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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.6249

FRI0384 PRIMARY SJÖGREN'S SYNDROME STRATIFIED BASED ON THE SEVERITY OF PATIENT-REPORTED FATIGUE

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Background: fatigue is one of the most common symptoms reported by patients affected by primary Sjögren's syndrome (pSS), and a major contributor to impaired quality of life.

Objectives: to analyse the clinical, serological and histological features of pSS patients stratified according to the severity of their self-reported fatigue.

Methods: among pSS patients undergoing clinical evaluation in our Sjögren's Clinic in a six-months period (January-June 2017), 86 consecutive unselected patients, fulfilling the latest ACR/EULAR pSS classification criteria, accepted to report their degree of fatigue on a 10 cm VAS (range 0–100) and to complete the ESSPRI questionnaire. Four subgroups of fatigue severity were defined, as previously published¹: no fatigue (VAS=0); low fatigue (VAS=1–24); moderate fatigue (VAS=25–74); high fatigue (VAS=75–100). For each subgroup demographic, serological, histological features and ESSDAI score were collected, as well as the prevalence of pSS-related lymphoma, fibromyalgia (FM), autoimmune thyroiditis, and anaemia.

Results: fatigue was reported by the 87.2% (n=75) of pSS patients, distributed in subgroups as following: 25.3% (n=19) with low fatigue, 58.7% (n=44) with moderate fatigue and 16% (n=12) with high fatigue. Lymphoma was significantly (p=0.0133) more frequent in the pSS subgroup with high fatigue (33.4%, by considering active lymphoma cases; 50%, by considering also the cases with lymphoma in remission). FM patients were a minority (4.7% ; n=4), and never complained of high fatigue, all of them reporting moderate fatigue. A significant correlation was finally found between fatigue severity and ESSPRI (p<0.0001), but not with ESSDAI (p=0.31). No significant age or sex difference was observed between subgroups. Also autoimmune thyroiditis, anaemia, anti-SSA and/or anti-SSB positivity, rheumatoid factor positivity, and cryoglobulinemia showed no significant difference between subgroups.

Conclusions: when fatigue is better stratified in pSS, it appears that it is usually moderate or severe, rather than mild. Furthermore, it is unrelated to FM. Overall, fatigue appears as a consequence of pSS itself. Of note, severe fatigue was related in this study with the most important complication influencing patient survival in pSS, i.e., lymphoma. Further studies are needed to disclose the pathogenetic events leading to fatigue in pSS, and investigation of lymphoma in pSS might be also helpful to this end.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.6348

FRI0385 CLINICAL CHARACTERISTIC FEATURES OF BRAINSTEM ENCEPHALITIS IN NEUROPSYCHIATRIC SYSTEMIC LUPUS ERYTHEMATOSUS

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Background: Neuropsychiatric manifestation in systemic lupus erythematosus (NPSLE) is one of the most serious complications of the disease. The American College of Rheumatology (ACR) developed standardised nomenclature and case definitions for neuropsychiatric involvement in SLE (NPSLE) in 1999. Although myelopathy is included in the ACR nomenclature, no space is provided for the brainstem encephalitis. Moreover, it is still unclear whether the brainstem encephalitis is an independent clinical entity in NPSLE.

Objectives: The present study was designed in order to disclose the clinical characteristic features of the brainstem encephalitis in NPSLE.

Methods: We prospectively collected the patients who presented brainstem lesions from 2005 to 2015. The diagnosis of the brainstem encephalitis was made by the elevation of cerebrospinal fluid (CSF) IL-6, Magnetic Resonance Imaging and response to steroid. Serum autoantibodies, including anti-ribosomal P protein antibodies (anti-P), anti-NMDA receptor NR2 antibodies (anti-NR2), anti-Sm antibodies and anti-aquaporin 4 antibodies (anti-AQP4) and anti-cardiolipin antibodies (anti-CL), were measured by ELISA.

Results: Seven patients presented the brainstem encephalitis during the period of 10 years. All the patients showed the elevation of CSF IL-6 (8.3–2716 pg/ml, median 377 pg/ml), whereas CSF cell count was elevated in 6 patients. The most common clinical symptoms were headache (4 patients), vertigo (3patients) and consciousness disturbance (3 patients). All the patients showed high intensity lesions in FLAIR images on MRI. Notably, 5 of the 7 patients also showed the high intensities periventricular regions, including cerebral aqueduct and the 4th ventricle (figure 1). Anti-NR2 and anti-P were positive in 7 patients and 6 patients, respectively. By contrast, anti-CL, anti-Sm and anti-AQP4 were positive in 4 patients, 3 patients and 1 patient, respectively. Six patients recovered after steroid therapy, while 1 patient died due to severe brainstem damages at the onset.

Cerebral aqueduct

The 4th ventricle

Conclusions: These results indicate that the brainstem encephalitis is an independent clinical entity in NPSLE, characterised by reversible high intensity lesions, especially in periventricular areas. Moreover, the data demonstrate that anti-AQP4-related neuromyelitis optica disorder is a minor population within the brainstem encephalitis. By contrast, it is suggested that anti-NR2 and anti-P might play an important role in the pathogenesis of the brainstem encephalitis.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4056

FRI0386 WORK DISABILITY AND QUALITY OF LIFE AMONG MULTI-ETHNIC MALAYSIAN SYSTEMIC LUPUS ERYTHEMATOSUS (SLE) PATIENTS

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Background: Patients with Systemic Lupus Erythematosus (SLE) are at risk of work disability due to the substantial impact of the disease towards their physical and mental health.

Objectives: To study the prevalence of work disability (WD) and unemployment rate among SLE patients, and their associations with the quality of life (QOL) in National University of Malaysia Medical Centre (UKMMC)

Methods: This was a cross-sectional study which recruited consecutive SLE patients who attended the Nephrology and Rheumatology clinic at Universiti Kebangsaan Malaysia Medical Centre (UKMMC) from March 2017 to July 2017. Information on their current and past employment history were obtained from a customised questionnaire and WD was defined as unemployment or an inability to do paid work due to illness at the time of study or at any time after the diagnosis of SLE being made. The disease characteristics, disease activity and damage were determined from the medical records. The quality of life was measured using SF-36 questionnaires. Statistical analyses were subsequently performed to determine the factors associated with WD. This study was approved by the UKMMC ethics research committee (FF FF-2017–109).

Results: A total of 197 patients were recruited, and their median age was 37 (32 to 47) years with median disease duration of 12 (8 to 17) years. Majority of them were Malays (n=116, 58.9%), followed by Chinese (n=69, 35%), Indians (n=8,