HYPERFERRITINAEMIA AND SYSTEMIC DISEASE : SLE OR STILL’S DISEASE? A Kella, N Bouziani, A Belabbas, A Khedim, D Hakem*. Internal Medicine Department, University Hospital Center, – Mostaganem – Algeria

Abstracts

Introduction Major hyperferritinemia - after exclusion of hemochromatosis - constitutes a major biological argument of inflammation which characterizes Still’s disease (SD), a condition bordering on auto-inflammatory and inflammatory diseases. Despite the well-established diagnostic criteria for SD, its diagnosis requires the exclusion of other exclusion of other conditions such as systemic SLE and lymphomas. The clinical similarities between SLE and SD are causes of diagnostic wandering.

Purpose To report case of a SLE simulating SD posing the nosological boundaries between inflammatory diseases and autoinflammatory diseases.

Case Report 16 years old woman was explored for a diffuse macular eruption with recrudescence in the evening, with deterioration in general condition, odynophagia, hectic fever, tachycardia with heartbeat at 120/min and joint damage with wrist flexion. The evolution was enameled by a macrophage activation which responded favorably to bolus of corticosteroids. Biologically, serum ferritin level was over 1200 with low level of glycosylated ferritin. The initially negative autoimmunity assessment came back polymorphically positive including anti-Sm+ AAN. The inflammatory showed ERS at 105, polyclonal major gammaglobulinemia at 24 g/l. albuminemia at 23.97g/l, the creatinine clearance at 68.35ml/min, increase levels of ASAT at 241 (x7N), and ALAT (X4n) prothrombin at 100%, Hb level at 10.5g/dl, leucopenia at 3500 and platelet rate at 97000. The medullogram showed no suspicious cells, no leishmianiasis or activated macrophages. The thyroid tests, the blood ionogram, the phosphocalcic and the hemostasis balance were without abnormalities. The triglyceride (TG) level was over 5g/l with a low HDL level of 0.30 g/l and a cholesterol level of 1.4g/l. The chest radiography revealed a small amount of pleural fluid effusion and echocardiography found a pericardial effusion with preservation of myocardial functions. The evolution was enameled by flare-ups confining the patience to bed, myocardiitis and proteinuria leading to a renal biopsy which revealed an aspect of WHO stage 2 lupus nephropathy and signs of vasculitis. Treatment with corticosteroids, synthetic antimalarials, methotrexate allowed clinical stabilization.

Discussion The presence of evening febrile eruptions, odynophagia and hyperferritinemia associated with articular manifestations and macrophage activation argued for SD disease. The positivity of the immunological assessment and the successive occurrence during the follow-up of renal damage and myocarditis made us opt for a SLE disease in its pseudo-Still form. The background treatment remains the same for both situations.

Conclusion SLE can take on the appearance of an SD. This clinical similarity is overcome by the positivity of an autoimmunity assessment - element of exclusion of SD - which can appear secondarily. This difficult clinical situation nevertheless raises the nosological boundaries between his two conditions which fortunately benefit from a common therapy (corticosteroids, immunosuppressants such as methotrexate, biotechnology such as anti-IL 6 and anti-IL1).

Thursday 06 October 2022 from 13:00 to 14:10

Po.3 E-poster session 3: cardiovascular manifestations, cytokines and complement system in SLE, difficult cases, organ damage and survival, innate immunity

PREDICTORS OF CARDIOVASCULAR EVENTS IN SYSTEMIC LUPUS ERYTHEMATOSUS N Farina, J Webster, W Luo, D Isenberg, A Rahman*. Centre for Rheumatology Research, Division of Medicine, University College London – UK

Purpose Systemic Lupus Erythematosus (SLE) is associated with an increased cardiovascular risk. Several traditional and disease-specific risk factors have been shown to correlate with the occurrence of cardiovascular events (CVE) in patients with SLE. However, results of previous studies are heterogeneous. The objective of this study was to evaluate the potential predictors of CVE in a large, multiethnic, monocentric cohort of patients with SLE and a long follow-up duration.

Methods Medical records of patients treated at the Lupus Clinic at University College London Hospital (UCLH) between 1979 and 2022 were reviewed. Data about CVE, traditional cardiovascular risk factors, demographic and disease features, and treatment history were collected. Only patients with complete information available were included in the study. Firstly, descriptive analysis was performed to compare features of patients who had a CVE and patients who did not. Subsequently, inferential statistical analyses with ad-hoc proportional hazards models were performed to identify predictors of CVE.

Results Four hundred and nineteen patients were included in the study. Maximum follow-up length was 40 years. Demographic and disease features, as well as prevalence of traditional cardiovascular risk factors are shown in Table 1. Seventy-one (17%) patients had at least one CVE, whereas two different CVE were observed in nine (2%) cases. Forty (12%) patients had venous thrombosis, 26 (6%) had stroke, and 14 (3%) had coronary disease. Mean time to CVE was...
14 (SD 8) years. While both diabetes and antiphospholipid antibodies positivity were associated with the outcome at univariable analysis (figure 1), multivariable analysis showed that only antiphospholipid predicted the occurrence of CVE (Hazard Ratio [HR] 2.95, 95% Confidence Interval [CI] 1.79 – 4.85, p-value < 0.001). Dedicated subanalyses showed that also cumulative dose of glucocorticoid (HR 1.0002, 95% CI 1.00003 – 1.000045, p-value=0.028) and Systemic Lupus International Collaborating Clinics Damage Index score (HR 1.47, 95% CI 1.15 – 1.88, p-value=0.002) were associated with CVE. While both venous thromboembolic events and strokes were predicted by antiphospholipid antibodies positivity (p-value < 0.001 and = 0.007, respectively), only male sex was associated with the specific diagnosis of coronary artery disease (p-value=0.002).

Conclusions Cardiovascular disease is highly prevalent in patients with SLE and is strongly associated with anti-phospholipid antibodies positivity, glucocorticoid therapy, and damage.

Objective The aim of this study was to investigate the relationship between the concentration of hydroxychloroquine (HCQ) in serum and adverse cardiac effects including prolonged QTc interval and cardiomyopathy in patients with systemic lupus erythematosus (SLE) undergoing long-term treatment with HCQ.

Methods The concentration of HCQ in serum from 96 SLE patients treated with HCQ was determined using liquid chromatography – high resolution mass spectrometry. QTc intervals at electrocardiograms were calculated using Bazett’s formula. Cardiomyopathy was determined during follow-up in accordance with the SLICC/ACR-damage index definitions.

Results There were 83% women and the median (range) age at study was 47.5 (21–82) years. As a prerequisite, all 96 SLE patients included in the study were treated with Plaquenil, with 96% stating that they were taking a dose of 200 mg