HEMOPHAGOCYTIC SYNDROME IN SYSTEMIC LUPUS ERYTHEMATOSUS. A MONOCENTRIC STUDY OF 20 CASES

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Objectives We reviewed the medical records of adult patients with SLE for a recent 8 years period and identified patients who had developed HS. The diagnosis of SLE was made using ACR criteria (4 or more criteria) and HS has been diagnosing using Hunter criteria (5 or more). We conducted statistical analyses to identify the characteristics of those patients in comparison with SLE patients without HS.

Results Among 208 consecutive lupus patients, 20 patients (19 women) was identified having HS. The mean age of patients was 35.4±11.4 years (21–68). HS revealed lupus in 7 patients; in the others the delay between diagnosis of SLE and HS was 33 months (1 months – 108). Fever, pericarditis and splenomegaly were found in 95%, 70% and 50% of patients at presentation of HS. Bone marrow aspiration indicated hemophagocytosis in all patients. Cutaneo-mucous and arthritis were present in 95% and 70% of patients at presentation of HS. Lower serum C-reactive protein level, a higher ferritinemia, higher SLEDAI and H score and, splenomegaly.

Conclusion HS was observed in 9.26% Moroccan patients with SLE. Recognition of the cause of HS was particularly challenging because it may mimic the clinical features of the underlying disease or be confused with an infectious complication.

IDENTIFYING THE RATES OF IRON DEFICIENCY AND ANAEMIA IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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Background Haematological abnormalities are commonly seen in patients with systemic lupus erythematosus (SLE). Anaemia is frequently reported although there is little data in the literature regarding iron deficiency.

Purpose To identify the prevalence of anaemia and iron deficiency in a large cohort of patients with SLE.

Methods From February 2017, patients fulfilling revised ACR criteria of lupus were prospectively recruited from the Lupus
Clinic at University College London Hospital, UK. Standard haematological measures including Haemoglobin (Hb), Red Blood Cell Distribution Width (RDW) and Mean Corpuscular Volume (MCV) were recorded. Anaemia was defined by World Health Organisation (WHO) criteria; Hb <120 g/L in women and <130 g/L in men. Iron status was assessed by serum Ferritin and Transferrin saturations.

Results
A total of 284 patients were recruited. 93% (265/284) of patients were female with ages ranging between 16–82 years old (median 44; IQR 32–56). Hb ranged from 88–167 g/L (median 127; IQR 117–135). A third of patients (33%; 95/284) were found to be anaemic. Of those who were anaemic, 89 were female and 6 were male. A low MCV is suggestive of iron deficiency; 5.6% (16/284) of patients were microcytic while 19% (54/284) were macrocytic. In those who were anaemic, 13% were microcytic (12/95) and 18% were macrocytic (17/95). RDW was typically at the upper limit of the normal range (median 14.1%; NR 11.5%–14.5%). 38% (107/284) of patients had an elevated RDW (>14.5%). Ferritin levels were highly variable, ranging from 6–2536 ng/mL (median 62 ng/mL; IQR 38–123 ng/mL). Ferritin <20 ng/mL (suggestive of iron deficiency) was seen in 12% (35/284) of patients. Transferrin saturations<16% (suggestive of iron deficiency) were seen in 20% (57/284) of patients. These results are summarised in figure 1.

Conclusions Anaemia is a common feature of SLE (affecting one third of patients in this cohort). It is however difficult to verify how many cases are due to iron deficiency. Ferritin is a poor marker of total iron and is likely to be elevated in the context of inflammation. RDW (a marker of early iron deficiency) is however elevated in 38% of patients with SLE in this cohort.

Fatigue is the main symptom in up to 80% of SLE patients. Its origin is presumably multifactorial and it is an important factor in the reduced quality of life. This study tries to identify possible causes of fatigue in SLE patients.

In preparation of this study, the literature discussing fatigue in SLE patients was reviewed to identify possible associated factors. These and supposed additional factors were then investigated in a retrospective study of 332 SLE outpatients from the Polyclinic for Rheumatology of the medical faculty of the Heinrich-Heine-University Düsseldorf. This study population included 297 females and 35 males aged 19–81 years with an average disease duration of 13 years (range 0–41 years). Patient data were collected during their baseline visits in 2014 and 2015. The Fatigue Severity Scale (FSS) was used to measure fatigue. This instrument covers nine items associated with fatigue and allows patients to assess its severity on a scale from 1–7. An average result in the FSS >4 points is considered as severe fatigue. In this population the average result of the FSS was 3.76 points (range 0.89–7.0), in total 44% of all patients reached 4 points or more. In a univariate logistic regression a pathological result in the FSS >4 was then compared with patients’ demographic and clinical data such as age, gender, disease activity and duration, depression, physical activity, pain, anaemia, vitamin D deficiency, sleep quality, target organ damage, obesity, hypothyroidism, infection, and current medication. A linear regression analysis was adjusted for potential confounders such as age, sex and disease activity.