Background Multiple organ systems including skin, musculoskeletal, neurological, renal and hematologic systems can be involved in patients with SLE. To date extensive research has been conducted to identify unique gene expression signatures using heterogeneous SLE cohorts, however little research has been conducted to delineate SLE signatures in patients with less common disease manifestations, such as cerebral lupus. The diagnosis and monitoring of patients with cerebral lupus is particularly challenging as traditional markers of lupus disease activity in peripheral blood are often negative, and the clinical symptoms overlap with many other non-immunological diseases. The aim of this study was to identify and interrogate immunological pathways that may be aberrant in this particular SLE disease subtype.

Methods Whole blood RNA was extracted from 46 SLE patients, 5 Autoimmune encephalitis and 20 healthy controls. Gene expression was measured using a Nanostring nCounter mRNA expression assay incorporating over 500 immunological genes. Data was analysed using Partek Genomics Suite to identify differentially expressed genes and find pathways that may be of interest to interrogate further in the context of SLE disease manifestations.

Results Clear differences in gene expression between healthy controls and SLE patients were evident. In our cohort of 46 SLE patients, seven had symptoms of cerebral lupus (seizure and/or psychosis). Cerebral lupus patients showed significantly different expression in 5 genes. TNFRSF1B, CD14 and IL1B expression was increased in cerebral lupus compared to other SLE, while PSMB7 and IKBKB was decreased in cerebral lupus compared to other SLE.

Conclusions The differences observed in this group of genes (CD14, PSMB7, IKBKB, IL1B and TNFRSF1B) implicate myeloid cell dysfunction as a possible point of difference between cerebral lupus and other forms of SLE. This preliminary finding may help explain the overlap between the innate and the adaptive immune dysregulation in lupus pathogenesis, and the use of TNF inhibition acutely in patients with a similar autoimmune cerebral disease, cerebral vasculitis.

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antihypertensive and corticosteroids. The immunosuppressant treatment used was: mycophenolate mofetil, azatioprine, cyclophosphamide, tacrolimus and rituximab. Considering that the time of follow-up of patients varies, each temporary space was analyzed according to the number of patients. Figure 1 shows that at 6 months, 38% of patients had an improvement >50% and 7.7% of patients achieved remission. After one year of treatment, 42% of patients presented improvement >50% and 17% achieved remission but in 23% of patients two changes of immunosuppressive treatment was needed. At 24 months, 50% of patients improved >50% and 25% achieved remission; in 13% of patients it was necessary to make another treatment change. Finally, after 24 months, it was observed that 50% of patients achieved remission, 33% of patients presented an improvement >50% and only 17% presented renal failure, and it was necessary to make another treatment change. Globally, in 36% of patients 1 or more changes of immunosuppressive treatment were necessary to achieve improvement >50% or remission.

Conclusions Patients who do not experience an improvement >50% in a period of 6 months are more likely to improve if a change in immunosuppressive treatment is made. It is necessary to extend the series to reach conclusions with statistical value.

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DAMAGE ACCRUAL IN SWEDISH SYSTEMIC LUPUS ERYTHEMATOSUS: SECONDARY SJÖGREN'S SYNDROME IS AMONG THE FACTORS ASSOCIATED WITH INCREASED RISK

Background Although the expected survival of patients with systemic lupus erythematosus (SLE) has improved during the last 50 years, accrual of damage remains a critical concern. Acquired damage is tightly linked to decreased quality-of-life and premature death, and could be due to raised disease activity, drug-related side-effects or co-morbidities.

Methods Accumulation of organ damage was studied in 543 well-characterized and from 1998 and onwards consecutively recruited prevalent/incident SLE cases meeting the 1982 American College of Rheumatology (ACR82) and/or the 2012 SLICC criteria. The SLICC/ACR damage index (SDI) was used to estimate damage. Disease variables were evaluated regarding association with damage accrual, and time to first damage. Detailed information on clinical and immunological features, as well as damage in each SDI domain, where at hands. Standardized mortality rate was calculated and causes of death recorded. Comparisons between groups were performed using Chi-square or Mann-Whitney U-tests, p-values<0.05 were considered significant.

Results 59% of the patients had an SDI score 1% and 25% had extensive damage defined as SDI 3. Patients with presence of damage (SDI 1) were significantly older at disease onset, had longer SLE duration and fulfilled further classification criteria. Caucasian ethnicity was more common among cases with damage. Having ACR82-defined neurologic disorder, antiphospholipid syndrome (APS), any anti-phospholipid antibody (positive IgG anti-2-GPI or a lupus anticoagulant test, separately) as well as concomitant co-morbidities such as hypertension, hyperlipidemia, diabetes, depression and secondary Sjögren's syndrome were associated with presence of damage (SDI 1). In addition to factors associated with SDI 1, serositis, renal and hematological disorder as well as interstitial lung disease and positive IgG anti-cardiolipin were associated with extensive damage (SDI 3). Time to first damage was significantly shorter for males and for cases with a positive lupus anticoagulant test, whereas APS patients were borderline significant. Cases with malar rash and anti-La/SSB antibodies had significantly longer time to first damage. Malignancy was the most common cause of death.

Conclusions Despite that Swedish health care is tax-funded and offers universal access, a considerable number of patients are affected by irreversible damage over time. We confirm previous observations for several damage associations and

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