Limbic encephalitis (LE) is a rare and serious neurological syndrome that selectively affects structures of the limbic system. The main clinical manifestations are seizures associated with episodic memory and behavioral disorders. They are essentially of viral or paraneoplastic origin. Dysimmunitary etiologies, in particular systemic lupus erythematosus, are rarely described and have recently been demonstrated (4–6). However, there remains a lack of understanding about the frequency and strength of this association.

Methods We report a clinical case of a patient with limbic encephalitis complicating SLE.

Results A 36-year-old woman was admitted to our service because of fever, headache, encephalopathy followed by severe, generalized seizures, memory disorders and altered state of consciousness. The patient had a history of SLE for 9 years, under hydroxychloroquine. At the admission, general examination revealed memory disorders. The antiphospholipid antibodies were negative. T2-weighted, fluid-attenuated inversion recovery magnetic resonance imaging showed hyper signals in the hippocampus and amygdala. At the lumbar puncture, the cerebrospinal fluid was normal. Anti-neuronal antibodies were negative. Whole-body computed tomography and positron emission tomography (PET-CT) excluded malignancy. The diagnosis of limbic encephalitis secondary to SLE was retained. Acyclovir was given under the presumptive diagnosis of human herpes virus encephalitis, but neither HHV-6 virus DNA nor anti-N-methyl-D-aspartate receptor antibody was detected in the Cerebrospinal fluid. High-dose corticosteroid therapy was initiated associated with polyvalent immunoglobulin. Monthly cures (X6) of cyclophosphamide were associated followed by mycophenolate mofetil. The patient also received antiepileptic therapy, hydroxychloroquine was maintained. Brain MRI at 3 months of treatment revealed hippocampal atrophy. The evolution at 8 months showed a slight improvement in memory disorders

Conclusions ALE is a rare complication of SLE. Therapeutic management is not codified, but the use of immunoglobulins, high-dose corticosteroid therapy and immunosuppressants are recommended. Rituximab is also used in refractory cases. The prognosis depends on the early diagnosis of specific therapeutic management.

PO.1.19 LUPUS VASCULITIS MANIFESTING AS EIGHT-AND-A-HALF SYNDROME

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Introduction: eight-and-a-half syndrome is an uncommon neurological disorder of the pontine tegmentum and the 7th cranial nerve, manifesting as conjugate horizontal gaze palsy (CHGP) and ipsilateral peripheral nerve paralysis (PFP). The main causes are cerebrovascular disease, demyelinating disorders, and infections. Neuropsychiatric systemic lupus erythematosus (SLE) as the etiology of internuclear ophthalmoplegia is exceedingly rare.

Case report A 38-year-old White female with a 22-year history of SLE presented with diplopia and left PFP. Four months previously, she started complaining of intermittent fever and headache, for which she sought medical care several times in another service, repeatedly being given the presumptive diagnosis of an acute viral illness. Upon admission in our hospital, physical examination was significant for CHGP, nystagmus, and gait ataxia. A rash on her upper trunk, suggestive of acute cutaneous lupus, was also noticeable. Laboratory results were remarkable for high titer anti-dsDNA (223 U/mL), low complement (C3 of 58 mg/dL, C4 of 9.7 mg/dL), and lymphopenia (250 cells/mm3). Cerebrospinal fluid (CSF) analysis revealed a mild pleocytosis (7 cells, 70% lymphocytes), protein of 80 mg/dL, and glucose of 43 mg/dL. Brain magnetic resonance imaging evidenced hyperintense lesions, on T2 and FLAIR sequences, on the white matter of the left centrum semiovale and the left facial colliculus, with mild enhancement on the pons, as well as foci of restricted diffusion on the left lateral ventricle and left basilar part of the pons, inferring microthrombi. Chest computed tomography exhibited mild bilateral pleural effusion. Transthoracic echocardiogram demonstrated mild pericardial effusion. Empirical treatment with acyclovir, ceftriaxone and ampicillin was started, covering the main possible etiologies of infectious rhombencephalitis. Culture for fungi and bacteria, real-time PCR assay for Listeria, and viral CSF panel all resulted negative, though, and the patient showed no improvement on antibiotics. Given the extensive evidence of multisystemic lupus activity, the patient was pulsed with methylprednisolone and cyclophosphamide, with satisfactory evolution.

Conclusion we presented an atypical case of cerebral vasculitis due to SLE presenting as eight-and-a-half syndrome.

PO.1.20 PROGNOSTIC BIOMARKERS OF ORGAN DAMAGE IN PATIENTS WITH NEWLY DIAGNOSED SLE

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Purpose The urokinase plasminogen activator receptor (uPAR) is expressed on various cell types and is involved in proteolysis, migration and adhesion. A soluble form (suPAR) is yielded at shedding and has emerged as a severity biomarker in malignancies, inflammatory and infectious diseases.[1] Previously, high levels of suPAR at systemic lupus erythematosus (SLE) diagnosis was shown to associate with future organ damage in an international SLE cohort.[2] Herein, the aim was to confirm suPAR as a prognostic biomarker of organ damage in well-characterized Swedish patients with recent-onset SLE and to compare its prognostic value with other proteins detected in blood.

Methods Serum samples from 274 newly diagnosed (<6 months) SLE patients from four Swedish referral centers (Linköping, Lund, Stockholm, Uppsala) were analyzed for

suPAR (ELISA; Virogates, Denmark) and for 184 potential biomarkers (Inflammation and Organ damage panel, Olink, Sweden). Protein levels were related to organ damage development as defined by the Systemic Lupus International Collaborating Clinics/American College of Rheumatology damage index (SDI). Clinical follow up-data have so far been extracted for 210 of the patients.

Results A total number of 38 patients (18%) had accrued organ damage before the 1-year follow-up, 75 patients (37%) had developed damage after 3 years, and 88 patients (47%) after 5 years. In addition to suPAR, 9 proteins differed significantly ($p<0.01$, Mann-Whitney) between patients with SDI$\geq$1 and without damage (SDI=0) at both the 3- and 5-year follow-up. The top three significant proteins ($p<0.001$ at the 3-year follow-up) were G protein-coupled receptor 56 (GPR56), interleukin-18 receptor 1 (IL-18R1) and carbonic anhydrase 14 (CA14) of which the latter was decreased among patients who developed organ damage.

Conclusions The accumulation of organ damage is tightly associated with morbidity and mortality in patients with SLE. The biomarker suPAR, already validated for clinical triaging in acute care settings, could identify patients early who have a high risk to develop organ damage. These patients are thus in need of intensified follow-up. Additional data extraction and analysis will clarify the potentials of suPAR, GPR56, IL-18R1, CA14 and other proteins to predict development of organ damage, facilitate risk stratification, guide follow-up intensity, and provide tailored pharmacotherapy in SLE.

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PO.1.21 ATTAINMENT OF EQ-5D-3L FULL HEALTH STATE AFTER THERAPY IS ASSOCIATED WITH DECELERATED ORGAN DAMAGE ACCRUAL IN SYSTEMIC LUPUS ERYTHEMATOSUS

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Purpose Within the frame of a survey of the impact of patient-reported evaluation on long-term outcomes in systemic lupus erythematosus (SLE), we herein investigated whether attainment of self-reported full health state (FHS) following therapeutic intervention for active disease was associated with a lower probability to accrue organ damage.

Methods Data from the open-label (OL) extension periods of the BLISS-52 and BLISS-76 clinical trials of belimumab in SLE were used (N=973). FHS was defined as an experience of 'no problems' in all five dimensions of the EuroQol 5-Dimension health questionnaire three-level version (EQ-5D-3L). We also investigated potential associations between experience of 'no problems' in each one of the EQ-5D dimensions and organ damage accrual separately. Proportional hazards (Cox) regression analysis was employed for time-dependent associations between EQ-5D-3L responses at the OL baseline and the first documented increase in organ damage, as assessed using the Systemic Lupus International Collaborating Clinics (SLICC)/American College of Rheumatology (ACR) Damage Index (SDI).

Results FHS was associated with lower probability of and/or longer time to organ damage progression (adj. HR: 0.62; 95% CI: 0.38–0.99; P=0.047) after adjustment for potential confounders, as was experience of 'no problems' regarding mobility (adj. HR: 0.61; 95% CI: 0.43–0.87; P=0.006) and pain/discomfort (HR: 0.66; 95% CI: 0.44–1.00; P=0.049). No such association was seen in multivariable analyses for the other dimensions of EQ-5D; however, 'no problems' with regard to self-care showed a trend towards a statistically significant association with decelerated organ damage accrual (adj. HR: 0.65; 95% CI: 0.42–1.01; P=0.054).

Conclusions Experience of FHS and 'no problems' regarding mobility or pain/discomfort after therapeutic intervention for active SLE were associated with reduced subsequent risk of organ damage accrual, suggesting that optimisation of these health-related quality of life (HRQoL) aspects is a clinically relevant treatment target in patients with SLE.

PO.1.22 ONLINE EDUCATION SIGNIFICANTLY IMPROVED RHEUMATOLOGISTS' KNOWLEDGE AND CONFIDENCE IN MINIMIZING LONG-TERM ORGAN DAMAGE IN SLE

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Background/purpose Long-term organ damage (LTOD) is a key issue with SLE. SLE is often managed using glucocorticoids (GCs), which can lead to organ damage if used for longer or at higher dosages. It is important that physicians understand how to use GCs, what the drivers and risk factors are for accrual of organ damage, and which treatment strategies can minimize LTOD in clinical practice.

Methods Rheumatologists participated in two online activities: 'Addressing the Problems of Long-term Organ Damage in SLE' (launched 15 April 2021, data collection by 29 June 2021) and 'Appropriate Use of Glucocorticoids in the Treatment of SLE' (launched 24 June 2021, data collection by 22 September 2021). Educational effect was assessed using a repeated-pair design, pre-/post-assessment. A paired samples t-test was conducted for significance testing on overall average number of correct responses and for confidence rating. Cohen's d estimated the effect size of the education on number of correct responses (<0.20 modest, .20-.49 small, .59-.79 moderate, $\geq$.80 large). A series of McNemar's tests were conducted at the question level (5% significance level, $P<0.05$).

Results

Activity 1:

- Rheumatologists (n=71) significantly improved their knowledge of the prevalence of LTOD ($P<0.001$), the relationship between SDI (Systemic Lupus International