Methods: Twenty – two cases of systemic lupus erythematosus complicated with hemophagocytic syndrome were retrospectively analysed. The clinical manifestations, laboratory tests and prognosis were collected.

Results: Twenty-two patients with systemic lupus erythematosus complicated with hemophagocytic syndrome developed fever in 22 patients, including 18 with hyperpyrexia, 20 with hyperferritin, 20 with cytopenia, 10 with splenomegaly, 6 with hyperglycemia, 5 cases of hypofibrinogenemia, 14 cases of bone hemoptysis occurred in the bone wear, nk cell activity decreased or absent in 7 cases, 15 cases of lymphadenopathy, mostly bilateral armpit lymph nodes and neck bilateral lymph nodes, liver function Abnormalities/hepatic insufficiency in 11 cases, ultrasound showed 10 cases of cardiac involvement, MRI showed 5 cases of cumulative brain. 22 patients were diagnosed with high-dose glucocorticoid, 9 cases of hormonal impact, 17 cases of human immunoglobulin immunosuppression in 9 cases, 4 cases of ganciclovir, 1 cases of acyclovir, 1 case of relying on Park Glycosides. 21 cases improved, 1 case to persuade invalid to abandon treatment and discharge.

Conclusions: SLE and hemophagocytic syndrome have some similarities. The clinical diagnosis is easy to miss. When patients with SLE continue to have high fever, splenomegaly, blood cells decrease, phagocytosis occurs in bone wears, coagulation dysfunction should be combined with vigilance HLH, Improve the relevant inspection. High-dose glucocorticoid combined with immunosuppressive agents, human immunoglobulin therapy can effectively improve the prognosis.

REFERENCE:

- [1] Henter JL, Horne A, Arico M, etc. HLH-2004: Diagnostic and therapeutic guidelines for hemophagocytic lymphohistiocytosis[J]. *Pediatr Blood Cancer* Infections associated with haemophagocytic syndrome. Reactive hemophagocytic syndrome in adult systemic disease:report of twenty-six-cases and literature review. *Advances in Macrophage Activation Syndrome* Ma Jie, Zheng Wenjie, Tang Fulin.

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AB0576 FEATURES OF NEUROLOGICAL MANIFESTATIONS OF SYSTEMIC LUPUS ERYTHEMATOSUS IN THE KYRGYZ COHORT OF PATIENS

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Background: The development of a variety of neuropsychiatric symptoms in SLE patients starts with the dysregulation of the immune system. Involvement of the CNS in the pathological process during the onset of the disease can significantly worsen the patient's condition and complicate the prognosis.

Objectives: To study the features of neurological manifestations of SLE in the Kyrgyz cohort of patients.

Methods: The basis of the study were the results of an initial examination of 460 patients with prospective observation, with a reliable diagnosis of SLE. Out of them, 112 patients were neurological, at the age of 30 (median) [26, 41], predominantly of Kyrgyz nationality (87), with duration of the disease from 6 months to 7 years (median – 2.0) treated in the National Cardiology and Therapy Centre named after M. Mirrahimov (NCTC) from January 2012 to December 2017. Neurologic manifestations of SLE were diagnosed by clinical examination by a neurologist using some instrumental methods of investigation.

To assess the neuropsychiatric state of patients, the classification criteria by the American College of Rheumatology (1999) were used. Neuropsychiatric disorders were diagnosed through a clinical examination by the psychotherapist using the classification of mental disorders and behavioural disorders according to ICD (International Classification of Diseases) – 10.

Results: The damage of the nervous system was observed in 112 (24.35%) Kyrgyz patients from 460, mainly of the central nervous system – in 77 (68.75%), to a lesser extent – the peripheral nervous system (PNS) – in 35 (31.25%) out of 112. In half of the observed patients with CNS lesions, various neuropsychiatric disorders were registered – in 39 (50.65%) out of 77, with psychosis prevalence with visual and auditory hallucinatory syndrome – in 34 (87.18%) out of 39 patients. There were 14 patients with cerebrovascular infection (18.18%) out of 77, mainly with ischaemic stroke in the SMA basin. There was one patient with aseptic meningoencephalitis with manifestations of mild hemiparesis and with the disruption of the function of the pelvic organs in the form of mandatory urges. Symptoms of myelopathy with malfunction of the pelvic organs were registered in 9 (11.69%) patients out of 77.

Five patients had symptoms of combined CNS and PNS lesions. In one of them cases the patient had encephalopolyneuroradiculopathy of the Guillain-Barre type with the phenomena of coarse tetraparesis and with disruption of the function of pelvic organs in the form of delay, with a single epicrisim.

Conclusions: Neurological manifestations of SLE in the majority of Kyrgyz patients were mainly caused by CNS lesions (68.75%), mostly in the form of neuropsychiatric disorders (50.65%), with a predominance of psychosis with visual and auditory hallucinatory syndrome (87.18%).

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AB0577 CHARACTERISTICS OF VALVULAR HEART DISEASE IN A MONOCENTRIC MALAYSIAN LUPUS COHORT

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Background: Valvular heart disease (VHD) is one of the commonest cardiovascular manifestations of autoimmune disease, including systemic lupus erythematosus¹. Its prevalence ranges from 18% to 74%, with predominantly regurgitant lesions and valvular thickening. The cause of such abnormalities is yet unclear. There have been very few studies performed in Asian patients, and only one in Malaysian patients to date.

Objectives: We sought to evaluate the valvular abnormalities of a multiracial cohort of lupus patients and also examine the association of these abnormalities with potentially related factors.

Methods: The medical records of all patients with SLE who had attended the Rheumatology Clinic of Kuala Lumpur Hospital between January 2008 and December 2016 were reviewed. All patients fulfilled the 1997 updated ACR criteria for SLE. Transthoracic echocardiographic examinations as part of their standard of care were performed by trained technicians using a standardised protocol. Multivariable logistic regressions were conducted to determine factors associated with valvular abnormalities in these patients.

Results: There were 207 patients in this study. VHD was found in 67.1% of patients: they were asymptomatic and did not have murmurs. The main abnormalities were tricuspid regurgitation (TR) (45.4%), mitral regurgitation (MR) (40.6%) and valvular thickening (19.2%). The majority of cases comprised mild TR (37.2%), mild MR (25.1%) and mitral valve thickening (10.6%). There were more patients with moderate to severe MR compared to TR (15.5% vs 8.2%). Median pulmonary artery systolic pressure was 23.0 mmHg (IQR 13.2). Risk of VHD was higher in patients with lower C3 levels (<0.83 g/L) (OR 3.36, 95% CI 1.74–6.66). MR (83.3%) and mitral valve (MV) thickening (58.3%) were the commonest abnormalities in patients with APS. Having APS greatly increased the risk of MR (OR 8.10, 95% CI 1.65–77.95) and mitral valve thickening (OR 16.30, 95% CI 3.93–73.84). Risk of mitral valve thickening was increased by presence of lupus anticoagulant (OR 20.82, 95% CI 3.64–223.08), anticardiolipin antibody (OR 5.24, 95% CI 1.42–17.76), anti-β2GPI antibody (OR 6.35, 95% CI 1.18–37.14) and diabetes (OR 7.85, 95% CI 1.43–40.24). Risk for MR was increased by presence of lupus anticoagulant (OR 3.55, 95% CI 1.10–12.43). Hypertension was associated with an increased risk for developing aortic regurgitation (OR 4.70, 95% CI 1.67–13.44). Anti-histone antibody positivity appeared to protect against development of MR (OR 0.46, 95% CI 0.213–0.968).

Conclusions: More than half of our patients had VHD, with regurgitant lesions and valvular thickening being the commonest abnormalities. Our finding of the association of MR and mitral valve thickening with APS and individual antiphospholipid autoantibodies concurs with earlier studies. However, what were unusual about our cohort were that TR was the prevalent abnormality and the association of VHD with low C3. We suggest that all patients with SLE have at least one echocardiogram, especially the patients with APS or antiphospholipid antibodies.

REFERENCE:

- [1] Bourre-Tessier J, Huynh T, Clarke AE, Bernatsky S, Joseph L, Belisle P, et al. Features associated with cardiac abnormalities in systemic lupus erythematosus. *Lupus*. 2011;20(14):1518–25

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AB0578 ANALYSIS OF CLINICAL FEATURES AND RISK FACTORS OF IN-HOSPITAL MORTALITY IN CYTOMEGALOVIRUS (CMV) DISEASES WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: Cytomegalovirus (CMV) is known as a major cause for life-threatening complications in immunocompromised hosts, including status post allogeneic bone marrow transplantation, solid organ transplantations, and acquired

immunodeficiency syndrome (AIDS). The reactivation of CMV depends on the immune status of host. Systemic lupus erythematosus (SLE) often requires steroid and immunosuppressive agents to induce remission and lower disease activity. This study presents the clinical presentations, laboratory characteristics, medical profile (including steroid, immunosuppressive agents, and biological agents) and clinical outcomes in SLE patients with diagnosis of CMV diseases. Further, we attempted to investigate the mortality risk factor in these patients.

Objectives: To analyse the clinical features, the mortality risk factors and all-cause mortality of Cytomegalovirus (CMV) diseases in patients with systemic lupus erythematosus (SLE) in single centre of Taiwan.

Methods: A retrospective study was performed to investigate the clinical features and identify the mortality risk factors associated with CMV diseases in patients with systemic lupus erythematosus (SLE). We reviewed the medical records in patients with SLE who were diagnosed with CMV diseases between Jan. 2006 and Dec. 2016 from Taipei Veterans General Hospital in Taiwan. Clinical and laboratory parameters as well as treatment outcomes were analysed.

Results: Fifty-six patients diagnosed with CMV diseases were enrolled in the study and separated into survivors ($n=24$) and non-survivors ($n=32$) groups. All patients in CMV disease demonstrated significantly high incidence of CMV pneumonitis (71.43%). The higher SLEDAI-2000 score ($p=0.009$, HR1.154, 95% CI 1.037–1.285), percentage of recent pulse therapy ($p=0.013$, HR 4.569, 95% CI 1.313–15.902), and plasmapheresis during hospital course ($p=0.005$, HR6.905, 95% CI 1.637–29.122) was more common characteristics in non-survivor group than in survivor group. Non-survivors had significantly higher percentage of pancytopenia ($p=0.001$, HR9.667, 95% CI 2.307–40.511), CMV-positive PCR of blood and bronchoalveolar (BAL) lavage fluid (Blood: $p<0.001$, HR 15.000, 95% CI 3.932–57.223, BAL fluid: $p=0.021$, HR 6.176, 95% CI 1.151–33.151), and presence of concurrent infections (bacteremia: $p=0.026$, HR 4.833, 95% CI 1.122–20.824, other fungal infections: $p<0.001$, HR 11.424, 95% CI 2.722–47.952) than survivors. Septic shock ($n=10$, 41.2% of non-survivor group) is the most common cause of in-hospital mortality in CMV diseases.

Conclusions: The recent pulse therapy, pancytopenia, and concurrent infections are risk factors of in-hospital mortality in CMV diseases of patients with Systemic Lupus Erythematosus. The serological data of non-survivor groups showed negative findings of CMV immunoglobulin M (IgM) with detection of CMV DNA by polymerase chain reaction (PCR) was observed in CMV diseases. The pulmonary haemorrhage and acute respiratory distress syndromes (ARDS) were the factors of in-hospital mortality in CMV pneumonitis.

REFERENCES:

- [1] N Berman and HM Belmont Disseminated cytomegalovirus infection complicating active treatment of systemic lupus erythematosus: an emerging problem. *Lupus*. 2017 Apr;26(4):431–434. DOI:10.1177/0961203316671817
- [2] Tsai WP, Chen MH, Lee MH, Yu KH, Wu MW, Liou LB. Cytomegalovirus infection causes morbidity and mortality in patients with autoimmune diseases, particularly systemic lupus: in a Chinese population in Taiwan. *Rheumatology international*. 2012; 32(9):2901–8. DOI:10.1007/s00296-011-2131-4

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AB0579 IMMUNOLOGICAL RESULTS OF SALIVARY GLAND BIOPSY AND THEIR RELATIONSHIP WITH CLINICAL AND SEROLOGICAL PARAMETERS IN PRIMARY SJÖGREN'S SYNDROME

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Background: Positive minor salivary gland biopsy (MSGB) is a major criteria for the diagnosis of primary Sjögren's Syndrome (PSS). In our centre the MSGB analysis is carried out by Immunology and Pathology Department in parallel. The immunological analysis identifies the lymphocytic composition of the inflammatory infiltrate. Their results show: the number of T and B cells, the ratio between CD4 and CD8 T lymphocytes and other non infiltrating lymphocytes.

Objectives: The goal of our study was to evaluate whether there is an association between the lymphocytic composition of the MSGB with the clinical and serological findings of PSS patients.

Methods: Patients diagnosed of PSS according American-European criteria (2002) underwent . MSGB between February and November of 2017

Demographic (sex and age), clinical (disease duration, xerostomia, keratoconjunctivitis sicca, Schirmer test, systemic disease) data were collected. Present or previous treatment with steroids and/or immunosuppressive therapy, serological studies such as ANA, RF, anti Ro and anti La were also included. MSGB data with the number of infiltrates, quantitative composition of T and B lymphocytes, CD4/CD8 ratio and presence of other non infiltrating lymphocytes were registered. Pathology data concerning Chisholm-Mason scale, presence of fibrosis, atrophy and size of infiltrate (small, moderate and severe) were also registered. A multiple logistic regression for each item of the immunological analysis adjusted for sex and age was made. We also measured the odds ratio and performed correlation test for all variables included.

Results: Table 1 and 2 summarise our cohort characteristics. The presence of T lymphocyte was associated with B lymphocyte, OR 99.21 (IC95% 5.12–1921, $p=0.002$) and with higher CD4/CD8 ratio, OR 17.34 (IC95% 1.45–206.15, $p=0.024$). CD4/CD8 ratio was also associated with the presence of T lymphocytes OR 10.54 (IC95% 2.16–51.50, $p=0.004$) and B lymphocytes were associated with the presence of T lymphocytes as well OR 5.38 (IC95% 1.63–17.72, $p=0.006$). Other non infiltrating lymphocytes were composed of CD8 T cells and were associated with a positive Schirmer test OR 17.47 (IC95% 1.62–188.13, $p=0.018$) but inversely associated with Chisholm-Mason grade ≥ 3 OR 0.09 (IC95% 0.014–0.58, $p=0.011$). There was no other association observed with clinical or analytical parameters. Colinearity test between pathological and immunological analysis was negative.

Abstract AB0579 – Table 1. Immunology, serological and treatment characteristics of patients with PSS

Variable	Absolute number (patient)	Proportion %
Men	26	45.5%
Systemic disease	34	36.2%
ANA titration		
<1/80	15	25.9%
1/80	10	17.2%
1/160	13	22.4%
1/320	10	17.2%
1/640	7	12.1%
1/1280	3	5.2%
ANA Pattern	64	
Speckled	34	53.1%
Homogenous	13	20.3%
Speck & Homogenous	12	18.8%
Others	5	7.8%
Ac. Anti Ro	5	5.2%
Ac. Anti La	0	0%
Rheumatoid Factor	11	19.0%
Other		
Antiphospholipid Ab	5	8.6%
Crioglobulines	2	3.4%
Immunosuppressant	5	8.6%
<6mo		
Immunosuppressant	14	24.2%
>6mo		
Corticosteroids <6mo	8	13.8%
Corticosteroids >6mo	15	25.9%

Abstract AB0579 – Table 2. Immunological analysis

Variable	Absolute number (patient)	Proportion %
Number of Infiltrates		
0	3	5.2%
1	32	55.2%
2	6	10.3%
3	16	27.6%
Number of T cells		
<30	11	19.0%
30-50	17	29.3%
50-80	11	19.0%
80-100	2	3.4%
100-120	9	15.6%
≥120	8	13.8%
CD4/CD8 Ratio		
0	11	19.0%
1/2	2	3.4%
1/1	3	5.2%
2/1	19	32.8%
3/1	19	32.8%
4/1	4	6.9%
Number of B cells		
<30	17	29.3%
30-50	11	19.0%
50-80	6	10.3%
80-100	4	6.9%
100-120	9	15.5%
≥120	7	12.1%
Other infiltrates		
None	38	65.5%
CD8 cells	19	32.8%
CD4+CD8 cells	1	1.7%
Inadequate sample	1	1.7%

Conclusions: MSGB in our PSS patients demonstrated an association between T lymphocytes, B lymphocytes and CD4/CD8 ratio. The infiltrate is mostly based on CD4 more than CD8 T cells. Other significant findings were the association between CD8 T lymphocytes and Chisholm-Mason scale grade ≥ 3 , regardless of the number of infiltrates. No correlation or colinearity was observed with the number of infiltrates by immunological analysis and the Chisholm-Mason grade reported by the pathology analysis.

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