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POS0122 CENTRAL NERVOUS SYSTEM DEMYELINATING SYNDROMES IN SYSTEMIC LUPUS ERYTHEMATOSUS: RESULTS FROM “ATTIKON” LUPUS COHORT

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Background: Central nervous system (CNS) demyelinating syndromes that occur in the context of SLE may represent a manifestation of neuropsychiatric lupus, or an overlap of SLE and multiple sclerosis (MS). The differential diagnosis between the two entities has important clinical implications, because the therapeutic management differs between the two conditions.

Objectives: To characterize CNS demyelinating syndromes in a large SLE cohort as neuropsychiatric SLE or SLE-MS overlap, using a multidisciplinary approach and existing criteria (MS international diagnostic criteria) and SLE criteria.

Methods: Patients from the ‘Attikon’ lupus cohort (n=707) were evaluated for demyelinating syndromes. Clinical, laboratory and neuroimaging data were recorded for each patient. Following multidisciplinary evaluation and application of criteria, the demyelinating syndrome was attributed to either SLE or MS. Patients with transverse myelitis were not included in this study.

Results: We identified 26 patients with demyelinating syndromes (3.7%) with mean age at diagnosis 46.9 (SD 12.3) years and median disease duration at last follow-up 60 (IQR 52) months. Of them, 12 were diagnosed as primary SLE-demyelination (46.2%) and 14 as overlap SLE-MS (53.8%). The two groups did not differ with respect to rheumatologic and neuropsychiatric manifestations, or serologic findings (ANA, dsDNA, C3/C4, aPL, EANA). SLE patients with demyelination manifested a lower degree of extra-CNS disease mainly involving joints and muscles, while severe non-CNS manifestations were rare. However, SLE patients with SLE-demyelination were less likely to have elevated IgG index (OR 0.055 95% CI: 0.008-0.40) and positive oligoclonal bands (OR 0.95 95% CI: 0.014-0.56). SLE patients with primary demyelinating syndrome were less likely to exhibit brain lesions in the spinal cord, infratentorial, periventricular and juxtaocular regions. A single brain region was affected in 8 SLE-demyelination patients (75%), while all MS-SLE overlap was associated with increased likelihood of neurologic relapses (OR 18.2, 95% CI: 1.76-188), while SLE-demyelination patients were less likely to exhibit neurological deficits (EDSS=0) at last follow-up visit (50% vs. 78.6%in SLE-MS, respectively).

Conclusion: Demyelination in the context of SLE follows a more benign course compared to a frank SLE-MS overlap. Prolongation of follow-up will ascertain whether SLE-demyelination patients evolve to MS, or this is a bona fide NPSLE syndrome.

Disclosure of Interests: None declared


POS0123 MAJOR SALIVARY GLAND ULTRASONOGRAPHY AND MRI WITH DIFFUSION WEIGHTED IMAGING (DWI) AS COMPLEMENTARY TOOLS TO IDENTIFY FEATURES OF MALT IN PRIMARY SJÖGREN’S SYNDROME (PSS): A SINGLE CENTER CROSS SECTIONAL STUDY

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Background: Salivary MALT lymphoma represents the major complication of primary Sjögren’s Syndrome (pSS). However, the early recognition of MALT lymphoma may be challenging due to its indolent, slow clinical course.

Objectives: 1. to identify salivary gland ultrasonographic (SGUS) features and magnetic resonance (MRI) abnormal findings with Diffusion Weighted Imaging (DWI) and Apparent Diffusion Coefficient (ADC) of MALT in pSS patients 2. To evaluate the correlation between salivary gland ultrasonography (SGUS) and MRI in pSS patients with suspected lymphoma.

Methods: consecutive patients with pSS (2016 ACR/EULAR criteria) and suspected MALT lymphoma undergoing histological examination were included in this cross sectional study from September 2017 to November 2021. The US echoroyst of each gland on B-mode images was graded using the latest OMERACT semiquantitative SGUS scoring systems (0-3). Sonographic features of focal lesions were described. Conventional MRI techniques (i.e T1WI, T2WI, and STIR images) combined with MR sialography was performed in all the cases. DWI was acquired at b-value 0, 500 and 1000. ADC values were calculated. Patients’ clinical and histological data were collected. Data were presented as means±SD, or percent frequency as appropriate. Intergroup comparisons were made using the t-test/Mann–Whitney test for continuous variables and Fisher’s exact test for categorical variable.

Results: 45 pSS (mean age, S.D=55.15 yrs) were included. MALT lymphoma was histologically confirmed in 14/45 pSS patients and, specifically in 18/180 major salivary glands (17 parotids and 1 submandibular gland). At SGUS examination, MALT salivary glands presented an OMERACT grade 3 in 16/18 and a grade 2 in 2/18, significantly higher than the OMERACT scoring observed in no-MALT pSS glands (p=0.001). The sonographic features more commonly detected in MALT were: hypoechoic microaureas with posterior enhancement, presence of septa or hyperechoic strands and anacral intralesional vascularization. At MRI, 15/18 (83.3%) MALT lymphoma appeared as intraglandular solid lesions: 9/15 (60%) were solid-cystic lesions and 6/15 (40%) were solid lesions without cystic changes. The frequency of solid lesions in pSS patients without lymphoma was 3/124 (2.4%), significantly lower than in MALT-pSS. Furthermore, 15/18 (83.3%) MALT lymphoma showed glandular fatty substitution. The presence of fatty substitution did not differ in MALT lymphoma and in no-MALT pSS glands. The mean (SD) ADC value of MALT lesions was significantly lower than the ADC of the parotid glands in pSS lymphoma (0.63±0.07 x 10^-3 mm2/s, p =0.001). A negative correlation between SGUS OMERACT score and mean glandular ADC values (r = – 0.776, p < 0.001) was found; patients with OMERACT score 3 in their SGUS group showed a careful screening for lymphoma.

Disclosure of Interests: None declared


Through the looking glass...