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THU0274

QUALITATIVE AND QUANTITATIVE ANALYSIS OF THE IMMUNOLOGIC CHARACTERISTICS OF THE MINOR SALIVARY GLAND BIOPSY IN SJÖGREN'S SYNDROME

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Background: Minor salivary gland biopsy (MSGB) is the most important diagnostic test of Sjögren's Syndrome (SS). It demonstrates the presence of the inflammatory infiltration in the most affected site. It's possible role as a biomarker in the disease is still unknown. The Immunology Department of our center conducts a detailed analysis of the MSGB about the leukocyte infiltration and quantifies number of each cell.

Objectives: To describe the immunologic features of the MSGB and carry out an association analysis with clinical variables

Methods: Clinical variables, ESSDAI index at the moment of diagnosis and laboratory parameters were recorded. As from the MSGB, number of infiltration focus (1, 2 or several), big infiltrations (>100 cells), number of B and T cells, CD4/CD8 ratio and presence of isolated lymphocyte were collected. Categorical variables were described as frequencies and analysed using Fisher exact test. T student and Wilcoxon Rank Sum Test were used for comparison of means (μ).

Results: In 2017, a total of 104 MSGB were carried out in our center. Among them 58 were diagnosed as SS by medical and ACR/EULAR 2016 criteria. Finally 41 patients with SS and abnormal MSGB result were included for this study.

Basal characteristics of patients	Frequency (number/percentage)
Men	8/19,5%
Extraglandular disease	18/43,9%
ESSDAI $\geq$ 2	17/41,46%
Other autoimmune diseases	2/4,87%
Ac. Ro/La	0/0
CRP	13/31,71% (μ 9,79mg/dL)
ANA pattern	8/19,51%
Negative	1/2,44%
Homogenous	16/39,02%
Speckled	10/24,39%
Speckled and Homogeneous	6/14,64%
Other	

Biopsy: Patients with active disease (ESSDAI $\geq$ 2) had greater amount of cells (μ 159 cells vs 509 cells; $p=0,055$) as well as those with extraglandular disease (μ 160 vs 488; $p=0,08$). Patients with active disease also had larger number of infiltration focus ($p=0,062$). The presence of isolated CD8+ T cells was observed in 13 patients and they had lesser cells (μ 136 vs 381; $p=0,35$). In those samples with predominance of T cells over B cells had larger number of infiltrate focus (7/20; 35% vs 12/21; 57.14%; $p=0,155$). No association with disease activity or extraglandular manifestation was found.

Extraglandular manifestation and disease activity: 18 patients had extraglandular disease. Moderate or severe ESSDAI activity was found in 14 of these patients (34.2%, $p=0,00$). The biopsy of patients with extraglandular disease had larger amount of cells (μ 200 vs 145; $p=0,01$). Patients with active disease had more infiltrate focus (6/22 vs 11/19; $p=0,047$). Disease evolution time was similar with a mean duration of 8-9 years in both groups.

Corticosteroids: There were 3 patients with active steroid treatment ($\geq$ prednisone 10mg/d) at the moment of the biopsy. All 3 of them had >1 focus in the sample and 2 of them had large infiltrate with >150 cells. Eight of them had received steroids in the last 5 years, 6 of them had large infiltrate with >150 cells and 4 had >1 infiltrate focus in the biopsy. A study with more sample should be carried out to study the influence of steroids in the biopsy results.

Conclusion:

- Patients with extraglandular disease have larger amount of cells in the composition of infiltration. Those with more disease activity had more number of infiltration focus.

- In 14 patients specific antibodies and antinuclear antibodies were negative. In these patients the biopsy is the most useful diagnostic test.
- Possible association of those variables that were statistically not significant should not be ruled out due to the small sample size of the study.

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THU0275

NEW PROSPECTIVE OF COGNITIVE IMPAIRMENT IN SYSTEMIC LUPUS ERYTHEMATOSUS PATIENTS: A PRAGMATIC LANGUAGE EVALUATION

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Background: Cognitive impairment (CI) in Systemic Lupus Erythematosus (SLE) is a frequent neuropsychiatric manifestation affecting several domains, even in apparently asymptomatic patients. Current research revealed that the typical CI pattern affects frontal-subcortical circuit and thus executive functions. The impairment of non-literal language or Pragmatic Language (PL), including metaphors, idioms, inferences or irony has been well described in several conditions such as autism disorders, Parkinson's disease, brain injury and even in earlier phases of neurodegenerative processes. Even if PL neuro-anatomy remains controversial, correlation between executive dysfunctions and non-literal language involvement has been reported both in traumatic injury and mild cognitive impairment patients. Nonetheless, no specific study has been performed to evaluate PL impairment in SLE patients so far.

Objectives: We aimed at assessing the PL domain in a monocentric SLE cohort in comparison to healthy controls, matched to age and education, through a specific battery, BLED [1]. Secondly, we focused attention on possible correlations between CI and clinical and laboratory SLE-related features.

Methods: Forty adult patients affected by SLE, according to the ACR criteria, and thirty healthy subjects were enrolled consecutively in this cross-sectional study. The protocol included complete physical examination, extensive clinical and laboratory data collection (comprehensive of demographics, past medical history, co-morbidities, disease activity, chronic damage evaluation, previous and concomitant treatments) and cognitive assessment for five different domains: memory, attention, pragmatic language, executive and visuospatial functions. Self-reported scale for anxiety and depression were performed to exclude the influence of mood disorders on cognitive dysfunction.

Results: We enrolled forty Caucasian SLE patients (M/F 3/37; mean $\pm$ SD age 45.9 $\pm$ 10.1 years, mean $\pm$ SD disease duration 120.8 $\pm$ 81.2 months) and thirty healthy subjects (M/F 9/21; mean $\pm$ SD age 41.3 $\pm$ 13 years). According to the low level of disease activity and damage (mean $\pm$ SD SLEDAI-2K of 1.3 $\pm$ 2.3, mean $\pm$ SD SDI of 0.2 $\pm$ 0.5), only 30% of patients was on glucocorticoid treatment at the study entry. PL was the most compromised domain in terms of Mean Domain Z scores (Fig. 1). As regards the Domain Cognitive Dysfunction score, a deficit of PL was observed in 45% of patients and resulted significantly more prevalent than memory, executive and visuospatial functions impairment ($P=0,0002$, $P=0,0002$ and $P<0,000001$, respectively). According to Global Cognitive Dysfunction score 25% of patients experienced a mild impairment and 7.5% a moderate one. Anti-phospholipid antibodies positivity was significantly associated with memory impairment ($P<0,0005$), whereas the presence of other neuropsychiatric events was associated with executive dysfunctions ($P<0,05$); neither further significant association nor correlation were identified.

Abstract THU0275 – Figure 1. Distribution of neurocognitive impairment expressed in Mean Domain Z score.

Conclusion: We evaluated for the first time PL in a single center SLE cohort finding a dysfunction in almost half of patients enrolled, significantly more frequent than the other assessed domains.

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Disclosure of Interests: Carmelo Pirone: None declared, Fulvia Ceccarelli: None declared, Concetta Mina: None declared, Alfredo Mascolo: None declared, Carlo Perricone Speakers bureau: BMS; Lilly, Celgene, Sanofi, Barbara Mazzotta: None declared, Laura Massaro: None declared, francesca spinelli: None declared, cristiano alessandri: None declared, Guido Valesini: None declared, fabrizio conti: None declared

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THU0276

CHARACTERISTICS OF NEUROLOGIC INVOLVEMENT AND ITS RELATED FACTORS IN PRIMARY SJÖGREN SYNDROME

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Background: Neurological manifestations seem common in primary Sjögren's syndrome (pSS) but their reported prevalences vary in Chinese. And few studies reveal if the disease activity is associated with neurological involvement.

Objectives: To analyze the clinical neurological manifestations of primary Sjögren syndrome(pSS), and to evaluate the relationship with disease activity.

Methods: 112 patients(7 male, 105 female) who fulfilled the 2002 American-European Consensus Group criteria for pSS were enrolled in the study. For each patient, the clinical features were evaluated by medical data including clinical, laboratory and immunologic data, and neurological examinations including electromyography, magnetic resonance imaging, cerebrospinal fluid, and electroencephalogram. Statistical methods used were t-test, chi-square test and Logistic regression.

Results: Data at inclusion were available for 112 patients, whose mean age was 55±10 years. Neurological involvement was noted in 19.6(22/112) patients, including 17(15.2%) with peripheral nervous system(PNS) manifestations, 3(2.8%) with central nervous system (CNS) manifestations and 2(1.8%) with both PNS and CNS involvements. Optic neuritis and trigeminal neuralgia were revealed frequently in cranial neuropathy. Anti-aquaporin 4 antibody was detected in two patients with optic neuritis. The clinical spectrum of peripheral neuropathies encountered in Sjögren's syndrome patients was wide with sensory neuropathies being the most common. Tibial nerve, peroneal nerve and sural nerve were the most likely involved and lower limb involvement accounted for 68.4%(13/19). The frequency of Raynaud's phenomenon was significantly higher[31.8%(7/22) vs 4.4%(4/90), $P<0.01$] as well as acroanesthesia[72.7% vs 8.9%, $P<0.01$] in pSS with neurological involvement than in pSS without neuropathy. The median values of EULAR Sjögren's syndrome disease activity index(ESSDAI) were 5.6(range 2.6-7.6) and 3.2(range 1.4-5.2) in the NS and non-NS groups respectively($P<0.01$). We found a significant rise of neuropathy risk associated with Raynaud's phenomenon(relative risk 9.365, 95%CI 3.191 40.093, $P=0.003$) and ESSDAI(relative risk 1.628, 95%CI 1.169 1.969, $P=0.001$). Elevated titers of rheumatoid factor($P<0.05$) and ANA($P<0.01$) were common in patients with neuropathy.

Conclusion: Neuropathy is not a rare manifestation of pSS. Prevalence of neurological involvement in pSS is 19.6%. Raynauds phenomenon and high disease activity may be the risk factors for neuropathy. Autoantibodies might contribute to the injury of the nervous system.

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THU0277

HOW THE AGE AT DIAGNOSIS MODIFIES THE PHENOTYPE OF PRIMARY SJÖGREN SYNDROME: ANALYSIS IN 11,420 PATIENTS (BIG DATA SJÖGREN PROJECT)

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Objectives: To analyse how the age at diagnosis modifies the phenotype of primary Sjögren syndrome (SS)

Methods: The Big Data Sjögren Project was formed in 2014 to take a "high-definition" picture of the main features of primary SS at diagnosis by merging international SS databases. By January 2019, the database included 11,420 patients from 24 countries of the 5 continents.

Results: Women (52.7 vs 54.6 yrs in men, $p<0.001$) and non-White patients (49.6 vs 53.5 yrs in Whites, $p<0.001$) were diagnosed at a younger age. Patients without sicca symptoms, with normal oral/ocular diagnostic tests and with positive biopsy were also diagnosed at younger ages ($p<0.001$ all comparisons).

Abstract THU0277 – Figure 1