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 these 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

DOI: 10.1136/annrheumdis-2017-eular.1736

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

F. Dall'ara¹, I. Cavazzana², M. Frassi², M. Taraborelli², M. Fredi², F. Franceschini², L. Andreoli¹, A. Tincani¹, P. Airò². ¹Rheumatology and Clinical Immunology, University of Brescia - Dscs; ²Rheumatology and Clinical Immunology, Spedali Civili di Brescia, Brescia, Italy

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-

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 (x10 ³ /uL)	2.1 (1.8-2.3)	ANA	6 (100%)
Neutrophils (x10 ³ /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 (x10 ³ /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	3 (50%)
LDH (U/L)	769 (535-915)	Ab/LA	
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: platelet; AST: aspartate aminotransferase; ALT: alanine aminotransferase; LDH: lactate dehydrogenase; ESR: erythrocyte 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

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; IQR 25–48). One patient died 44 months after MAS for pulmonary adenocarcinoma. Table 1: main clinical and laboratory features at diagnosis of SLE and MAS

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

DOI: 10.1136/annrheumdis-2017-eular.6692

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

I. Posso-Osorio¹, A. Echeverry¹, D. Aguirre-Valencia¹, G. Castaño², G. Tobón¹. ¹Rheumatology; ²Laboratory of immunology, Fundación Valle del Lili, CALI, Colombia

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.

References:

[1] Zou Q, Wen W, Zhang X. Presepsin as a novel sepsis biomarker. World J Emerg Med [Internet]. Second Affiliated Hospital of Zhejiang University School of Medicine; 2014 Aug 22;5(1):16–9. Available from: <http://www.ncbi.nlm.nih.gov/pmc/articles/PMC4129857>.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.2495

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

G. Hernandez-Molina¹, P. Almeda-Valdés², G. López-Carrasco², M. Astudillo-Angel¹, M. González-Acevedo², V. Zamora-Legoff¹, P. Sánchez-Fernández², C. Aguilar-Salinas², I. Cruz Bautista². ¹Immunology and Rheumatology; ²Endocrinology Department, Instituto Nacional de Ciencias Médicas y Nutrición Salvador Zubiran, Mexico City, Mexico

Background: Patients with primary Sjögren's 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 (±1 kg/m²) (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, Hamburg).

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