CNS 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 classification criteria (MS, SLE).

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, ENA). SLE patients with demyelination mainly involved the CNS, whereas extracranial disease mainly involving joints and skin was seen in severe non-CNS manifestations were rare. However, 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 0.09 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 9 SLE-demyelination patients (75%), while all MS-SLE patients had multiple affected brain regions. 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


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 u echosucture of each gland on B-mode images was graded using the latest 2019 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 variables.

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 score observed in no-MALT pSS glands (p=0.001). The sonographic features more commonly detected in MALT were: hypoechoic macroaeres with posterior enhancement, presence of septa or hyperechogenic strands and anisochronous intralobular 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 without lymphoma (0.63±0.07 x 10^-3 mm2/s vs 1.13± 0.19 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 presented the lowest mean salivary gland ADC when compared to the other OMERACT scores (0-2) (p<0.001).

Conclusion: OMERACT semiquantitative SGUS scoring systems and MRI with DWI represent promising complementary tools in the differential diagnosis of pSS MALT lymphoma, particularly useful to guide parotid biopsy. Patients with an OMERACT score 3 in their SGUS should receive a careful screening for lymphoma.

Disclosure of Interests: None declared


Through the looking glass...
Background: Hepatic involvement in AL amyloidosis is often asymptomatic but does affect prognosis and should be taken into account during follow-up. An increased plasma level of alkaline phosphatase (ALP) or increased liver span are part of the conventional diagnostic criteria for establishing hepatic involvement1, however these markers are nonspecific. 123I-labeled serum amyloid P component (SAP) scintigraphy is a specific and sensitive method to establish hepatic involvement but is not widely available. Liver stiffness measured by transient elastography is increased in AL amyloidosis patients with hepatic involvement and could be useful in establishing liver involvement and monitoring treatment response in AL-amyloidosis.

Methods: LS was measured prospectively in 49 treatment naïve patients with systemic AL amyloidosis and 9 patients with wild type transthyretin amyloidosis (ATTRwt) with cardiomypathy (cardiac controls). In addition, LS was longitudinally measured in 10 AL amyloidosis patients of whom 9 patients had liver involvement. SAP scintigraphy, laboratory assessments including ALP and measurement of liver span was performed in all patients.

Results: Of the 49 patients, 27 patients had liver involvement (of whom 24 also had heart involvement), 10 patients had heart involvement, 12 patients had no heart or liver involvement. Median LS was significantly higher in AL amyloidosis patients with liver involvement (22.8 kPa, range 4.3-75), than in AL amyloidosis patients without liver involvement (6.3 kPa, range 4.4-35.8) (p=0.000). Also a significant difference was seen between AL amyloidosis patients with liver involvement (22.8 kPa, range 4.3-75) versus, heart involvement (9.0, range 4.5-35.8) (p=0.02), no liver or heart involvement (5.7, range 4.4-10.1) (p=0.0001), and ATTRwt patients (2.9 kPa, range 4.0-14.6) (p=0.01). Furthermore, LS values seemed to significantly decrease over time in AL amyloidosis patients with liver involvement with a good hematologic response to treatment.

Conclusion: LS is a non-invasive tool which seems to be useful in clinical practice to establish liver involvement in patients with AL amyloidosis. In addition, it is a promising marker in the follow up of AL amyloidosis patients for establishing hematologic response over time.

REFERENCES:

Disclosure of Interests: None declared


Table 1. Abnormalities found on FDG-PET/CT scans

<table>
<thead>
<tr>
<th>No PET/CT result obtained</th>
<th>No PET/CT results found on any scan</th>
<th>One or more abnormalities found per scan*</th>
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<tbody>
<tr>
<td>3 (2.5)</td>
<td>59 (48.8)</td>
<td>59 (48.8)</td>
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* Fifteen of these abnormalities were found on the second PET/CT, the rest was found on the first scan. 11 abnormalities on the second PET/CT were the same as the one seen on the first scan, and 7 abnormalities resolved after the first scan. One scan can show multiple abnormalities, from different categories.

Follow-up action occurred in 21 (26.6%) patients, consisting of referral to a specialist or reassessing and/or scheduling diagnostics directly by the treating rheumatologist. In 5 (6.3%) patients, the rheumatologist followed-up. In 17 (21.5%) patients a consultation with a different specialist was scheduled. In five patients surgical/invasive intervention took place. In one patient a hemi-thyroidectomy was performed revealing a follicular adenoma. This resection was complicated by persistent recurrent laryngeal nerve paresis and hoarseness. In a second, an intra-uterine myometectomy took place. In a third, a colonoscopy was performed revealing two low-grade adenomas. In a fourth a benign cyst in the neck was extracted. A fifth patient underwent spinal marginal myotomy which turned out to be a benign schwannoma.

Nine patients (7.4%) were suspected of malignancy, none turned out to be malignant. Six clinical malignancies (bladder, penis, lymphoma, 2 melanoma and prostate) that developed during follow-up were all negative on baseline FDG-PET/CT. The malignancies were diagnosed after an interval of between 5 and 34 months (mean 13 months).

Conclusion: Whole-body FDG-PET/CT-scanning for arthritis imaging in RA patients results in frequent incidental extra-articular findings, while some who apparently had normal scans developed malignancies.

REFERENCES:

Disclosure of Interests: None declared


POSI025 EXTRA-ARTICULAR FINDINGS WITH FDG-PET/CT IN RHEUMATOID ARTHRITIS PATIENTS: MORE HARM THAN BENEFIT.

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Background: Whole-body Positron Emission Tomography with CT-scanning using fluorine-18 fluorodeoxyglucose (18F-FDG) is occasionally used in Rheumatoid Arthritis (RA) patients. Reasons to use FDG-PET/CT-scans are to diagnose arthritis or guide decisions on systemic therapy, as FDG uptake in affected joints may reflect disease activity [1]. FDG-PET/CT might also detect malignancies, but the frequency of incidental findings and the proportion of relevant malignant disease that could be missed are currently unknown.

Objectives: To study the malignancy screening performance of whole-body FDG-PET/CT in longstanding RA patients with low disease activity.

Methods: FDG-PET/CT-scanning was done in the intervention arm of the Dose REduction Strategy of Subcutaneous TNF-inhibitors (DRESS) study, a randomized controlled trial on dose-tapering of biological Disease Modifying Anti-Rheumatic Drugs (bOMARDs) [3]. Baseline and if applicable follow up whole-body FDG-PET/CT scans were performed in consenting patients in the tapering arm to assess predictive value of subclinical PET-arthritis for risk of flaring [4]. The scans were also read by experienced nuclear medicine specialists immediately after they were performed for any unexpected extra-articular finding, conform routine clinical care.

The reference standard was clinical diagnosis of malignancy during the 3 year follow-up. Prevalence of extra-articular abnormalities, follow-up, and received treatments were evaluated prospect to post-hoc.

Results: 121 scans were made in 79 patients. Extra-articular abnormalities were found in 59/121 (48.8%) scans (Table 1) in 45/79 (57%) patients.

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Table 1.

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Disclosure of Interests: None declared


POSI026 UTILITY OF THE SUBCHONDRAL BONE ATTENUATION COEFFICIENT OF THE SACROCILIAC MARGINS TO DIFFERENTIATE SPONDYLOARTHRITIS AND OSTEITIS CONDENSANS ILI.

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Background: Differentiating anklyosing spondylitis (AS) from osteitis condensans ili (OCI) remains challenging for clinicians.

Objectives: The aim of this study was to determine whether Subchondral Bone Attenuation Coefficient of the Sacroiliac margins (SBAC-SI) is different in AS, OCI and diffuse idiopathic skeletal hyperostosis (DISH).

Methods: A monocentric retrospective observational study was performed at a French University Hospital. Patients included were followed for AS, DISH or OCI and underwent CT scan including sacroiliac joint. Patients with tumor lesion of bone or a history of pelvic radiotherapy were excluded. AS and OCI patients were matched with a control of the same age and sex. All scans were acquired on the same CT-scan unit (Somatom 64 definition AS+, Siemens Healthineers, Erlangen, Germany), with a slice thickness of 0.625mm. In the coronal oblique plane of the SIJ, three slices (anterior, middle and posterior) and four quadrants per joint were defined. Twenty-four identical circular regions of interest (ROIs) (30 mm2), 8 per slice, were manually placed separately subcortical to the SIJ, four on the sacral side and four on the iliac side. The distance between the circle of the ROI and the cortical bone was 2 to 3mm. An overall score was obtained from...