syndrome (MAS) is a life-threatening syndrome characterized by excessive immune activation. It can be triggered by conditions affecting immune homeostasis, such as infections, malignancies and rheumatologic disorders, including Systemic Lupus Erythematosus (SLE). In previous studies, prevalence of MAS among SLE patients ranged from 0.9% to 4.6%.

Objectives: To describe the presentation and treatment of both MAS and SLE in patients with both syndromes.

Methods: Monocentric retrospective evaluation: patients with MAS according to HLH classification criteria were identified in our cohort of SLE patients (classified according to ACR and SLICC criteria)followed for at least 1 year between 1972 and 2014.

Results: Among 511 patients with SLE (mean age at diagnosis:31 years ±2),6 patients (1.2%)with MAS were identified (all female). Their main clinical and laboratory features are reported in table 1.Median HLH score was 226.5 (IQR 204–254),with a probability of having MAS of 96%.In all cases MAS happened simultaneously to the onset of SLE.Median age at diagnosis was 31.5 years, median SLEDAI was 12.All patients had fever above 38°C,lymphadenopathy,hematological involvement,and high titer ANA positivity. Workup for infections and malignancies was negative in all cases.All patients were treated with corticosteroids (100% received intravenous immunoglobulin pulse of methylprednisolone);concomitant medications were: cyclosporin A in 83%, IVIG in 67%, granulocyte colony-stimulating factor in 17%, mycophenolate mofetil in 17%, etoposide in 17% and plasma exchange in 17%. Two patients required haemotransfusion.All cases required hospital admission, and 2 were admitted in intensive care unit. No death from MAS was observed (median follow up: 34.5 months;IQR25–48).One patient died 44 months after after MAS for pulmonary adenocarcinoma.Table 1:main clinical and laboratory features at diagnosis of SLE and MAS

Table 1: main clinical and laboratory features at diagnosis of SLE and MAS

Clinical features of MAS n (%)		SLE ACR classification criteria	
Fever	6 (100%)	§Malar rash/Oral ulcers/Photosensitivity/ LED	5 (83%)
Hemorrhages	1 (17%)		
CNS dysfunction	0 (0%)		
Lymphadenopathy	6 (100%)		
Hepatomegaly	4 (67%)	Arthritis	2 (33%)
Splenomegaly	4 (67%)	Nephritis	0
		Serositis	3 (50%)
		CNS disease	1 (17%)
		Haematological involvement	6 (100%)
MAS Laboratory Parameters median (IQR)		Autoantibodies n (%)	
WBC (x10³/uL)	2.1 (1.8-2.3)	ANA	6 (100%)
Neutrophils (x10³/uL)	1.5 (0.87-1.8)	anti-ds DNA	5 (83%)
HGB (g/dL)	7.6 (7-8,2)	anti-RNP	2 (33%)
PLT (x10³/uL)	134 (72-142)	anti-Ro	1 (17%)
AST (U/L)	250 (166-402)	anti-Sm	1 (17%)
ALT (U/L)	111 (92-135)	antiphospholipid Ab/LA	3 (50%)
LDH (U/L)	769 (535-915)		
Ferritin (µg /L)	4607 (1897-31533)	*Direct Coombs Test +	5 (100%)
Fibrinogen (mg/dL)	97 (35-170)	C3 mg/dl (n.v 80-160)	34 (28-70)
Triglycerides (mg/dl)	511 (317-605)	C4 mg/dl (n.v 10-40)	13 (7-17)
ESR (mm)	17.5 (9-24)		
CRP (mg/dl)	37 (10-49)		

* this test was available only for 5 patients

§at least one of these criteria: malar rash; Oral ulcers; Photosensitivity; LED: Discoid Lupus Erythematosus

CNS: central nervous system; WBC white blood cells; HGB: hemoglobin; PLT: plateletAST: aspartate aminotrasferase; ALT: alanine aminotrasferase; LDH: lactate dehydrogenase; ESR: erytrocyte sedimentation rate; CRP: C-reactive protein; ANA antinuclear antibodies; anti-ds DNA: anti double stranded DNA antibodies; anti-RNP: anti-ribonucleoprotein antibodies; LA: Lupus anticoagulant: n.v.: normal values

Conclusions: MAS is a rare complication in our SLE cohort and can complicate the onset of SLE, but it seems to be a very uncommon manifestation during the course of the disease.Fever may be a red flag for possible MAS, particularly if temperature is persistently above 38° in absence of signs and symptoms of underlying infection.In our series, all cases were treated successfully with immunosuppressive drugs and cytotoxic agents such as etoposide were used only in one case.

Disclosure of Interest: None declared

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AB0498 SOLUBLE CD14 (PRESEPSIN) AS A POTENTIAL BIOMARKER TO DISCRIMINATE INFECTION VS. ACTIVITY IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: Differentiation of Systemic Lupus Erythematosus (SLE) activity and infection in a febrile SLE patient become difficult, since initial clinical presentation may be similar. Several biological markers (including procalcitonin and CRP) have been evaluated, with discordant results. Soluble CD14 (sCD14), also called presepsin, is the receptor for lipopolysaccharide - lipopolysaccharide binding protein (LPS-LBP) complexes. CD14 could activate a series of signal transduction pathways and inflammatory cascades, and lead to systemic inflammatory responses.(1)

Objectives: The aim of this study is to evaluate the utility of sCD14 as a biomarker to differentiate infection *vs.* activity in SLE patients admitted with systemic inflammatory response (SIRS).

Methods: We included 11 patients with SLE (ACR criteria 1997) and SIRS (International conference 2001) admitted to the ER and/or ICU. The measurement of sCD14 in plasma by enzymatic immunoassay of chemiluminescence *in vitro* was performed to differentiate active SLE *vs.* infection. Infection was considered if a positive culture/PCR was obtained. Mann-Whitney test was used to evaluate the association of variables with infection.

Results: All patients were female; mean age 37.9 years. An infectious disease was confirmed in 5 cases (3 bacterial including urinary tract infection, pneumonia and bacteremia; 1 viral infection by Chikungunya virus and 1 fungal by *histoplasma capsulatum*). sCD14 was elevated in the infected SLE patients (median: 1005 pg/mL- RIC: 533–1415-) vs patients with lupus flare (median: 431.5 pg/mL - RIC: 369–579-) (p=0,04).

Conclusions: High values of sCD14 levels seem to be useful to differentiate infections from activity in SLE patients with SIRS. More patients and further analysis are necessary to define the clinical use of this biomarker.

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AB0499 TOTAL BODY WATER AND ITS CORRELATION WITH SICCA SYMPTOMS IN PRIMARY SJÖGREN'S SYNDROME

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Background: Patients with primary Sjögrens' syndrome (PSS) suffer from severe alterations in both the quality and quantity of saliva and tears. Body water represents around 50–55% of the body weight. Tears contain 98% of water and saliva 99.5%.

Objectives: To evaluate the percentage of total body water (TBW) among patients with PSS and to assess its correlation with sicca symptoms.

Methods: We included 85 patients with PSS and 85 historical non diabetic controls matched by gender, age (±3 years) and body mass index (±1kg/m^2) (BMI). We assessed the presence of sicca symptoms, Schirmer-I test, non-stimulated whole salivary flow (NSWSF) and ocular staining. We also evaluated the ocular and oral domains of the ESSPRI, a validated scale for symptoms (a higher score implies worst symptoms). We obtained the TBW percentage with a bioelectric impedance analysis (BIA-SECA-514, Hamburgo).

Results: 80% were women, mean age 54.8±13.7 years and mean disease duration 11.5±7.52 years. The percentage of TBW was similar among patients and controls (PSS 46.85±4.6 vs. 46.9±4.5, p=0.88). Among the patients, the