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THU0282

EPIDEMIOLOGY OF CUTANEOUS INVOLVEMENT IN SJÖGREN'S SYNDROME: DATA FROM THREE FRENCH POPULATIONS OF PSS (TEARS, ASSESS, DIAPSS)

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Background: Cutaneous involvement is common during primary Sjogren's Syndrome (pSS) but prevalence and characteristics are difficult to establish precisely because of the limited number of patients studied in most cohorts, the variability of the disorders evaluated in each cohort, the rarity of some of them, and the heterogeneity of evaluations from previous studies (1).

Objectives: To determine the prevalence and significance of dermatological disorders in primary Sjogren Syndrome.

Methods: We use 2 French cohorts (ASSESS, in which prevalence of skin disorders in 395 pSS patients was evaluated, and DIAPSS in which 91 consecutive pSS patients had an examination by a dermatologist) and baseline data of the TEARS randomized trial (110 patients with recent or active pSS, treated with rituximab or placebo, and evaluated for skin dryness using a visual analogue scale out of 100).

Results: Skin manifestations included in the ESSDAI were rare in the ASSESS cohort (n=16/395, 4.1%, mainly purpura; only 3 had high activity) but associated with activity in the other ESSDAI domains (peripheral neurological (p<0.001), muscular (p=0.01), hematological (p=0.017) and biological (p=0.017)), history of arthritis (p=0.008), splenomegaly (p=0.024) and higher gamma globulin level (p=0.008) (Table). Compared to pSS patients not receiving a dermatological consultation, the pSS patients who had a dermatological consultation had significantly more dermatological involvement outside ESSDAI score [42% (29/69) versus 19.6% (11/56); p=0.008]. The TEARS study showed a high prevalence of cutaneous dryness (VAS>50; 48.2%) and that these dry skin patients had higher pain VAS (61.5±28.2 vs 46.8±27.0; p=0.003) and drought (79.4±15.2 vs 62.5±21.7; p<0.0001).

	Cutaneous involvement	No Cutaneous involvement	Total	p values*
Muscular	3/16 (18.8)	10/373 (2.7)	13/389 (3.3)	0.001
Peripheral nervous system PNS	4/16 (25)	34/373 (9.1)	38/389 (9.8)	0.00001
Biological	8/16 (50)	138/371 (37.2)	146/387 (37.7)	0.017
Hematologic	7/16 (43.8)	55/373 (14.7)	62/389 (15.9)	0.017
History of arthritis	12/16 (75)	154/374 (41.2)	166/390 (42.6)	0.008
History of splenomegaly	2/16 (12.5)	10/376 (2.7)	12/392 (3.1)	0.024
History of lymphoma	0/16 (0)	18/379 (4.7)	18/395 (4.6)	0.372
Mean (SD) ESSDAI score	14.5 (6.8)	4.4 (5.1)		0.00001
Mean (SD) ESSDAI score after excluding the points awarded by skin manifestations	8.1 (6.2)	4.4 (5.1)		0.014
Gammaglobulin levels (mean ± SD)	23.1 ±7.3	18.5 ±8.1	-	0.006

Conclusion: The most common skin disorder is dryness, which is associated with a higher level of pain and overall subjective dryness. ESSDAI skin activity is rare, associated with hypergammaglobulinemia and ESSDAI activity. Systematic dermatological examination is informative for non-specific pSS lesions

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THU0283

DISTINCT CLINICAL FEATURES OF LATE-ONSET SYSTEMIC LUPUS ERYTHEMATOSUS AMONG MALAYSIAN MULTI-ETHNIC COHORT

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Background: Systemic Lupus Erythematosus (SLE) commonly affects young women in their reproductive age group. However, there is an increase prevalence of late-onset SLE, parallel to the higher life expectancies among general populations worldwide. It has been reported that up to 25% SLE populations have a later onset of disease and their disease expression and course may be different.

Objectives: To determine the clinical features and outcomes of late-onset SLE patients in a multi-ethnic Malaysian cohort.

Methods: Medical records of SLE patients who attended regular follow-up clinics in Universiti Kebangsaan Malaysia Medical Centre (UKMMC) from 2011 until June 2019 were reviewed. Late-onset SLE was defined as the onset of SLE symptoms or diagnosis after the age of 50 years old. Information on their socio-demographics and disease characteristics were obtained from the clinical records. Disease damage was assessed using the SLICC/ACR (Systemic Lupus International Collaborating Clinics/American College of Rheumatology) Damage Index (SDI) scores. The disease characteristics and autoantibody profiles were compared between late-onset and younger onset patients. Damage accrual at disease onset and at 5 years was obtained and compared between the two groups.

Results: A total of 429 patients were included and majority of them were Malays (n= 225, 52.4%) followed by Chinese (n=180, 42%), Indian (n=21, 4.9%) and others (n=3, 0.7%). This multi-ethnic SLE cohort was consisted of predominantly female patients (n=372, 86.7%) with disease duration of 9.9 years ± 6.8 years. A total of 13.8% (n=59) had late onset SLE with mean onset of disease at 58.1 ± 6.3 years while younger group was 27.2 ± 9.4 years. The commonest system involvement among the late-onset group was haematological manifestation (69.5%). Compared to the younger-onset SLE, late-onset SLE occurred significantly higher among the Chinese (66.1%) as compared to Malay (32.3%), Indians and other ethnics (1.7%), p<0.01. Patients with late-onset SLE also had significantly less musculoskeletal (37.3% vs 62.4%) and renal (23.7% vs 71.1%), p<0.001 and tend to have less mucocutaneous manifestations (28.8 vs 42.4%, p=0.06). Meanwhile, pulmonary involvement was more common among the late onset SLE patients (11.9% vs 0.8%, p<0.001). Extractable nuclear antigen (ENA) results were available in 197 patients and patients with late-onset SLE had significantly higher rate of anti-RO positive (63% vs 3.9%), p=0.01. Otherwise, no significant difference in the other autoantibodies expressions including anti-La, anti-Sm, anti-RNP, anti-ribosomal P and anti-phospholipid antibodies. Patients with late-onset SLE tend to have more damage accrual at 5 years as compared to the younger age group (p=0.07). The mortality in the late onset group was 13.6% (n=8) as compared to 2.7% (n=10) in the younger age group, p=0.01. Majority of the cause of death in the later onset SLE was infection (87.5%) while in the younger age group was infection and active disease (90%).

Conclusion: Late onset SLE occurs more commonly among Chinese ethnics in Malaysia and Malaysian SLE patients with late onset of the disease have distinct clinical manifestations. Damage accrual at 5 years tend to be higher in the late-onset group and the mortality is significantly higher with the major cause of death is infection. The different disease expression and outcome in late onset SLE suggest different factors in influencing the disease course and hence further studies including their genetic profiles are warranted.

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THU0284 PREVALENCE OF NEUROPSYCHIATRIC LUPUS IN PSYCHOSIS PATIENTS WITH A POSITIVE ANTINUCLEAR ANTIBODY

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Background: Psychosis is a rare manifestation of Neuropsychiatric Systemic Lupus Erythematosus (NPSLE). Patients with SLE may have Psychosis as part of their initial presentation of disease¹. Current guidelines do not make a recommendation regarding the use of Antinuclear Antibody (ANA) in the assessment of patients with psychosis². There is limited evidence assessing the utility of ANA testing in this setting.

Objectives: Primary objective: Determine the prevalence of NPSLE in patients admitted to a mental health service with a diagnosis of a psychosis, who have had a positive antinuclear antibody test.

Secondary objectives: Determine the frequency and proportion of positive ANA testing in this patient group. Determine the pattern and titres of positive ANAs. Determine the subsequent investigation, referral and diagnosis of patients with positive ANAs.

Methods: Retrospective chart review of patients admitted to a mental health service of two metropolitan tertiary referral centres, Prince of Wales Hospital (POWH) and Royal Prince Alfred Hospital (RPAH), with a diagnosis of psychosis who had been tested for ANA. Patients were identified using their electronically entered diagnosis based on the International Classification of Disease³ codes. Assessment of patient data for SLE used the 2019 ACR/EULAR classification criteria⁴. Decisions regarding attribution of psychosis related events to SLE follows the criteria used by Bortoluzzi et al⁵.

Results: Between 1st of January 2010 and 31st of March 2018 there were 5585 (POWH) and 4620 (RPAH) mental health admission with an ICD diagnosis of psychosis representing 2451 and 2315 individual patients. 449/2451 (18%) and 462/2315 (20%) patients were tested for ANA. 78/449 (17%) and 57/462 (12%) were positive. Discharge data was available for all patients and long-term follow up data was completed for 53/78 (81% - POWH) patients and 50/57 (88% - RPAH). The mean follow-up time 43 ± 23 months and 51 ± 29 months respectively.

At discharge there were four patients who met 2019 ACR/EULAR for SLE. Of these, two patients met criteria for NPSLE. One was diagnosed clinically and treated specifically for NPSLE with intravenous methylprednisolone and rituximab.

There were no additional diagnoses of SLE or NPSLE clinically or by criteria found in the available follow up data. Hence the overall prevalence of NPSLE in patients admitted with psychosis was 1.3%, 95%CI [0.6,9%] and 1.8%, 95%CI [0.9,4%] respectively.

Conclusion: The prevalence of neuropsychiatric lupus in patients with psychosis and a positive ANA was 1/78 and 1/57 a two tertiary referral centres. This study expands significantly on the limited evidence available as to the expected outcomes of a positive ANA test in a patient with psychosis.

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THU0285 ANALYSIS OF THE RELATIONSHIP BETWEEN ORGAN DAMAGE AND QUALITY OF LIFE IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: Systemic lupus erythematosus (SLE) is an autoimmune disease that can not only cause systemic symptoms, such as fever and arthritis, but can also damage important organs, such as those of the central nervous system and the kidneys. Prevention of irreversible organ damage is important for better prognosis [1]. Additionally, the importance of maintaining the quality of life (QOL) of patients has recently been emphasized. However, only a few studies have examined the relationship between irreversible organ damage and patient QOL.

Objectives: To assess the relationship between organ damage and QOL, and to survey which organs have more significant effects on QOL.

Methods: We conducted a questionnaire-based survey of 183 patients with SLE at Kyoto University Hospital from September to December 2019. We used the SLICC/ACR Damage Index (SDI) to evaluate organ damage [2]. The following five scales were employed to evaluate QOL: the physical (PCS) and mental component summary (MCS) of the Medical Outcome Study (MOS) 36-Item Short-Form Health Survey version 2.0 (SF-36v2) [3], health (HRQOL) and non-health-related QOL (N-HRQOL) of LupusPRO [4], and SLE Symptom Checklist (SSC) [5].

Results: Linear regression analysis showed significant correlation between the SDI score and all QOL scales except for N-HRQOL, suggesting negative effects of organ damage on QOL (Table 1). Next, we analysed whether there was a significant difference in the SF-36 score between those who were positive and negative for each SDI item (41 in total), using the Wilcoxon rank sum test. Muscle atrophy or weakness ($p = 3.0 \times 10^{-10}$), osteoporosis with fracture or vertebral collapse ($p = 9.7 \times 10^{-8}$), claudication ($p = 7.4 \times 10^{-5}$), and cognitive impairment or major psychosis ($p = 9.9 \times 10^{-5}$) significantly correlated ($p < 1.2 \times 10^{-3}$) with PCS, and scarring chronic alopecia ($p = 3.4 \times 10^{-4}$) with MCS (Table 2). In addition, the five SDI items significantly correlated with the remaining three QOL scales (HRQOL, N-HRQOL, and SSC; $p < 0.05$).

Table 1. Relationship between the SDI score and QOL

	SF-36		LupusPRO		SSC
	PCS	MCS	HRQOL	N-HRQOL	
p-value	$<2.0 \times 10^{-16}$	1.7×10^{-3}	2.2×10^{-11}	0.23	1.9×10^{-8}

Table 2. Relationship between each SDI item and the SF-36 score ($p < 1.2 \times 10^{-3}$)

SDI item	PCS score		p-value
	Positive (Median (IQR))	Negative (Median (IQR))	
Muscle atrophy/weakness	33 (19-45)	50 (43-54)	3.0×10 ⁻¹⁰
Osteoporosis with fracture/vertebral collapse	24 (12-32)	49 (38-54)	9.7×10 ⁻⁸
Claudication	31 (19-35)	49 (38-54)	7.4×10 ⁻⁵
Cognitive impairment/psychosis	27 (17-33)	49 (38-54)	9.9×10 ⁻⁵
SDI item	MCS score		p-value
	Positive (Median (IQR))	Negative (Median (IQR))	
Scarring chronic alopecia	42 (29-51)	49 (39-54)	3.4×10 ⁻⁴

Conclusion: We demonstrated that organ damage has negative effects on patient QOL, indicating the importance of preventing irreversible organ damage for maintaining QOL. Moreover, muscle atrophy/weakness, osteoporosis with fracture/vertebral collapse, claudication, cognitive impairment/major psychosis, and scarring chronic alopecia significantly correlated with QOL deterioration, suggesting that these items should be examined with special care in clinical practice.

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