methylprednisolone (250 mg), AZA shifted to tacrolimus (3 mg/bd) and HCQ started. When discharged, prednisone was reduced to 25 mg/day; LMWH, LDA, HCQ, vitamin D and levothyroxine unchanged. Proteinuria rapidly decreased (1.3 g on 15th May and 0.260 g on 10th July).

On 23rd July she had vaginal delivery at 38 weeks' gestation, her baby was 3.390 kg and 51 cm high; APGAR 10/10. Tacrolimus was suspended on the delivery day and the patient continued with prednisone 5 mg every other day with LMWH during puerperium. A subsequent kidney biopsy confirmed Class V glomerulonephritis.

At last evaluation (October 2019), the patient and the baby were fine, urinalysis did not show proteinuria.

Discussion Points

- Pre-pregnancy counselling for women with a history of LN
- Management of acute flares of LN in pregnancy
- Diagnosing pre-eclampsia in women with active LN
- Discuss the use of immunosuppressive medications in pregnancy

Learning Objectives

- Explain the importance of pre-pregnancy counselling in women with LN
- Discuss the best management of flares of LN in pregnant women
- Discuss how to recognise pre-eclampsia in women with LN
- Distinguish lupus nephritis from APS nephropathy

18 RARE DISEASE MANIFESTATIONS IN SLE

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Case 1: A 20-year-old female with SLE and myocarditis

Zahir Amoura

A 20-year-old female with systemic lupus erythematosus (SLE) was referred for chest pain and skin lesions. The diagnosis of SLE was made 4 years earlier, when she developed arthritis, proteinuria, with positive antinuclear antibodies (1/1280), anti-dsDNA, anti-SS-A, and anti-Sm antibodies. The renal biopsy showed Class I lupus nephritis. The patient was treated with low-dose steroids and hydroxychloroquine. One year later, she had pulmonary embolism and received warfarin because of the presence of persistent lupus anticoagulant. Two months before admission, while the patient was receiving hydroxychloroquine (400 mg/day), hand skin lesions gradually developed.

At admission, the patient complained of chest pain and went to the emergency department. Her temperature was 36.6°C, blood pressure 130/70 mmHg, pulse rate 90 beats per minute, respiration rate 18 breaths/minute, and oxygen saturation 98% while she was breathing ambient air. There was normal breath sounds in the lungs' fields. Laboratory tests showed hemoglobin 10.3 g/dL, lymphocytes 0.9 G/L, normal platelets, international normalised ratio (INR) for prothrombin time 2.3, creatinine 69 µmol/L, elevated ultrasensitive troponin T (248 ng/L), low C3 (0.55 mg/dL), positive anti-dsDNA antibodies (45 U/mL) and proteinuria 1.5 g/24h. Serum protein electrophoresis showed a normal pattern and a diffuse increase

in immune globulin. The tests for lupus anticoagulant, anticardiolipin antibodies, were positive.

Discussion Point

- Diagnosis and management of lupus myocarditis

Learning Objectives

- Describe the clinical presentation of lupus myocarditis
- Discuss the diagnosis workshop for lupus myocarditis
- Discuss treatment options of lupus myocarditis

Case 2: A 22-year-old male with thrombotic microangiopathy

Richard Furie

A 22-year-old male was admitted to the hospital because of headache, confusion and fever. He had been previously well. At the time of admission, he was on no medications. Initial laboratory test results were notable for WBC 2,800, Hb: 6.7; platelets: 64,000 and creatinine: 1.9. The patient was given broad spectrum antibiotics for the treatment of sepsis and/or bacterial meningitis. However, the patient's mental status worsened and he became comatose. All cultures were sterile. A 'shotgun' diagnostic approach revealed ANA 1/2560 (H) and DNA 883 IU/dL. The creatinine continued to rise.

The impression was that of SLE complicated by anemia, thrombocytopenia, nephropathy and CNS disease. 'Pulse' steroids were administered for 3 days without subsequent improvement. Intravenous gamma-globulin failed to raise the platelet count. Rheumatology to the rescue!

Discussion Point

- Diagnosis and management of thrombotic microangiopathy (TMA)

Learning Objectives

- Describe the clinical presentation of TMA
- Discuss treatment options of TMA
- Review proposed modifications to the classification of lupus nephritis

Hot Topic

19 OPTIMAL ASSESSMENT AND MONITORING OF SLE PATIENTS IN CLINICAL PRACTICE

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Systemic lupus erythematosus (SLE) is a heterogeneous disease, characterised by a wide spectrum of clinical manifestations (ranging from arthritis and skin manifestations to renal insufficiency or central nervous system involvement) with an unpredictable relapsing-remitting course. SLE primarily affects young women in their childbearing lifetime, and perinatal complications due to SLE and/or the coexistence of the presence of antiphospholipid antibodies, can occur. Also, low complement levels and many antibodies can be found in SLE, among which anti-ds DNA and anti-Sm. During the disease course, irreversible organ damage can accrue due to SLE and/or its treatment. In addition, comorbidities can develop, including infections, osteoporosis and atherosclerosis. Compared to the general population, quality of life is reduced, frequently due

to severe fatigue or psychological issues, amongst other reasons. As such reduced work capacity and/or work loss may occur.

So, how should we assess and monitor patients with SLE?¹

² The (frequency of) assessments depends on the stage of disease (diagnosis and follow-up). Disease activity (active disease [i.e. every 1–3 months] *versus* stable disease [i.e. every 6–12 months]), and specific conditions (i.e. pregnancy, thrombosis and bone fractures). Measuring disease activity with validated instruments (i.e. SLEDAI-2k, BILAG 2004) at each visit is recommended, in addition to a history and clinical examination, laboratory assessments (including blood, urine and immunology test) and eventually other investigations (i.e. imaging or biopsy); using these instruments results in better outcome, although the evidence is of low-moderate quality. However, high disease activity is associated with poor outcome (higher mortality rate, higher level of organ damage and comorbidities). Conversely, lower levels of disease activity and remission are associated with better outcome. It is important that clinical symptoms can be due to one or any combination of the following: disease activity, thrombosis or active inflammation, drug toxicity, chronic damage due to the disease or to its treatment, or to comorbidity (i.e. infection). Yearly assessment of disease damage (with SLICC Damage Index, SDI) is recommended, although the evidence of better outcome is of low-moderate quality. However, early and late damage is associated with small to moderate increase in mortality, further damage in the future and reduced quality of life and therefore is justified to assess. Regular assessment of cardiovascular (CV) risk factors in particular as well as corticosteroid-associated adverse events is recommended, with moderate-high quality evidence for CV assessment.

The rationale and principles of treat-to-target (T2T) in SLE and how to incorporate T2T in daily clinical practice will be discussed.^{3–5}

Learning Objectives

- Describe the relevance of systematic assessment of disease activity, disease damage, comorbidities and quality of life
- Explain the current clinical practice guidelines in SLE, and define unmet needs in order to improve our daily practice
- Discuss the current state of T2T in SLE

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Neuropsychiatric (NP) involvement is one of the most complex and challenging features of systemic lupus erythematosus (SLE) encompassing the central (CNS), peripheral (PNS) and autonomous nervous system (ANS) as defined by the 1999 American College of Rheumatology standard nomenclature and case definitions. NPSLE has a negative impact on patient's quality of life and is associated with increased morbidity and mortality. The full disease burden of NPSLE is not clearly known, because robust epidemiology studies are lacking or biased by different methodology design. A realistic estimate of the prevalence of NP involvement in SLE is around fifty percent of SLE patients.

The challenge of diagnosis: As none of the NP syndromes that occur in SLE have features that are specific for SLE, determination of the correct attribution of NP events in SLE patients is a challenging but critical step in the treatment of individual patients and in performing research studies. In fact, erroneous attribution can lead to suboptimal treatment and to incorrect designation of patient groups in research studies. Approximately 30% of all NP events are attributable to SLE (NPSLE) and present most frequently around the time of SLE onset. Modern and rapidly evolving neuroimaging technologies can help clinicians in both diagnosis and follow up. A multidisciplinary expert team represents the best strategy for NPSLE.

The challenge of treatment: The main proposed pathogenetic pathways include both ischemic and neuroinflammatory mechanisms with evidence for complement and microglia activation. Following diagnosis and causal attribution, the treatment of NPSLE should be tailored to the type of NP event, the predominant putative pathogenetic mechanism, in addition to the history (acute or chronic), activity and severity of the clinical event. To treat NPSLE, in the absence of high-level evidence, it is necessary to develop pragmatic therapeutic strategies supported by expert opinion, published observational cohort data on NPSLE and extrapolation from experience with other organ system disease in SLE. To date, therapeutic options include symptomatic, anti-thrombotic and immunosuppressive agents. Therapeutic recommendations released by EULAR in 2010 and, more recently, by ACR/EULAR in 2019 are available.^{1–5}

Although neuropsychiatric manifestations of SLE have been recognised for over 100 years, unmet needs for patients with NPSLE still exist, including a lack of diagnostic biomarkers, lack of novel therapies and lack of clinical trials, which should be focused on future research agendas.

Learning Objectives

- Explain the diagnostic challenges in NPSLE with focus on the attribution and neuroimaging
- Discuss the current knowledge about the main pathogenetic mechanisms of NPSLE
- Explain the available and novel therapeutic options to treat NPSLE
- Describe unmet needs in the approach to the diagnosis and management of NPSLE

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