Case 4: Managing cardiovascular risk in a 35-year-old woman diagnosed with SLE 3 years ago

Eloisa Bonfá

A 35-year-old woman diagnosed with SLE 3 years ago, presented with malar rash, polyarthritis, thrombocytopenia, proteinuria, hypertension, normal creatinine, positive anti-dsDNA and low complement. She is a non-smoker. She was treated with MMF, prednisone, HCQ, anti-hypertensive with complete renal response within 6 months and sustained remission since then.

She currently has no complaints or features of lupus. Her BMI=24Kg/m2, BP 120/80 on MMF (1.0 g/day) and an ACE inhibitor. She self-discontinued HCQ 1 year ago. Presently her labs are normal, except for TC 168 mg/dL; LDL 120 mg/dL; HDL 30 mg/dL, TG 90 mg/dL.

Discussion Point

- What do you consider the most appropriate management for cardiovascular risk for this patient?

1. Low-risk patient (target LDL <100 mg/dL): Start on low dose statin and other measures (lifestyle intervention, smoking cessation, diet orientation, blood pressure control and check contraceptive drugs/hormonal replacement therapy).
2. Low-risk patient (target LDL <100 mg/dL): Start on low dose statin, HCQ and other measures.
3. Low-risk patient (target LDL <100 mg/dL): Start on HCQ 3 months and other measures if no improvement, add low-dose statin.
4. Low-risk patient (target LDL <100 mg/dL): Start on HCQ 3 months and other measures. Check for hypothyroidism. If no improvement, add low dose statin.
5. Low risk patient (target LDL <100 mg/dL): No need for specific therapy for dyslipidemia. Lifestyle intervention, smoking cessation, diet orientation, blood pressure control and check contraceptive drugs and hormonal replacement therapy. If no improvement, add low dose statin.

Learning Objectives

- Describe the present burden of atherosclerotic vascular disease in SLE
- Explain the occurrence of subclinical and preclinical disease
- Discuss approaches for better control of risk factors for the future including hypertension and hyperlipidemia
- Describe possible beneficial effects of antimalarials

17 FERTILITY AND PREGNANCY ISSUES IN PATIENTS WITH LUPUS NEPHRITIS

[1]Liz Lightstone, [2]Angela Tincani, [3]Sandra Navarra. *[1]Imperial College London, UK; [2]University of Brescia, Italy; [3]University of Santo Tomas, Manila, Philippines*

10.1136/lupus-2020-la.17

Case 1: Pre-pregnancy counselling for women with lupus nephritis

Liz Lightstone

An African Caribbean woman was diagnosed with systemic lupus erythematosus (SLE) in 2009 aged 21 years. At that time, she had severe Class IV lupus nephritis (LN) with crescents and acute kidney injury and was treated with steroids and cyclophosphamide. She achieved good clinical remission and was maintained for several years on low-dose prednisolone, mycophenolate mofetil (MMF), hydroxychloroquine (HCQ) and irbesartan. Antiphospholipid and lupus anticoagulant were negative, she was anti Ro positive with no extra renal manifestations.

She attended for pre-pregnancy counselling in July 2017 aged 29 years. She was off steroids but still taking MMF and irbesartan. Her creatinine was 97 umol/l with eGFR 70.8 mls/min (59 mls/min not corrected for race) and she had no proteinuria. Her labs at that time were ANA 1:320; dsDNA titer 10 units/ml; C3 normal; and C4 low 0.13 g/l. The counselling addressed fertility (in light of previous cyclophosphamide), contraception, as well as medicines management; in particular, what to stop (i.e. MMF and irbesartan), what to continue (i.e. HCQ) and what treatments might be added (i.e. azathioprine, aspirin, folic acid). The evidence base for advice regarding timing, risks to her and to the baby came from the PROMISSE study, metanalyses and the Italian series (Moroni G *et al*). Her key risk factors for adverse pregnancy outcomes were being non-white, on an antihypertensive, and having anti Ro antibodies but she was in a good remission from her lupus. I advised her to continue to use contraception whilst weaning off her MMF and to consider trying to conceive, if all remained well, when off her MMF for at least 3 months.

She presented pregnant in December 2017 but had not stopped either her MMF or irbesartan, despite the advice to do so back in July that year. We discussed the risks to the baby of first trimester exposure and she declined termination. Coincident with the pregnancy, she was serologically more active, and became symptomatic with joint pains, rising creatinine and hypertension and developed significant proteinuria. During the workshop we will discuss the pros and cons of renal biopsy in pregnancy and how we managed a really major flare. Also, the difficulty of diagnosing pre-eclampsia in a patient with active LN. The pregnancy was ultimately successful. She was treated with rituximab and MMF post-partum and again advised regarding contraception. The story continues in 2020!

Case 2: Lupus nephritis in pregnancy

Angela Tincani

Maria is a 39-year-old woman who consulted at 21 weeks of gestation for the sudden occurrence of proteinuria (3.8 g) during a previously uncomplicated pregnancy with normal fetal growth.

She had a history of undifferentiated connective tissue disease, diagnosed in 1997, because of thrombocytopenia (48,000 platelets per microliter) and Raynaud's; positive ANA, anti U1RNP. She was successfully treated with corticosteroids, which were stopped in 2000. She has had Hashimoto thyroiditis, treated with levothyroxine since 2000.

In 2014 she had vaginal delivery of a female baby at 40 weeks of an uncomplicated pregnancy without any treatment. In 2016 and 2017 she had three early miscarriages at 8, 7 and 9 weeks.

During our first evaluation (29th March 2019), she presented acrocyanosis and modest feet oedema. Proteinuria was 4 g with normal renal function; positive ANA and anti Sm/RNP; low titer anti ds DNA; positive anti-cardiolipin and anti β2 glycoproptein1 IgG.

In the last week she was treated with prednisone 50 mg/day plus low dose aspirin (LDA) and low molecular weight heparin (LMWH).

Azathioprine (AZA) was added, but because of increasing proteinuria (9.8 g) one week later she was admitted to the Obstetric Department. She was given three small pulses of

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

[1]Zahir Amoura, [2]Richard Furie, [3]David Isenberg. *[1]Pitié-Salpêtrière University Hospital, Paris, France; [2]Zucker School of Medicine at Hofstra/Northwell, New York, USA; [3]University College London, UK*

10.1136/lupus-2020-la.18

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

Alexandre Voskuyl. *Amsterdam University Medical Centers, Netherlands*

10.1136/lupus-2020-la.19

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