PO.2.44 CATASTROPHIC ANTI-PHOSPHOLIPID SYNDROME AND PERIPARTUM: A NEW OBSERVATION!

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10.1136/lupus-2022-elm2022.74

Catastrophic anti-phospholipid syndrome (CAPS) is a rare (1%) severe, and accelerated form of antiphospholipids syndrome (APS) characterized by the almost simultaneous occurrence of microcirculatory thrombosis in multiple locations leading to multi-organ failure involving compromising vital prognosis (40% of deaths). Among the risky situations pregnancy has been reported by many authors who include the HELLP syndrome.

Observation 34 years old woman , P5P5A0 with a history of deep vein thrombosis of the lower limbs 9 months earliers present during a pregnancy at 25th week of amenorrhea successive hepatic, coronary, cerebral thrombotic events and pulmonary embolism. A HELLP syndrome imposes the extraction of a fetus at the 30th week of pregnacy. The diagnosis of APS was made on the history of DVT and the clinical context (coronary, neurological damage) and will be confirmed by the presence of anti-cardiolipin antibodies (ac CL) type IgG at 30UI. Speckled-type antinuclear antibodies and anti-SSA antibodies were positive at 1/320, beta2 glycoprotein antibodies and IGM-type anti-cardiolipin antibodies were positive at 36UI.The abdomino-pelvic ultrasound visualized a liver increased in size to 20mm with heterogeneous regular contours and a spleen increased in size to 164mm in regular contour with the Doppler finding a portal trunk and dilated but permeable superhepatic veins. Cardiac echo-doppler found an EF at 40%, a dilated left ventricle and kinetic disorders. Coronary angiography found healthy coronaries. In front of the table associating a successive attack of 3 organs (liver, heart, brain) in one week of interval associated with the presence of anti-phospholipid antibodies allowed to make the diagnosis of SCAPL. The patiente was then put on Lovenox at a curative dose and double anti-platelet aggregation (Aspégic 100 mg and Plavix 75 mg/d) associated with an enzyme-converting enzyme inhibitor and a cardio-selective beta-blocker. Similarly, a bolus of corticosteroid 1 g/d was initiated for 3 days with an oral relay of prednisone equivalent at 1 mg/d with progressive degression. The relay of heparin by Sintrom and the addition of plaquenil at 200 mg/d consolidated the treatment. Tubal ligation was discussed. The evolution was favorable on the biological and morphological clinical level. Cardiac reassessment and abdominal CT angiography were scheduled.

During APS, the obstetrical pathology is often in the foreground and frequently allows the diagnosis to be evoked. Patients with APS remain at high risk of developing obstetric complications involving the maternal and fetal prognosis. The successive thrombotic occurrence in the peripartum announces a SCAPL, darkens the prognosis and requires intensive multidisciplinary emergency management. Nevertheless, the prognosis of APS has improved in recent years, allowing patients to access maternity in the best possible safety conditions.

PO.2.45 AUTOANTIBODIES ASSOCIATED WITH SYSTEMIC SCLEROSIS (SSC) IN THREE DISEASES CHARACTERIZED BY TYPE I INTERFERONS: A COMPARISON BETWEEN SLE, PRIMARY SJÖGREN'S SYNDROME, SSC AND HEALTHY BLOOD DONORS

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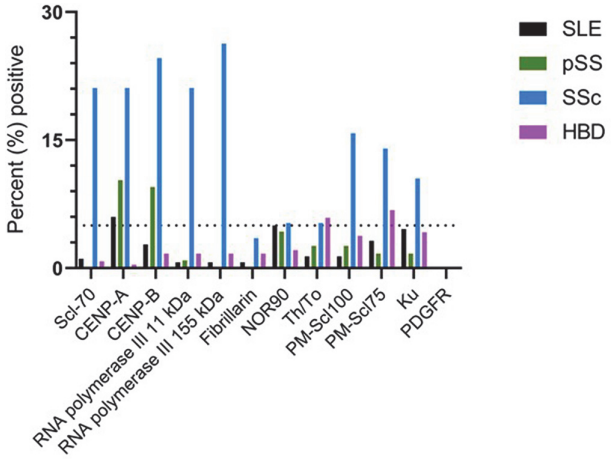
10.1136/lupus-2022-elm2022.75

Purpose SLE, primary Sjögren's syndrome (pSS) and systemic sclerosis (SSc) are heterogeneous autoimmune diseases with a dysregulated type I interferon (IFN) system. The diseases primarily affect females and often show overlapping clinical findings, e.g., arthritis and Raynaud's phenomenon (RP), which may result in diagnostic challenges. The Systemic Sclerosis Profile is an immunoblot test, including different autoantibodies (aab), which has been launched to aid the identification of patients with recent-onset SSc and to stratify patients into more homogenous subsets. Herein, we investigated whether SSc-associated aab also could be found in patients with well-characterized SLE and pSS, and whether these aab correlate to clinical phenotypes. In addition, we employed samples with clinical data from patients with SSc as well as from healthy blood donors (HBD).

Methods Serum samples from SLE (n=282), pSS (n=116), SSc (n=57) and HBD (n=236) were analyzed using a commercially available immunoassay (Systemic Sclerosis Profile Euro-Line [IgG]; Euroimmun AG, Lübeck, Germany). IFN-alpha was quantified in the sera using ELISA (Mabtech AB, Nacka,



Abstract PO.2.45 Figure 1 Presence of 12 autoantibody specificities (anti-Ro52/SSA not shown) among the patients with SLE, pSS and SSc as well as in HBD. The dotted line depicts an over all 5% positivity. A cut-off based on 5% positivity among HBD is frequently used for immunoassays

Sweden). Data on clinical manifestations of patients as well as self-reported information on RP of HBD were available.

Results Significantly higher proportion of subjects with SSc than SLE, pSS and HBD had aab against Scl-70, CENP-A, CENP-B, RNA polymerase-III 11kDa, RNA polymerase-III 155kDa, PM-Scl100, PM-Scl75 and Ku (Figure 1). No significant difference in prevalence was seen regarding aab against fibrillarin, NOR90, Th/To or PDGFR. Anti-Ro52/SSA was found in a higher proportion of patients with pSS (80.2%) and SLE (41.1%) than in SSc (22.8%). Among patients with SLE, anti-NOR90 was associated with discoid lupus (p=0.025) whereas anti-CENP-A was inversely associated with hematological disorder (p=0.005) and photosensitivity (p=0.024). Anti-CENP-A was inversely associated with mucocutaneous (ACR criteria 1–4, merged) involvement (p=0.014). Anti-CENP-B was significantly associated with serositis (p=0.005), anti-Ku with lupus nephritis (p=0.007) and anti-Ro52/SSA with neurological disorder (p=0.021). Among patients with SSc, 50/57 (87.7%) tested positive for ≥1 specificity and 6 of the 7 (85.7%) immunoblot negative patients were ANA positive with immunofluorescence microscopy. Pulmonary fibrosis was associated with anti-Scl-70 (p=0.008), anti-RNA polymerase-III 11kDa (p=0.003) and anti-RNA polymerase-III 155kDa (p=0.003). Pulmonary arterial hypertension was associated with anti-CENP-A (p=0.05). Among HBD, aab against RNA polymerase-III 155kDa (p=0.027) and PM-Scl100 (p=0.018) were associated with RP.

Conclusions We demonstrate that the 13 aab included in the Systemic Sclerosis Profile immunoassay are commonly detected among patients with SSc, but they are also frequent among individuals with other diseases characterized by type I IFNs. This information is important to convey to clinicians, especially as patients may present with similar manifestations or overlap syndromes. In SLE, we observed associations between clinical manifestations and SSc-associated aab which have not previously been reported.

PO.2.46 CLINICAL AND IMMUNOLOGICAL CONSIDERATIONS OF CERULOPLASMIN ACTIVITY SHIFTS IN SYSTEMIC LUPUS ERYTHEMATOSUS

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10.1136/lupus-2022-elm2022.76

Background Ceruloplasmin is an oxidoreductase considered to be a key intracellular antioxidant factor that substantially contributes in SLE pathogenesis. There is disequilibrium between ROS production and disposal in SLE shifted to accumulation of toxic products of the free radical reactions. The origin of such imbalance still remains unclear. Production of autoantibodies to ceruloplasmin and other antioxidant enzymes could be putative source of their dysfunction.

Purpose Assessment of interrelations between oxidase activity of ceruloplasmin, anti-ceruloplasmin antibodies presence and basic clinical and laboratory parameters of SLE with the use of ceruloplasmin immobilized on the magnetic beads.

Methods The research was conducted according to the WMA Helsinki Declaration after approval of the local ethics committee. 63 SLE patients were included in the research when admitted to rheumatology unit of Volgograd Municipal Hospital of Emergency Care #25. The diagnosis was verified using EULAR/ACR classification criteria (2019). Serum samples were collected after inclusion as well as before the discharge. Disease activity was assessed by means of ECLAM index. 30 healthy volunteers were included in the reference group. Anti-ceruloplasmin antibodies were detected by ELISA using ceruloplasmin coated magnetic polyacrylamide beads that have been made by original technology. Oxidase ceruloplasmin activity was measured using paraphenylenediamine oxidation technique. The values were expressed as optical density units (ODU). Results were expressed as means ± standard deviations.

Results Mean ECLAM index in SLE group was 10.2±6.7 points. Mean anti-ceruloplasmin antibody value in the reference group was 0.020±0.014 ODU. None of individual values was outside reference interval (M+3SD) meaning that there were no antibody positive cases in the reference group. 30 (47.6%) SLE patients were demonstrated serum anti-ceruloplasmin antibodies, and its mean concentration was 0.134 ±0.026 ODU. Ceruloplasmin activity in anti-ceruloplasmin antibody positive SLE patients was significantly lower than in antibody negative patients (1.234±0.315 ODU and 1.822 ±0.154 ODU, respectively, p<0.001). SLE activity positively correlated with anti-ceruloplasmin antibody concentration in contrast to its negative correlation with ceruloplasmin activity. Moderate but significant increase of ceruloplasmin activity and decrease of the antibody concentration were found after in-hospital treatment.

Conclusion Anti-ceruloplasmin antibodies, which can be found in a large proportion of SLE patients, are eventual inhibitors of ceruloplasmin oxidase activity, thereby leading to insufficiency of the extracellular portion of the antioxidant system. The study of antibody formation to ceruloplasmin, as well as its enzymatic activity, expands the existing understanding of SLE pathogenesis and outlines ways for further research in the field of immunological considerations of antioxidant system.

PO.2.47 HOW SMALL DETAILS AFFECT THE SENSITIVITY OF DIFFERENT SLE CRITERIA? INVESTIGATING A LARGE NUMBER OF PATIENTS IN A HUNGARIAN CENTER

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10.1136/lupus-2022-elm2022.77

Introduction The new 2019 EULAR/ACR classification criteria includes compulsory ANA positivity as an entry criterion and contains 7 clinical and 3 immunologic domains. Each domain involves different clinical signs followed by weighted score from 2 to 10.

Aim To assess the performance of the 2019 European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) classification criteria for SLE patients against the ACR 1997 and the SLICC 2012 criteria.

Method We performed a retrospective observational study between 2018.01.01. and 2021.04.27. from 382 patients with lupus. The diagnosis of SLE was established by the rheumatologist in rutine care and these diagnosis rates were compared against those that were determined based on the three classification criteria to identifying the sensitivities.

Results Among the patients the ACR 1997 sensitivity was 81% (310 patients) and the SLICC 2012 criteria achieved 95% sensitivity (361 patients). The 2019 EULAR/ACR