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THU0261

NEW 2019 SLE EULAR/ACR CLASSIFICATION CRITERIA ARE VALID FOR IDENTIFYING SLE AMONG PATIENTS ADMITTED FOR PERICARDIAL EFFUSION

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Background: The new 2019 SLE European League Against Rheumatism/American College of Rheumatology (EULAR/ACR) classification criteria for systemic lupus erythematosus (SLE) have been recently published. Seritis is a prominent -often inaugural- feature of active SLE. Low titers of antinuclear antibodies (ANA) have been frequently reported in patients with idiopathic pericarditis. Of note, ANA positivity at a titer $\geq 1/80$ is now mandatory as an entry criterion in the 2019 SLE EULAR/ACR classification criteria.

Objectives: Although classification criteria have theoretically no individual diagnostic purpose, we aimed at testing this new criteria set in unselected patients with pericardial effusion

Methods: In a retrospective study performed in the Department of Internal Medicine, University Paris Diderot, a French competence centre for rare systemic autoimmune diseases (AID), all consecutive adult patients hospitalized from January 2009 to January 2019 for pericardial effusion were reviewed. Clinical and biological data collected at time of the diagnosis of pericardial effusion were analyzed. The characteristics of the patients are listed in Table 1. Three sets of lupus criteria (SLE ACR-1997, SLE SLICC and 2019 SLE EULAR/ACR criteria) were applied in all ANA-positive patients

Results: Over a 10-year period, 137 patients were admitted for pericardial effusion. Search for ANA was systematically performed at diagnosis in all but 8 ($n=129$) and measured at a titer $\geq 1:80$ on Hep-2 cells in 49 patients (38%) that were eventually separated in three groups: 17 (34.7%) patients with a final diagnosis of SLE based on senior clinician judgement, 6 (12.2%) patients with a final diagnosis of autoimmune disease (AID) other than SLE (primary Sjögren's syndrome ($n=2$), undifferentiated connective-tissue disease ($n=2$) and systemic sclerosis ($n=2$)) and 26 (53.1%) patients with a diagnosis of idiopathic pericarditis after exclusion of malignancy, tuberculosis and systemic inflammatory diseases with a median 12.3 [1.6-29.8] months follow-up

The 2019 SLE EULAR/ACR criteria were met in 100% of patients with SLE, 33.3% of patients with non-SLE AID and 11.5% of patients with idiopathic pericarditis. Thus this new set of criteria for SLE offered a higher sensitivity (100%) but a lower specificity (84.38%) as compared to the former criteria, for the diagnosis of SLE in patients with pericardial effusion. Interestingly, the 2019 SLE EULAR/ACR classification score was higher in SLE patients (median: 30 [11-45]) as compared to non-SLE AID (median: 8 [6-12], $p=0.0006$) and idiopathic pericarditis patients (median: 6 [5-12], $p<0.00001$). Moreover, the 2019 classification set score strongly correlated with the SLEDAI activity score [6] as shown Figure S1 ($R^2=0.8105$, $p<0.00001$). Setting the 2019 SLE EULAR/ACR classification threshold score >12 (out of a theoretical maximum of 51) instead of ≥ 10 increased the specificity of 2019 SLE EULAR/ACR criteria from 84.38% to 100%. Overall, in patients with pericardial effusion and positive ANA, the diagnosis of SLE could be ruled out when 2019 SLE EULAR/ACR criteria score was < 10 and confirmed when the score was > 12 .

Conclusion: This study shows that the new 2019 SLE EULAR/ACR criteria for SLE are helpful in clinical practice for the diagnosis of SLE in patients admitted for pericardial effusion

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THU0262

LEVELS OF OSTEOCALCIN AND PROCOLLAGEN TYPE I C-TERMINAL PROPEPTIDE IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS, THEIR ASSOCIATION WITH BONE MINERAL DENSITY AND LEVEL OF INTERLEUKIN-6

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Background: Osteoporosis and fractures associated with it are considered to be one of the most severe complications of systemic lupus erythematosus (SLE). The role of a systemic inflammatory process, vitamin D deficiency, hypogonadism and peculiarities of disease treatment in reduced bone mineral density (BMD) is being discussed. Even though the frequency of osteoporosis in patients with SLE is being studied extensively by scientists from different countries, data on the peculiarities of bone tissue metabolism and the factors that provoke disorders of bone remodeling in such individuals are quite limited. The association between markers of bone tissue metabolism and BMD, and how they change during an inflammatory process is poorly studied.

Objectives: The objective of our research is to study the levels of osteocalcin (OC) and procollagen type I C-terminal propeptide (PICP) in patients with systemic lupus erythematosus and to estimate their association with BMD and inflammatory activity based on the levels of interleukin-6 (IL-6).

Methods: A total of 58 women with SLE (the average age was 45.11 ± 1.03 years old) and 29 individuals from the control group (the average age was 46.79 ± 2.30 years old) were examined. The diagnosis of SLE was established on the basis of 2019 EULAR/ACR classification criteria for SLE. Levels of IL-6, OC and PICP in serum were determined by enzyme immunoassay. Changes in BMD of the lumbar spine at the level of L1-L4 and the proximal femur were determined by dual-energy X-ray absorptiometry. In postmenopausal women, the diagnosis of osteoporosis was established by the T-score ≤ -2.5 SD. Osteopenia met T-score from -1 to -2.5 SD. In women of reproductive age, the Z-score was used to determine BMD. Values of the Z-score ≤ -2.0 SD were considered as "below expected range for age".

Results: The average OC level in serum of practically healthy individuals equaled 17.64 ± 0.59 ng/ml, and in patients with SLE it was 13.96 ± 0.40 ng/ml, i.e. it was 20.9% lower. The average PICP level in the control group equaled 107.8 ± 4.28 ng/ml, in the main group it was 92.9 ± 5.01 ng/ml, i.e. 16% lower. Overall, the decrease in the bone turnover markers (PICP and/or OC) was noticed in 28 patients with SLE (48.3%) and only in 4 practically healthy individuals (13.8%).

In women with decreased bone turnover markers, the T-score of the lumbar spine and hip was 2.3-2.6 times lower ($p < 0.05$) than in the group with adequate levels of bone turnover markers. Z-score was also lower among patients with decreased levels of OC and PICP. In this group, the average BMD level was 0.81 ± 0.05 g/cm² and was 13.8% lower than in the group of patients with no signs of bone tissue metabolism disorder - 0.94 ± 0.02 g/cm². Among the group of women with signs of suppression of biosynthetic processes in bone tissue, there were twice more individuals with decreased BMD. In patients with critically high levels of IL-6 (above 20.0 ng/L), OC level was lower than in patients with high (12.5-20.0 ng/L) and adequate (< 12.5 ng/L) levels of IL-6 (by 17.3 and 19% respectively). The proportion of individuals with low OC levels increased from 31.2% in the last group to 70.6% among patients with critically high levels of IL-6.

PICP level was also lower (38.1% and 39.7% respectively) in case of critically high IL-6 levels compared to its high and adequate levels. The proportion of individuals with low PICP levels increased from 6.3% in the group with adequate IL-6 level to 58.8% in the group with critically high IL-6 level.

Conclusion: Women with SLE have bone tissue metabolism disorder in the form of decreased bone turnover markers (procollagen type I C-terminal propeptide and osteocalcin) associated with the inflammatory activity. In the group of patients with the signs of suppression of biosynthetic processes in the bone tissue, there were more individuals with decreased BMD.

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THU0263

LUNG INVOLVEMENT IN PRIMARY SJÖGREN SYNDROME – AN UNDER-DIAGNOSED ENTITY

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Background: Interstitial lung disease (ILD) represents a frequent extra-glandular manifestation of primary Sjögren's Syndrome (pSS). Limited published data regarding phenotyping and treatment exists. Advances in managing specific ILD phenotypes have not been comprehensively explored in patients with coexisting pSS.

Objectives: This retrospective study aimed to phenotype lung diseases occurring in a well-described pSS cohort and describe treatment course and outcomes.

Methods: Between April 2018 and September 2019, all pSS patients attending our Outpatient clinic were screened for possible lung involvement. Clinical, laboratory and computer tomography (CT) findings were analysed. Patients were classified according to CT findings into 5 groups: usual interstitial pneumonia

(UIP), non-specific interstitial pneumonia (NSIP), desquamative interstitial pneumonia (DIP), combined pulmonary fibrosis and emphysema (CPFE) and unspecific-ILD.

Results: Lung involvement was confirmed in 24/240 patients (10%). Clinically manifest pSS occurred later in patients with ILD vs. non-ILD (53.2 [42.0-61.7] vs. 62.3 [55.6-68.8] years; $p=0.0016$). The commonest phenotype was UIP $n=10$ (41%), followed by NSIP $n=7$ (29%), DIP $n=2$ (8%), CPFE $n=2$ (8%) and unspecific-ILD $n=3$ (13%). Forced vital capacity (FVC) and carbon monoxide diffusion capacity (D_{LCO}) appeared lower in UIP and DIP, without reaching a significant difference. Treatment focused universally on intensified immunosuppression, with 12/24 patients (50%) receiving cyclophosphamide. No anti-fibrotic treatments were used. Follow-up was median 13.2 [7.9-72.3] months, during which 6/24 (25%) patients exhibited a further decline in FVC. No deaths occurred.

Conclusion: Lung involvement in pSS is heterogeneous. Better phenotyping and tailored treatment may improve outcomes and requires further evaluation in larger prospective studies.

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THU0264

MYELOID MALIGNANCIES, SYSTEMIC AUTOIMMUNE DISEASES AND CARDIOVASCULAR RISK: AN UNDER-REPORTED ASSOCIATION?

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Background: The association between systemic autoimmune diseases (ADs) and lymphoproliferative malignancies is well established; nonetheless, few studies have investigated the prevalence and prognostic impact of myeloid malignancies on systemic autoimmune conditions.

Objectives: To investigate the frequency of myeloid malignancies (i.e. myelodysplastic syndrome (MDS) and chronic, either Philadelphia-positive or Philadelphia-negative, myeloproliferative disorders (MPNs)) in patients with ADs and their influence on the ADs clinical course and vice-versa.

Methods: A retrospective systematic search through the electronic health records of the patients admitted at our Rheumatology University Hospital from 2009 and 2019 was performed to select those presenting with ADs and MDS or MPNs. To refine the search the ICD-9-CM diagnosis codes for MDS/MPNs were utilized. Medical charts of eligible patients were retrieved and data were collected with regard to demographics, type of AD, AD duration, prior treatments, serum laboratory indices, bone marrow aspiration and biopsy data. Categorical variables were compared using chi square test and Fisher's test; continuous variables were compared using Student's t-test. A 2-tailed value of $p < 0.05$ was taken to indicate statistical significance.

Results: Out of the medical records of 5040 patients, we identified 51 patients (31 F: 20 M, mean age: 61 years (15)) with AD and myeloid malignancies: 17/51 with AD and MDS and 34/51 with AD and MPNs. No demographic differences were observed in the two subgroups. Regarding MDS, anaemia was the most common haematologic presenting finding (15/17, 88%), while the most common diagnosis was refractory anemia with excess of blasts (RAEB I/II) (5/17, 29%) followed by sideroblastic anemia (2/17, 12%). In the MPNs subgroup, 12/34 patients (35%) had a diagnosis of chronic myeloid leukemia (CML), 9/34 (26%) had a myelofibrosis (MF), 7/34 (21%) had an essential thrombocythemia (ET) and 6/34 (18%) had a polycythemia vera (PV). The JAK2 V617F mutation was detected in 100%, 57%, and 66% of PV, ET, and MF patients. Regarding the temporal appearance of myeloid malignancy, MDS occurred concurrently (9/17) or followed (7/17) the diagnosis of ADs in the vast majority of the cases whereas MPNs generally preceded the diagnosis of ADs (19/34). In MDS the most commonly diagnosed ADs were seronegative arthritis (5/17, 29%), large and small vessel vasculitis (4/17, 23%) and Systemic Lupus Erythematosus (3/17, 17%). In patients with MPNs the diagnosis of rheumatoid arthritis (2/9, 22%), and antiphospholipid syndrome (3/9, 33%) were often associated with MF, whereas anti-Ro52 (TRIM21) positive systemic connective tissue disorders (4/7, 57%) were more frequently detected in ET. Cardiovascular events were observed in 14/51 (27%): 4/17 (23%) in MDS, 3/12 (25%) in CML and 7/22 (32%) in Philadelphia-negative MPNs. The latter seven cardiovascular events were all observed in patients presenting JAK2 V617F mutation ($p=0.05$).

Conclusion: Our study is limited by its retrospective design. However, our results documented that the frequency of MDS and MPNs in ADs is not negligible and might be considered in the assessment of cardiovascular risk in systemic autoimmunity. Moreover, it has been reported that, under viral infection, TRIM21 is up-regulated by activation of the IFN/JAK/STAT pathway; interestingly, anti-Ro52 (TRIM21) were over-represented in MPN, where the JAK/STAT signal is hyper activated. This could explain also our observation that frequently the onset of ADs follows the diagnosis of MPN.

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THU0265

THYMIC STROMAL LYMPHPOIETIN (TSLP) AS A BIOMARKER OF PRIMARY SJÖGREN'S SYNDROME (PSS) AND RELATED LYMPHOMA: VALIDATION IN INDEPENDENT COHORTS

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Background: Thymic stromal lymphopoietin (TSLP) has been implicated in primary Sjögren's syndrome (pSS) and related B-cell lymphoproliferation/lymphoma (NHL) by tissue studies on salivary glands (SG) (1). It resulted significantly higher in the serum of pSS patients compared to non-pSS sicca and to healthy subjects, with the highest levels found in NHL.

Objectives: The purpose of this work was to confirm that serum TSLP is elevated in pSS by the study of independent cohorts.

Methods: Serum TSLP levels were measured by ELISA in 91 pSS patients ($F=86$, 94.5%; mean age 57.2 years, 25-80) from the Udine cohort (cohort 1, UD), Italy. One additional multicentre cohort (cohort 2) from the Italian SS Study Group (GRIS) was studied, including 125 pSS patients from the Universities of Roma (RO), L'Aquila (LAQ), Pisa (PI) and Perugia (PG). pSS patients with active NHL ($n=12$ in cohort 1; $n=1$ in cohort 2) were excluded from comparative analyses to avoid bias. Secondly, additional serum samples from pSS-related NHL in stable and complete remission, from both cohort 1 and 2, were analysed in a separate subgroup ($n=12$). Thirdly, a preliminary evaluation of serum TSLP was performed in pSS patients from a different geographical area (University of Athens, Greece; cohort 3).

Results: Cohort 2 included 125 pSS patients ($F=114$, 91.2%; mean age 58.1 years, 23-84): 124 benign, 1 with NHL. In this cohort, serum TSLP levels were confirmed to be high (mean 30.26 pg/mL, 0.41-95.21) and comparable to cohort 1 (mean 33.81 pg/mL, 0-140.8; $p=ns$). No difference was found by the separate analysis of pSS from each single Centres (RO $n=49$, mean 33.21, 1.4-95.21; LAQ $n=34$, mean 38.6, 16.31-85.11; PI $n=28$, mean 20.23, 0.41-56.67; PG $n=13$, mean 19.39, 1.03-68.38; $p=ns$), and vs cohort 1 ($p=ns$). The only patient in cohort 2 with NHL showed serum TSLP of 160.91 pg/mL, comparable to the mean TSLP in the 12 UD pSS-NHL (151.96 pg/mL). Importantly, in pSS-related NHL in stable remission, serum TSLP resulted undetectable (7/13) or detectable at very low levels (6/13) (mean 10.46, 0-38.5), and significantly lower than in benign pSS patients from the two cohorts ($n=203$, mean 31.48, 0-140.8; $p=0.0022$). Metachronous samples from one patient, at the stage of NHL activity and then at NHL remission, showed a decrease in TSLP from 128.04 pg/mL to undetectable levels. Finally, TSLP levels were increased also in the Greek cohort (mean 54.9, 26.72-78.95), and significantly higher than the two Italian cohorts ($p=0.0085$ and $p<0.0001$, vs cohort 1 and 2, respectively).

Conclusion: Serum TSLP levels are increased in pSS, as herein confirmed in independent cohorts. TSLP might be important in the disease pathophysiology and mirrors the course of pSS-related B-cell lymphoproliferation itself. It may thus represent a novel important biomarker.

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[1] Gandolfo S. et al, Clin Exp Rheumatol. 2019 May-Jun;37 Suppl 118(3):55-64.

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