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AB0392

THE EFFECT OF HYDROXYCHLOROQUINE ON THE RISK FACTORS FOR ATHEROSCLEROSIS DEVELOPMENT

R. Wakiya¹, K. Ueda¹, H. Shimada¹, S. Nakashima¹, M. Mahmoud Fahmy Mansour¹, M. Kato¹, T. Miyagi¹, K. Sugihara¹, Y. Ushio¹, T. Kameda¹, H. Dobashi¹. ¹Kagawa University, Division of Hematology, Rheumatology and Respiratory Medicine, Department of Internal Medicine, Kagawa, Japan

Background: Systemic lupus erythematosus(SLE) patients, especially patients with lupus nephritis have poor vascular endothelial function and increased cardiovascular(CV) mortality.

Meanwhile, several studies showed hydroxychloroquine(HCQ) has effect on reduction in lipids and thrombosis(1), but the mechanism is unclear.

Objectives: We examined effect of HCQ on adipocytokine expression in SLE patients.

Methods: 52 SLE patients with low disease activity started with HCQ were analyzed before and 3 months after HCQ treatment. 21 SLE patients has past history of lupus nephritis. Serum S100 proteins and adipocytokines were measured by ELISA, and serum inflammatory cytokine levels were evaluated by Multiplex assay (TNF- α , IL-6, VEGF-A).

Results: Serum adiponectin level was increased significantly 3 months after HCQ treatment compared with those at baseline (mean change 1.35, Figure 1). SLE patients who achieved LLDAS had a greater increase than those who did not. Additionally, the changes of serum adiponectin levels were associated with those of TNF- α , IL-6, VEGF-A and S100A9 protein, which plays an important role of SLE pathogenesis.

Figure 1. Serum adiponectin levels at baseline were compared with levels after 3 months of HCQ treatment. Serum adiponectin levels significantly decreasing during HCQ treatment in SLE patients. For statistical analyses * $p < 0.0001$, P value: Wilcoxon signed-rank test

Conclusion: A HCQ could reduce the risk factors for atherosclerosis along with control of SLE disease activity.

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AB0393

EFFICACY OF HYDROXYCHLOROQUINE FOR LUPUS NEPHRITIS IN MAINTENANCE PHASE

M. Watanabe¹, Y. Haji¹, M. Kato¹, T. Ito¹, Y. Banno¹, M. Okada². ¹Daido Hospital, Nagoya, Japan; ²St.Luke's International Hospital, Tokyo, Japan

Background: Hydroxychloroquine (HCQ) is an essential drug for systemic lupus erythematosus. But in Japan, chloroquine and HCQ remain unavailable until mid

2015 because of a series of lawsuits about its retinal toxicity in the 1970s. There is insufficient knowledge regarding renal protective effect of HCQ.

Objectives: We aimed to investigate its efficacy of adding HCQ for Lupus nephritis (LN) as a maintenance-phase therapy.

Methods: We conducted an observational retrospective cohort study included patients with LN ($n=42$) in maintenance-phase in Japan. We reviewed medical records of LN patients aged > 18 years who were initiated HCQ from May 2015 to May 2018. Maintenance phase was defined as stabilization in serum creatinine and urinary segment after induction therapy and who achieved complete or partial remission. The annual change of proteinuria was compared between patients on HCQ who have proteinuria ($>0.5\text{g/gCr}$) or not. Other outcome measures were disease flare, dose of steroids, renal and immunologic features.

Results: A total of 42 patients were analyzed and allocated to two groups based on their amount of daily urine protein level: HCQ with proteinuria as group A ($>0.5\text{g/gCr}$, $n=14$) and HCQ without proteinuria as group B ($\leq 0.5\text{g/gCr}$, $n=28$). Both groups were comparable, with mean (SD) age of 36.1 (12.9) years and 37.5 (13.8), female 78.6% and 92.9% in each group, mean (SD) disease duration until HCQ of 3.5 (3.25) and 3.3 (2.9) years in group A and group B, with prednisolone dose at base line of 10.3 (7.1) mg and 7.9 (4.4) mg, respectively. The mean (SD) proteinuria at base line was 1.38 (1.11) g/gCr in group A and 0.20 (0.09) g/gCr in group B and after 12 months, proteinuria decreased in group A (-1.34g/gCr in group A vs $+0.03\text{g/gCr}$ in group B; $p < .001$; 95% CI, 0.305-0.736). No relapse was experienced in group A during the study period.

Conclusion: In patients with clinically stable LN but with proteinuria, hydroxychloroquine is a good therapeutic option for achievement of complete remission.

Disclosure of Interests: None declared

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17. SLE, Sjögren's and APS - clinical aspects (other than treatment)

AB0394

DOES HIGH DISEASE ACTIVITY IN SYSTEMIC LUPUS ERYTHEMATOSUS PATIENTS INCREASE THE RISK OF CANCER INCIDENCE?

A. Abdalla¹, F. I. Abdelrahman². ¹Aswan University, Rheumatology and Rehabilitation, Aswan, Egypt; ²Zagazig University, Rheumatology and Rehabilitation, Zagazig, Egypt

Background: Cancer is known as one of the causes of morbidity and mortality in systemic lupus erythematosus (SLE) patients. It has been thought that SLE activity and stimulation of the immune system predisposes the risk of cancer (1).

Objectives: To investigate the correlation between SLE disease activity and the cancer incidence.

Methods: The study included a cohort of SLE patients, diagnosed according to the American College of Rheumatology classification criteria⁽²⁾ attending the Rheumatology department, Aswan University in the period from January 2018 to June 2019. We used a questionnaire to screen patients who were diagnosed with cancer. We collected demographic and laboratory data on all screened patients and their disease activity using the Systemic Lupus Erythematosus Disease Activity Index (SLEDAI)⁽³⁾. For the patients diagnosed with cancer, we recorded age of onset of SLE, age of diagnosis of cancer, type of cancer, treatment received, immunosuppressive regimen (dosage and duration) and cancer outcomes.

Results: The study included 117 patients (95 female, 22 male), mean age (25.6 ± 6.5) years with mean SLE duration (7.3 ± 6.3) years and mean SLEDAI (9 ± 8.9). 76% had lupus nephritis, 62.2% had hematological abnormalities and 17.8% had neuropsychiatric lupus. 91% were on corticosteroids (CCS), 33% on mycophenolate mofetil (MMF), 43% on azathioprine, 14% on cyclosporin. 56.8% were either receiving or had received intravenous cyclophosphamide (CYC) with a mean cumulative dose (7.5 ± 4.7) gm. We found 18 (15.3%) patients (13 female and 5 males) were diagnosed with cancer during the course of SLE with mean age at onset (31 ± 3.7), mean age at cancer diagnosis (39.28 ± 10.77), mean SLE duration (18.17 ± 6.02) and mean SLEDAI (7.39 ± 4.19). Most of SLE patients with cancer had lupus nephritis (89%) and all cancer patients were on a median dose of CCS 10 (2.5- 20) mg daily for median 10 (4-24) years. 83.5% of them had received intravenous CYC prior to the development of cancer with mean total cumulative dose of (6.7 ± 4.6) gm, 67% received MMF, 33% received cyclosporine and 50% received azathioprine. Types of cancer were as follow; 22.2% lymphoma, 16.7% cancer cervix, 16.7% cancer breast, 11.1% colorectal cancer, 11.1% squamous cell carcinoma, 5.6% leukemia, 5.6% bronchogenic carcinoma, 5.6% prostate cancer and 5.6% cancer thyroid. 66.7% of them

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

N. Acharya¹, S. Jha¹, D. Mishra¹, A. Chattopadhyay¹, S. Jain¹, S. K. Sharma¹, A. Sharma¹, S. Jain¹, V. Dhir¹. ¹Post Graduate Institute of Medical Education and Research, Division of Rheumatology, Department of Internal Medicine, Chandigarh, India

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/mm3)	1400 (1025–3175)
<1000	3 (18.8)
1000 – 2500	9 (56.3)
2500 – 4000	1 (6.3)
Platelet (cells/mm3)	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

J. Álvarez Troncoso¹, C. M. Oñoro López¹, C. Soto Abánades¹, E. Ruiz Bravo², L. Ramos Ruperto¹, L. Blasco Santana², R. Sorriguieta Torre¹, Á. Robles Marhuenda¹, E. Martínez Robles¹, A. Noblejas Mozo¹, J. J. Ríos¹, F. Arnalich Fernández¹. ¹Hospital Universitario La Paz, Department of Internal Medicine, Madrid, Spain; ²Hospital Universitario La Paz, Pathological Anatomy, Madrid, Spain

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.