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SAT0222 CLINICAL SPECTRUM AND LONG TERM FOLLOWUP OF SYSTEMIC LUPUS ERYTHEMATOSUS-ASSOCIATED MACROPHAGE ACTIVATION SYNDROME

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Background: Systemic lupus erythematosus (SLE) patients present with variable clinical features ranging from mild joint and skin involvement to life-threatening organ involvement such as nephritis, neuropsychiatric involvement, diffuse alveolar hemorrhage, and hemophagocytic lymphohistiocytosis (HLH). HLH is rare, but fatal complication of SLE. Recently, European League Against Rheumatism, the American College of Rheumatology, and the Pediatric Rheumatology International Trials Organization was to develop a set of classification criteria for MAS complicating systemic juvenile idiopathic arthritis (PRINTO criteria) [1]. Sung Soo Ahn and his colleagues reported PRINTO criteria predicted mortality of adult SLE patient, but they followed only one year [2].

Objectives: To reveal association PRINTO criteria with long term mortalities in SLE patient in our Hospital.

Methods: We performed a retrospective analysis of SLE patients who received moderate dose glucocorticoid therapy (>0.4mg/kg/d) in our hospital between April 2008 and April 2019. Patients were evaluated for HLH using the 2016 PRINTO

classification criteria for MAS. Clinical features and laboratory findings were compared and overall survival rate was analyzed.

Results: Among 164 episode (144 patients) with SLE, 31 episode (31 patients) 5.2% were considered to have MAS on admission.

The overall survival rate was significantly lower in patients with MAS than without MAS (86.2% vs. 95.3%, $p = 0.048$). Interestingly, SLEDAI had no association with mortality, relapse rate, and MAS complication. SLEDAI more focused on renal and neuropsychiatric symptoms than hematologic features. So SLEDAI might not be associated with MAS secondary to SLE. Furthermore, we observed no death patient with MAS after one year, and only 1 case relapse in MAS patient. So MAS might have fatal but less relapsing property compared with other lupus cases.

Conclusion: PRINTO criteria may be useful to differentiated fatal MAS patients from others. Further investigations are required to confirm our findings. **Limitation** : The main limitations of our study include its retrospective design, single center site, and that the number of admitted patients with SLE was small.

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SAT0223 TUBULO-INTERSTITIAL INFILTRATES IN LUPUS NEPHRITIS

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Background: Lupus nephritis (LN) occurs in up to 60% of patients affected by Systemic Lupus Erythematosus (SLE). The presence of inflammatory infiltrates in glomeruli and/or in tubulo-interstitium (TI) plays an important role in terms of prognosis (1). Ectopic lymphoid structures (ELSs) are clusters of organized lymphoid infiltrates forming at sites of chronic inflammation in non-lymphoid organs (2). Data on ELSs in kidneys of SLE patients are scant (3-5).

Objectives: The aim of the study was to evaluate the tubule-interstitial infiltrates (TI-I) organization and to investigate the presence of ELSs.

Methods: Kidney sections of SLE patients undergoing a renal biopsy for diagnostic purpose were studied; LN was diagnosed according to the 2003 International Society of Nephrology / Renal Pathology Society classification criteria. Clinical, laboratory and histological data were collected in a standardized, computerized and electronically filled form, including past medical history. The presence of lymphoid aggregates and ELS in the kidney sections were firstly evaluated by hematoxylin-eosin (HE). By using immunohistochemistry (IHC), we characterized the infiltrates by identification of T cells (CD3), B cells (CD20) and follicular dendritic cells (CD21).

Results: By HE we evaluated 53 kidney samples from LN patients (F:M = 51:2, mean age at biopsy $\pm$ SD years 35 ± 7.7 ; mean disease duration at date of biopsy $\pm$ SD 8 ± 8.3 years). TI-I were found in 33 kidney specimens. By HE we identified a well-defined infiltrate pattern resembling ELS in 13 renal samples. In these samples, we confirmed the presence of organized infiltrates by IHC, with evidence of segregated T and B cells areas in most of them. In one sample, the CD21 staining was positive confirming the presence of ELS (Figure 1). Interestingly, this last patient, who failed cyclophosphamide and mycophenolate, responded to rituximab administration and is now in complete LN remission. Moreover TI-I was negatively correlated with renal remission after induction therapy ($P=0.03$) independent of the histological class and the induction treatment.