FRI0266 COMPARISON OF CLINICAL AND SEROLOGICAL CHARACTERISTICS BETWEEN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS WITH AND WITHOUT ASSOCIATED JACCOUD ARTHROPATHY

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Background: Jaccoud arthropathy (JA) is a chronic non-erosive reversible joint disorder commonly associated with systemic lupus erythematosus (SLE), and occurs in roughly 5% of all cases. Some studies suggest different profiles of clinical and immunological features between SLE patients with or without JA.

Objectives: To compare the profile of clinical and serological manifestations of patients with SLE with and without JA followed in a tertiary care hospital of Madrid. **Methods:** We performed a retrospective observational study of a cohort of patients diagnosed with SLE (4 or more ACR criteria) between June 1977 and December 2015. The variables evaluated included demographics, clinical, analytical and radiological manifestations. The definition of JA was based on the presence of clinical criteria (reversible joint deformities) and absence of radiographic erosions typical of rheumatoid arthritis. Statistical analysis was performed using SPSS software version 21.0.

Results: We included 108 patients representing a sample of 24% of the total number of patients with SLE treated at our center during that period. The majority of patients were women (89.8%), mean age at diagnosis was 30±12.29 years (range: 7–75) and duration of disease was 127 months (range: 2–411). Thirteen patients (12.03%) had findings compatible with JA. There were no significant differences in age, sex or race, but the duration of disease was higher in JA patients (190 vs. 118.2 months, $p=0.0299$). There were significant differences in the presence of malar rash ($p=0.0009$), photosensitivity ($p=0.0050$), oral ulcers ($p=0.0032$) and pericarditis ($p=0.00001$), which were more frequent in patients without JA, but arthritis, nephritis, pleuritis, seizures, psychosis, Raynaud's phenomenon and antiphospholipid syndrome had a similar distribution between both groups. Among the immunological features, no significant difference was found in relation to hemolytic anemia, lymphopenia, thrombocytopenia, ANA, anti-ENA, anti-DNA, anticardiolipin, anti- β_2 glycoprotein I and lupus anticoagulant, but leukopenia was also more frequent in patients without JA ($p=0.0041$).

Conclusions: In the analyzed sample of patients in our center JA was a relatively frequent finding and was associated with a longer duration of the disease. It was not possible to corroborate other JA associations suggested in previous studies such as a lower frequency of lupus nephritis or major secondary antiphospholipid syndrome, probably due to limited sample size, but there are also other studies that do not demonstrate significant differences in relation to clinical and serological findings in patients with SLE with JA with respect to those who do not present it. Studies involving a larger number of patients with JA associated with SLE are required in order to better identify these differences, so a multicenter initiative could be of great help.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.6980

FRI0267 HEMOPHAGOCYTIC SYNDROME IN PATIENTS FROM SLE REGISTRY FROM THE SPANISH SOCIETY OF RHEUMATOLOGY (RELESSER)

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Background: Systemic Lupus Erythematosus (SLE) is an autoimmune systemic rheumatic disease that, in our area, presents hematologic manifestations in approximately 70% of cases¹. Some of them are very rare so there are no large series whose analysis could provide relevant information.

Objectives: To study the characteristics of patients with Hemophagocytic Syndrome (HS) in a large sample of SLE patients.

Methods: SLE patients from RELESSER database were studied. We analysed SLE manifestations present at 12 different domains (mucocutaneous, renal, musculoskeletal, constitutional, hematologic, vascular, cardiac, respiratory, neuropsychiatric, gastrointestinal, ophthalmic and serological) before, during and after HS diagnosis and until the last available assessment. We also studied activity (SELENA-SLEDAI) and damage (SLICC/ACR DI) indices at those moments. We evaluated the treatment received, HS recurrences and the number of deaths by this entity.

Results: 3,656 SLE patients (≥ 4 ACR criteria) from 45 Rheumatology Units across Spain were studied. Seven patients ($<0.5\%$) with HS were identified. 71.4% were women, with a mean age ($\pm$ S.D.) at SH diagnosis of 35.1 ($\pm$ 17.1) years. In 5 of the 7 cases the HS occurred 115.5 ($\pm$ 162.9) months after SLE diagnosis. In the other 2 cases the diagnosis of both entities was simultaneous. The main triggers of HS were infections, followed by SLE activity flares. At the time of HS diagnosis, they had high SLE activity with a mean SLEDAI score of 13.1 ($\pm$ 11.3) and 1.4 ($\pm$ 2.3) SDI score.

Clinically, 100% of the patients presented fever and alterations of the liver profile, 85.8% cytopenias and 71.5% dermatological manifestations. Respiratory manifestations and hemolytic anemia were present in 57.2% of the cases, both; lymph nodes and coagulopathy in 42.9%. Hepatomegaly was detected in 28.6%, as well as neuropsychiatric, digestive and renal manifestations. Splenomegaly was detected in 14.3%. The mean hemoglobin level was 8.6 ($\pm$ 1.1) g/dl, platelets 85,585 ($\pm$ 83,390)/mm³, ferritin 7,410 ($\pm$ 6,470) ng/ml and triglycerides 404.7 ($\pm$ 235.6) mg/dl. All patients were admitted and undergone bone marrow study, requiring a mean of 2.2 ($\pm$ 1.5) treatment lines, using 2.8 ($\pm$ 1.7) drugs. One patient died during the HS episode by the HS itself and another 2 patients had 2 and 3 recurrences, respectively.

The following table shows the characteristics of each patient.

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6	Patient 7
Number of organ systems affected by SLE before HS diagnosis	9	4	5	5	6	Simultaneous diagnosis of SLE and SH	Simultaneous diagnosis of SLE and SH
Number of organ systems affected by SLE at HS diagnosis	4	1	2	4	2	4	3
Number of organ systems affected by SLE until last assessment	Died	2	2	Follow-up lost	1	5	1
SLEDAI/SLICC-ACR DI at HS diagnosis	4/6	4/0	5/0	29/3	4/1	25/0	21/0
SLEDAI/SLICC-ACR DI 1 year after HS	*	0/0	0/0	*	2/1	0/0	2/13
SLEDAI/SLICC-ACR DI at last assessment	*	0/0	0/0	*	0/1	2/0	2/13
Number of treatment lines	1	0	3	2	2	4	3
Number of treatments administered	2 GC and CsA	0	5 GCs, etoposide, iv Ig, CsA, platelets, red cells	3 GC, CYP and iv Ig	2 Amphotericin B, miltefosine	4 GC, iv Ig, CsA, MM	4 GC, CsA, anakinra and CYP
Relapses	0	2	0	3	0	0	0
Deaths	Yes	No	No	No	No	No	No
Follow-up time (months)	Died	45	80	Follow-up lost	Unknown	26	24

GC: Glucocorticoids, iv Ig: intravenous immunoglobulins, CYP: Cyclophosphamide, CsA: Cyclosporine A, MM: Mycophenolate mofetil

Conclusions: HS is a rare life-threatening SLE manifestation. It must be suspected in SLE patients with persistent fever who do not respond to antibiotics, cytopenias and evidence of multiorgan involvement. Relapses and death are common in HS associated to SLE.

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Acknowledgements: Work supported by Spanish Society of Rheumatology, FIS/ISCIII (P11/02857), BIOCAPS from the European Union 7th Framework Programme/REGPOT-2012–2013.1 (316265), GSK, Roche, Novartis, UCB.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.2542

FRI0268 ULTRASOUND VS. MAGNETIC RESONANCE IMAGING IN DIAGNOSIS OF PRIMARY SJÖGREN'S SYNDROME

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Background: Salivary gland (SG) biopsy is the gold standard for the diagnosis of primary Sjögren's syndrome (pSS). Ultrasound (US) and magnetic resonance imaging (MRI) of the SG are emerging techniques. The comparison of both imaging methods for diagnosis and assessment of functional limitation is yet not well investigated.