Results Patients with SLE had higher cfDNA (1.69 ± 0.23 vs 1.42 ± 0.31 ng/mL, $p=0.0003$), MPO (1607 ± 2353 vs 1503 ± 1106 , $p<0.05$) and BAFF levels (160735 ± 2353.23 vs 891.16 ± 184.31 , $p<0.05$). DNase concentration was also lower in healthy controls (5.84 ± 5.72 vs 9.38 ± 6.97 , $p<0.05$). BAFF showed strong correlation with anti-dsDNA antibodies determined by ELISA method ($\rho=0.564$, $p=0.000$), MPO activity ($\rho=0.256$, $p=0.021$), DNase I concentration ($\rho=0.27$, $p=0.012$) and cfDNA concentration ($\rho=0.262$, $p=0.014$). DNA determined by ELISA test showed correlations with DNase concentration ($\rho=0.249$, $p=0.022$), DNA determined by CLIFT ($\rho=0.341$, $p=0.001$) and C3 complement component ($\rho=-0.4$, $p=0.023$). MPO activity showed correlations with cfDNA levels ($\rho=0.386$, $p=0.001$), DNase concentration ($\rho=0.501$, $p=0.000$), and anti-MPO antibodies ($\rho=0.293$, $p=0.006$). Cell free DNA levels additionally correlated with DNase activity ($\rho=0.288$, $p=0.007$) as well with netolytic activity ($\rho=0.244$, $p=0.026$). Netolytic activity also correlated with anti-dsDNA antibodies determined by ELISA ($\rho=-0.299$, $p=0.039$).

Conclusions Increased NETs' footprints (myeloperoxidase and cfDNA) are present in lupus sera. As probably compensatory mechanism increased DNase I concentrations also were found in lupus sera. NET burden is followed by production of various antibodies recognising different NET structures.

S3d – APS

S3D:4 REAL LIFE SINGLE CENTRE RESULTS ON RITUXIMAB TREATMENT OF PATIENTS WITH PRIMARY AND LUPUS ASSOCIATED ANTIPHOSPHOLIPID SYNDROME

E Kiss, A Szappanos, A Wiedemann, A Balogh, M Szabo, G Poor. *National Institution of Rheumatology and Physiotherapy, Budapest, Hungary*

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Recurrent thrombotic events occur in 20% of patients with APS despite of adequate antithrombotic therapy. As phospholipid-cofactor antibodies are key elements of pathophysiology, B cell depleting rituximab is a rational therapeutic approach.

Aims were to identify and characterise APS patients receiving rituximab. All together 179 (60 primary, 119 lupus-associated) APS patients were found in our database of whom 15 (8.4%) were treated with RTX. As younger and male patients dominated in RTX +group, 15 from RTX naive patients were matched regarding age and gender for a case control study. First symptoms appeared at the age of 30 in both groups. Patients were followed for 4 and 7 years, the diagnosis delayed 4 and 1 year in RTX +and RTX- groups, respectively. This 3 year difference might contribute to higher disease severity. Primary APS (47% vs 32%), cerebrovascular events (13 vs 2), valvular heart disease (5 vs 2), and high risk triple aPL positivity (13 vs 5) were more prevalent in RTX +group. All 4 deep venous thrombosis cases in the RTX +group were complicated with pulmonary embolism in comparison with 1 PE within 6 RTX- DVT patients. Raynaud syndrome resulted in digital ulcer more frequently in RTX +than in RTX- group (4/4 vs 1/6). Antimalarials were given to 3 and 10, cumarin or NOAC were initiated in 15 and 5 patients in the RTX +and RTX- groups, respectively. Rituximab was started 2

(0.5–6) years after diagnosis. Within 10 (3–60) months follow-up on RTX there were no incident thrombotic events. Two thirds of patients could stop steroid. RTX was stopped in 6 cases: 3 due to remission (of whom 2 relapsed), 1 LFU, and 2 adverse events (1 longstanding B cell depletion, 1 peroneus paresis). Other AEs were 1 mild infusion reaction, 1 leukopenia, 1 UTI. As summarised, delay in APS diagnosis and male gender were associated with more severe disease and the need for rituximab. Antimalarials and associating lupus were identified as markers of more favourable disease outcome. In patients with recurrent thrombotic events despite of adequate anticoagulant therapy, RTX can be a rational and effective choice with favourable safety profile.

S3D:5 PLASMA SOLUBLE TRIGGERING RECEPTOR EXPRESSED ON MYELOID CELLS-1 IS ELEVATED IN PATIENTS WITH THROMBOTIC PRIMARY ANTIPHOSPHOLIPID SYNDROME

¹Y Molad, ^{1,2}Y Edel, ^{1,2}E Pokroy-Shapira, ^{1,2}S Oren, ^{1,2}A Dortort, ¹Y Pri-Paz Basson, ^{2,3}T Shochat, ^{2,4}V Kliminski. ¹Rheumatology Unit, Beilinson Hospital-Rabin Medical Centre, Petach Tikva, Israel; ²Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel; ³Bio-Statistical Unit, Beilinson Hospital, Rabin Medical Centre, Petach Tikva, Israel; ⁴Laboratory of Inflammation Research, Felsenstein Medical Research Centre, Rabin Medical Centre, Petach Tikva, Israel

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Background Antiphospholipid antibodies (APLA) are necessary, but not sufficient for the development of thrombosis in APS. Soluble triggering receptor expressed on myeloid cells-1 (sTREM-1) is an innate-immune receptor found in the blood and reflects innate immune cells activation.

Aim To determine if plasma sTREM-1 can be served as a biomarker for thrombosis in patients with PAPS.

Methods A cross-sectional, case-control study. Plasma level of sTREM-1 was analysed by ELISA in a group of consecutive patients diagnosed with either PAPS (Sapporo criteria) or asymptomatic persistently positive APLA, and healthy controls (HC).

Results The study group comprised of 33 patients with PAPS (age 52.0 ± 17.4 years.), 10 asymptomatic APLA-positive patients (50.6 ± 17.9 years.) and 73 HC (42.6 ± 13.3 years.). The mean plasma sTREM-1 level was significantly higher in the PAPS group compared to HC (316.3 ± 119.3 pg/ml, vs 230.2 ± 85.5 pg/ml, $p=0.0002$), as well as past obstetric APS (195.12 ± 58.52 pg/ml, $p=0.014$), and asymptomatic APLA ($215.8 \pm$ pg/ml vs HC, $p=0.019$). Plasma sTREM-1 in PAPS patients with an acute event of thrombosis was significantly higher than in patients with past thrombotic event ($p=0.012$), past obstetric APS ($p=0.0001$) and HC ($p<0.0001$). Plasma sTREM-1 level was significantly higher in PAPS patients who ever had stroke ($p=0.007$) or venous thromboembolic event ($p=0.018$). On receiver operator curve (ROC) analysis, plasma sTREM-1 showed an area under the curve (AUC) 0.7292 in differentiating between thrombotic APS (ever) and non-thrombotic APS or asymptomatic APLA-positivity. A multivariate regression model to predict sTREM-1 level by thrombotic PAPS ever, age and sex found that sTREM-1 level is independently associated with thrombotic PAPS ($p<0.004$) as well as with female gender ($p=0.017$) and older age ($p=0.0006$). Plasma sTREM-1 level was neither associated with anti-cardiolipin, anti- β_2 glycoprotein I (IgG/IgM/IgA) Abs' titers and/or

lupus anticoagulant positivity, nor with single-, double- or triple- APLA positivity.

Conclusion Our data show for the first time that plasma sTREM-1 level is significantly elevated in patients with thrombotic PAPS. We suggest that soluble TREM-1 might be used as a biomarker for thrombosis in patients with primary APS and our results support the possible role for the innate immune system in the pathogenesis of thrombosis in PAPS.

S3D:6 MICROANGIOPATHIC MANIFESTATIONS OF THE PRIMARY ANTIPHOSPHOLIPID SYNDROME

J Zhao, Y Sun, Q Wang, M Li, X Zeng. Peking Union Medical College Hospital, Beijing, China

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Objective Thrombotic microangiopathy (TMA) is a relatively rare but severe vascular complication of antiphospholipid syndrome (APS) and may involve any organ. It's crucial for prompt diagnosis and early intervention, while the clinical features and outcomes of patients with TMA in APS is quite scant. The aim of this study was to investigate characteristics of primary APS with TMA patients.

Patients and methods One hundred and twelve primary APS patients who attended in Peking Union Medical College Hospital from January 2004 to December 2016 were enrolled. Patients were subdivided into two groups: PAPS with TMA and PAPS without TMA. Demographic data, clinical characteristics, laboratory features and treatment were retrospectively collected. Bivariate statistical analysis and logistical regression test were performed to compare the discrepancy between the two groups. Survival rates were studied by Kaplan-Meier method, and COX proportional hazard model was adopted to perform the analysis of predicting factors for mortality.

Results Twenty-one patients with TMA were identified, presenting in 18.8% of all PAPS patients. Both groups were similar with respect to age of onset, gender, clinical course and pregnancy morbidity. However, the frequency of anti-b2-glycoprotein I antibodies (anti-b2GPI) (66.7% vs 33.0%, $p=0.004$) was significantly higher in the TMA group than the non-TMA group. In addition, thrombocytopenia (85.7% Vs 54.9% $p=0.007$) and elevated high-sensitivity C-reactive protein (hsCRP) (71.4% Vs 54.9% $p=0.007$) were observed more frequently among TMA group. In multivariate analysis, only thrombocytopenia (OR=4.055, 95% CI: 1.006 to 16.345,

Abstract S3D:6 Figure 1

$p=0.049$), hsCRP elevation (OR=6.789, 95% CI: 2.018 to 22.840, $p=0.002$) and positivity for anti-b2GPI (OR=4.723, 95% CI: 1.409 to 15.830, $p=0.012$) were independent risk factors for the occurrence of TMA in PAPS patients. The overall 1, 3, and 5 year survival rate of PAPS with TMA was 69.2%, 60.6% and 50.5% respectively; while 90.4%, 88.2%, and 88.2% in APS patients without TMA ($p<0.05$).

Conclusion Microangiopathic antiphospholipid syndromes should be an independent subset with mainly small vessels involved and worse prognosis, compared with classic APS.

S3D:7 CEREBRAL HYPOPERFUSION DETECTED BY PERFUSION-WEIGHED MRI MAY ASSIST THE DIAGNOSIS OF PRIMARY DIFFUSE NEUROPSYCHIATRIC LUPUS ERYTHEMATOSUS

¹E Papadaki, ²A Fanourakis, ¹E Kavroulakis, ¹D Karageorgou, ³P Sidiropoulos, ³G Bertias, ⁴P Simos, ²D Boumpas. ¹University of Crete, Department of Radiology, Heraklion, Greece; ²National and Kapodestrian University of Athens, 4th Department of Internal Medicine, Athens, Greece; ³University of Crete, Department of Rheumatology, Heraklion, Greece; ⁴University of Crete, Department of Psychiatry, Heraklion, Greece

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Objective Abnormalities in regional or global cerebral perfusion have been reported in SLE but their value in distinguishing lupus from non-lupus related diffuse neuropsychiatric events has not been determined. We examined whether the addition of dynamic susceptibility-contrast-enhanced T2*-weighted perfusion MRI (DSC-MRI), a non-invasive assessment of brain haemodynamic status, to the standard MRI examination suggested by the EULAR recommendations, may be of added value in the clinical diagnosis and attribution of neuropsychiatric SLE (NPSLE).

Patients and methods Seventy-six SLE patients (53 NPSLE, 23 non-NPSLE) and 31 healthy controls underwent conventional MRI (cMRI) and DSC-MRI. Attribution of NPSLE to lupus (primary NPSLE: $n=37$) or not ($n=16$) was based on multi-disciplinary assessment including cMRI results and response to treatment. Cerebral blood volume (CBV) and cerebral blood flow (CBF) values were estimated in 18 normal-appearing white matter (NAWM) and deep grey matter (NADGM) areas. Perfusion differences among subgroups and their diagnostic utility were assessed using Analysis of Variance, Receiver Operating Characteristics, and Binary Logistic Regression analysis.

Results The most common manifestations were mood disorder, cognitive disorder and headache. The most common manifestations were mood disorder, cognitive disorder and headache. Primary NPSLE patients had lower cerebral blood flow and volume in several NAWM areas compared to controls ($p<0.0001$), and lower cerebral blood flow in the semioval centre bilaterally compared to non-NPSLE and non-primary NPSLE patients ($p<0.001$). A cut-off for cerebral blood flow of 0.77 in the left semioval centre discriminated primary NPSLE from non-NPSLE/non-primary NPSLE with 80% sensitivity and 67%–69% specificity. Blood flow values in the left semioval centre showed substantially higher sensitivity than cMRI (81% versus 19%–24%) for diagnosing primary NPSLE and the combination of the two modalities yielded 94%–100% specificity in discriminating primary from non-primary NPSLE.

Conclusion Primary NPSLE is characterised by significant hypoperfusion in white matter and deep grey matter areas that appear normal on conventional MRI sequences. Addition