had been successfully treated, 27.8% had metastasis, 5.6% had died. There was no significant difference in SLEDAI between patients with cancer and patients without. Whereas malignancy is correlated to longer disease duration ($p = 0.01$) and older age of SLE onset with significant difference ($p = 0.001$).

Conclusion: Although we have detected an increasing incidence of cancer in SLE patients in comparison to normal population, our study didn't find a significant correlation between SLE disease activity and the risk of cancer. We should closely observe SLE patients with old age at onset and/or long disease duration because of their higher risk for cancer development.

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AB0395 SYSTEMIC LUPUS ASSOCIATED MACROPHAGE ACTIVATION SYNDROME – A SINGLE CENTRE EXPERIENCE FROM INDIA

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Background: Macrophage activation syndrome (MAS) is a potentially fatal complication of systemic lupus erythematosus (SLE). (1) Overlapping clinical features of SLE activity with MA makes it a clinical diagnosis. There is limited data from developing countries like India.

Objectives: To study the clinical features, treatment, and outcomes in SLE-MAS.

Methods: This single-centre, retrospective study included patients fulfilling SLICC 2011 criteria for SLE, admitted from January 2017 to November 2019, and diagnosed as MAS by a team of attending physicians. Their demographic and clinical data, treatment, and outcomes were recorded. H score and SLEDAI were calculated. Patients were assessed for HLH classification criteria (2004) and preliminary diagnostic criteria for MAS in childhood SLE (cSLE).

Table 1. Patient characteristics

Characteristic	N=16
Age (years)	26 (19.5 – 30)
Duration of symptoms (days)	60 (18.7 – 90)
Fever	16 (100)
Pancreatitis	1 (6.3)
Renal	7 (43.8)
Neurological	7 (43.8)
Myocarditis	6 (37.5)
Hepatomegaly	7 (43.8)
Splenomegaly	3 (18.8)
Lymphadenopathy	12 (75)
Anaemia	16 (100)
Leukopenia	13 (82)
Thrombocytopenia	14 (88.2)
Coagulopathy	7 (43.8)
Concomitant infection	3 (18.8)
H score	222 (193 – 254)
cSLE-MAS diagnostic criteria	16 (100%)
HLH 2004 (≥5)	7 (43.8)
Corticosteroids	16 (100%)
Cyclophosphamide	8 (50%)
Intravenous immunoglobulin	2 (12.5%)
Cyclosporine	6 (37.5%)
Death	4 (25%)

All data presented as n (%) and median (IQR)

cSLE-MAS – Childhood systemic lupus erythematosus – Macrophage activation syndrome, HLH – Hemophagocytic lymphohistiocytosis

Results: Sixteen patients (median age – 26 years, 15 females) were included. Twelve patients (75%) had MAS as the initial presentation of SLE. The common clinical features were fever (100%) and cytopenias (100%). The mean duration of symptoms was 60 days. The most frequent biochemical abnormalities were high ferritin (>500 ng/ml, 100%) and elevated transaminases (100%, aspartate transaminase > alanine transaminase). Common complications were renal (43.8%), neurological (43.8%), and coagulopathy (43.8%). Seven and 16 patients fulfilled the HLH 2004 and cSLE – MAS preliminary criteria, respectively. The median H score was 222, giving a cumulative probability of 96%. All the patients received high-dose steroids. Cyclophosphamide pulse and cyclosporine were administered to 8 (50%) and 6 (37.5%) patients respectively. There were four (25%) in-hospital mortalities.

Table 2. Laboratory features

Parameter	N = 16
Haemoglobin (g/dL)	6.9 (5.7 – 7.8)
Total leucocyte count (cells/mm ³)	1400 (1025–3175)
<1000	3 (18.8)
1000 – 2500	9 (56.3)
2500 – 4000	1 (6.3)
Platelet (cells/mm ³)	57500 (23500 – 95250)
<20000	3 (18.8)
20000 – 50000	3 (18.8)
50000 – 1 lac	8 (50)
Serum ferritin > 500 ng/ml	16 (100%)
Fibrinogen	2.48 (1.6 – 4)
Triglycerides (mg/dL)	375 (294–470)
AST/ALT (U/L)	153 (113 – 234) / 90 (68 – 170)
Procalcitonin (pg/mL)	0.8 (0.3 – 1.4)
Lactate dehydrogenase (U/L)	1674 (1081 – 2184)
Erythrocyte sedimentation rate (mm in 1 st hour)	61 (44 – 69)
C-reactive protein (mg/L)	36 (6.3 – 52)
Bone marrow examination	14 (87.5)
Increased histiocytes with hemophagocytosis	5 (32.5)

All data presented as n (%) and median (IQR)

ALT – Alanine transaminase, AST – Aspartate transaminase

Conclusion: Fever, cytopenia, high ferritin, and elevation of transaminases were the commonest features in this series of SLE-MAS. SLE-MAS carried a high mortality (25%) despite aggressive treatment.

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AB0396 DIAGNOSTIC PROTOCOL INCLUDING MINIMALLY INVASIVE MINOR SALIVARY GLANDS BIOPSY IN SJÖGREN'S SYNDROME IN A SPANISH REFERENCE CENTER

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Background: Sjögren's syndrome (SjS) is a systemic autoimmune disease with a broad clinical presentation from dry syndrome to systemic extraglandular manifestations. The diagnosis can be complex since none of the markers, except anti-Ro, is sufficiently sensitive or specific. Although, minor salivary glands biopsy (MSG), Schirmer's test and unstimulated whole salivary flows (UWSF) are the hallmark for the diagnosis of this entity, its use is not widespread in some centers.

Objectives: The aim of the study was to analyze the usefulness and safety of the diagnostic protocol for the classification of SjS and the immunological and analytical markers in dry syndrome due to SjS.

Methods: Prospective observational study of a cohort of patients with sicca syndrome from a reference center. The diagnostic protocol (Schirmer's test, UWSF and minimally invasive MSGB) was applied in the same consultation. Demographic, clinical, analytical and histological data were reviewed.

Results: Over a period of 6 months, 48 patients with dry syndrome were analyzed, of which 39 women (81.2%). The main suspicion was SjS (39), followed by sarcoidosis (3), IgG4-related disease (2) and other diagnoses (4). The mean age was 59.1 ± 4.4 years. Almost half (45.8%) reported xerostomia and 41.6% xerophthalmia. Recurrent parotidomegaly was described in 6 patients (12.5%) and arthralgias in 12 (25%). Immunologically, 23 (47.9%) presented anti-nuclear antibodies, 13 (27.1%) anti-Ro, 4 (8.3%) anti-La, 12 (25%) rheumatoid factor and 15 (31.2%) low C4. Schirmer test was positive in 32 patients (66.7%), UWSF in 22 (45.8%) and 9 (18.8%) had a focus score ≥ 1 , although 16 (33.3%) had focal lymphocytic sialadenitis in the MSGB. A total of 21 (43.8%) patients were classified according to the 2016 ACR/EULAR criteria. 12 (57.1%) were seropositive SjS and 9 (42.9%) seronegative SjS. MSGB sensitivity was 71% and specificity 96%. Patient reported symptoms were unhelpful to differentiate SjS from other causes of dry syndrome. The number of protocols needed to diagnose a SjS was 2.28 (5.33 in seronegative SjS). Complications associated with the procedure were low (1 of 48) and mild (self-limited paraesthesia). Patients with SjS, unlike those with dry syndrome of another etiology, had more anemia ($p < 0.001$), lymphopenia ($p = 0.022$), ESR ($p = 0.030$), beta-2 microglobulin ($p = 0.011$), ANA ($p < 0.001$), anti-ENA ($p = 0.006$), anti-Ro ($p < 0.001$), low C4 ($p < 0.001$) and hypergammaglobulinemia ($p = 0.002$).

Conclusion: Immunological and histological manifestations were more predictive than clinical ones to differentiate SjS from other causes of dry syndrome. MSGB is a simple, sensitive, specific and safe procedure. The application of the diagnostic protocol (Schirmer test, UWSF and MSGB) allowed to standardize the classification of SjS and increased the diagnosis of patients with seronegative SjS.

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AB0397

LUPUS FATIGUE PROBLEMS IN THE RUSSIAN COHORT OF PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS (RENAISSANCE).

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Background:

Objectives: To determine the dependency of fatigue from SLE activity, irreversible organ damage and HRQoL in SLE patients of the Renaissance cohort.

Methods: 328 Russian SLE patients were enrolled in the study (M/F 30/298) who fulfilled SLICC 2012 criteria. The SLEDAI 2K activity, SLICC damage index, Facit Fatigue scale, and health related quality of life (HRQoL) using the LupusQoL questionnaire were evaluated.

Results: Based on the Facit Fatigue scale scores fatigue was verified in 148 (45%) out of 328 patients with SLE. Following lupus fatigue status patients were divided into two groups - 148 and 168 patients respectively. The groups were perfectly matched in terms of age, duration of the disease, duration of GCs therapy, and damage scores (Table 1)

The SLEDAI 2K activity scores were significantly higher in the group with lupus fatigue - 9.6 ± 6.0 vs 6.7 ± 4.2 ($p = 0.01$) as compared to values in the group without

fatigue, as well as the level of antibodies to DNA ($p = 0.02$), 110.2 ± 34.2 and 82.3 ± 20.5 , respectively.

There was a significant decrease in HRQoL in patients with lupus fatigue based on scores in all eight scales of the questionnaire ($p = 0.00$),

Table 1. Comparative analysis of 328 SLE patients with and without lupus fatigue

Characteristics mean (SD)	Patients with lupus fatigue (n=148)	Patients without lupus fatigue (n=168)	P (Mann-Whitney)
Age in years	35,31 $\pm$ 12,03	33,68 $\pm$ 10,68	0,13
Disease duration in years	9,57 $\pm$ 9,47	10,1 $\pm$ 9,17	0,6
Duration of glucocorticoid therapy in month	72,81 $\pm$ 71,58	79,43 $\pm$ 75,98	0,6
SLICC Dal, score	1,55 $\pm$ 1,37	1,69 $\pm$ 1,23	0,44
C3 Mu/ml	0,88 $\pm$ 0,33	0,87 $\pm$ 0,31	0,75
C4 Mu/ml	0,15 $\pm$ 0,12	0,16 $\pm$ 0,14	0,36
ANF, hep2	641 $\pm$ 453	550 $\pm$ 402	0,4
Fatigue scores by the Facit fatigue scale	25,16 $\pm$ 6,58	42,43 $\pm$ 4,56	0,000

Figure 1. Comparative assessment of HRQoL using the LupusQoL questionnaire in 328 SLE patients with and without fatigue.

Conclusion: Almost half (45%) SLE patients in the Russian cohort experience fatigue. It is associated with disease activity on the SLEDAI 2k scale and high levels of antibodies to DNA. All patients experiencing lupus fatigue have significantly worse HRQoL based on scores in all eight scales of the questionnaire LupusQoL.

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AB0398

TNF-LIKE WEAK INDUCER OF APOPTOSIS / FGF INDUCIBLE MOLECULE 14 PATHWAY IN UNTREATED LUPUS NEPHRITIS: SERUM OR URINE TWEAK LEVELS MORE ACCURATE IN RELATION TO RENAL ACTIVITY?

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Background: Lupus nephritis (LN) is one of the most serious manifestations of SLE, affecting 70% of patients and the most critical predictor of morbidity and mortality of the disease [1]. Tumor necrosis factor-like weak inducer of apoptosis/ fibroblast growth factor inducible molecule 14 (TWEAK/Fn14) activation is involved in various pathological processes that occur locally in kidneys, facilitating the pathogenesis of LN [2].

Objectives: To assess serum and urine TWEAK levels as well as renal Fn14 expression in newly diagnosed patients with LN and its correlation to disease activity.

Methods: The present study included 30 selected newly diagnosed previously untreated SLE patients divided into 2 groups; 15 patients with LN and 15 without LN as well as 30 age and sex matched healthy subjects who served as control group. Written consent was obtained from all patients and