infections. Complement deficiency was the most frequent genetic variant (40%), followed by DNase variants (23.3%). Interestingly, genetic analysis revealed 12 novel gene variants. All patients were treated aggressively with corticosteroids and sequential conventional immunosuppressive drugs. Twenty-eight patients received biologic agents (14 belimumab, 12 rituximab, and 2 JAK inhibitors), while 23 received IVIG. Most of them showed a poor response to treatment. There were five deaths related to serious infections.

Conclusions This report expands the pathogenic variants and the clinical spectrum of monogenic lupus. Patients with various variants share clinical characteristics. The outcome is guarded by a high mortality rate. Unfortunately, to date, there is no effective standard treatment.

LSO-079 DYNAMIC CHANGES OF IMMUNE CELLS IN SYSTEMIC LUPUS ERYTHEMATOSUS WITH NEPHRITIS USING LONGITUDINAL PERIPHERAL BLOOD SINGLE-CELL RNA-SEQ

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Background Single-cell RNA-sequencing (scRNA-seq) has been recently applied in systemic lupus erythematosus (SLE) to define distinct cellular composition and transcriptional signatures, greatly expanding our understanding of SLE pathogenesis. However, since most of the studies were cross-sectional approach from a single moment in time, dynamic feature of immune cells over the disease course is yet to be revealed.

Methods We analyzed serial longitudinal PBMC samples (N=19) obtained at various time points of SLEDAI ranged from 4 to 19 in lupus nephritis (N=6) and from controls (N=33) using scRNA-seq (10X Chromium 5', TCR sequencing).

Results SLE showed different cellular composition; reduction of CD4+ T, monocytes, cDC, pDC, and increases of CD8+ T and B cells compared to controls. Expression of type 1 IFN signatures were increased in the most cell clusters. Cell to cell interaction analysis revealed activated CD8+ T cells were the most interactive cell population. Further analysis of CD8+ T cell showed increased T cell exhaustion markers and highest cytotoxicity in GZMH+ CD8+ Tem population in SLE. We also observed significantly decreased CD8+ T cell TCR diversity in SLE. To investigate dynamic transformation of immune cells during flare, especially CD8+ T cells, we analyzed selected paired samples before and after flare of lupus nephritis (N=12). In flare state, GZMH+ CD8+ Tem were expanded, displaying increased cytotoxicity with reduction of TCR diversity. We also observed expression of T cell exhaustion markers, but reduced Type 1 IFN signalling in flare state. Interestingly, the paired TCR repertoire and mitochondrial mutation analysis reveals expanded effector CD8+ T cell clonotypes are heterogeneous in each flare from the same patient, directing towards unique antigen convergence in flare stage.

Conclusions These results uncover dynamic transition of peripheral blood immune cells during flare of lupus nephritis, especially in CD8+ T cell population, suggesting its pivotal roles in SLE pathogenesis.

LSO-080 MACHINE-LEARNING APPROACH ON LUPUS LOW DISEASE ACTIVITY PREDICTION

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Background The development of lupus low disease activity state (LLDAS) as a treat-to-target endpoint for SLE patients has been validated. Its attainment has been associated with improved outcomes. This study aims to show whether a machine learning model can yield good results in predicting whether a patient will achieve LLDAS on their succeeding assessment.

Methods A total of 42,355 records of patients were retrieved from the APLC longitudinal study database. Three machine learning models – XGBoost, Random Forest, and Naive Bayes – were tested for their predictive power. Eighty percent of the data was used to train the models while thirty percent was used for validation. The data were normalized and all models were subjected to 10-fold cross-validation to prevent overfitting. Additionally, we compared the top ten most significant features of each model.

Results Various metrics were used to measure the model's predictive power. The results of our study showed that the Random Forest model scored the highest for specificity, PPV, and accuracy with 0.8450, 0.8182, and 0.8338, respectively. The XGBoost model topped the NPV metric with 0.8559 while the Naive Bayes model got the highest score for sensitivity with 0.8986. It is good to note that the score difference of Random Forest with the top sensitivity and NPV scores were only 0.0629 and 0.0085, respectively.

For the significant features, only two features were present on all three models, namely the current LLDAS and