17 MANAGEMENT OF NEUROPSYCHIATRIC SLE

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10.1136/lupus-2022-la.17

Case 1: A 38-year-old woman with NPSLE

A 38-year-old patient presented for a follow-up consultation for her systemic lupus erythematosus (SLE). She was on 5 mg/day of prednisone and hydroxychloroquine 400 mg/day. She describes headaches that have been present for a month. On examination her pulse rate was 80 BPM, BP 122/74mmHg and temperature: 37°C. Neurological examination was normal. Respiratory, cardiovascular, joint and skin examinations were normal.

Discussion Points: What course of action do you propose?

Case 2: A 31-year-old woman with SLE and acute psychosis

A 31-year-old woman hospitalised in January 2022. She has had a medical history of SLE diagnosed in December 2018. Malar rash (acute cutaneous lupus), photosensitivity, diffuse alopecia, oral ulcerations, bilateral pleurisy, proteinuria with a kidney biopsy showing Class IV glomerulonephritis according to ISN classification, positive anti-double stranded DNA antibody test (Farr assay 78 UI; N< 9 UI), low C3 fraction, positive lupus anticoagulant, negative anticardiolipin antibody ELISA (IgG and IgM), negative anti-B2GP1 ELISA (IgG and IgM). She had no thrombotic or obstetrical history.

She first received three pulses of methylprednisolone (1000 mg each) followed by oral prednisone 0.5 mg/kg/day + mycophenolate mofetil (MMF) 2 g/day + ACE inhibitors. Steroids were tapered to 5 mg/day at 6 months. Daily proteinuria decreased to 1 g at Month 3 and 0.5 g at Month 6. C3 returned to normal level at Month 6. Steroids were stopped at Month 24 and hydrocortisone 20 mg/day was given instead. MMF 2 g/day was decreased to 1 g/day in September 2015.

She was hospitalised in January 2022 for altered sleep-wake cycles, hyperactivity, intense anxiety, ideas of persecution and auditory hallucinations. She had no arthritis and no mucocutaneous manifestation. Her physical examination was normal with no neurological abnormalities. Laboratory test showed: normal red and white blood cell and platelet counts; creatinine: 69 µmol/L; proteinuria: 0.2 g/L, Urine tests were sterile with no haematuria, creatininuria: 8.9 mmol/L = ratio 0.02 g/mmol; albuminemia: 43 g/L; C reactive protein: <5 mg/L. Farr assay 18 UI; N< 9 UI, normal C3 fraction. At that time, she was treated with prednisone 5 mg/day + MMF 1 g/day. A diagnosis of acute psychosis given done by the psychiatrist.

Learning Objectives

- Describe how to manage headaches in patients with SLE
- Describe how to manage myelitis in patients with SLE
- Describe how to manage psychiatric manifestations in patients with SLE

18 MANAGEMENT OF PREGNANCY IN SLE AND APS

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10.1136/lupus-2022-la.18

Case 1: A pregnant woman with SLE and APS

JB is a Caucasian female (born in 1978), smoker; she consulted us in 2003 (25 years old). She complained frequent episodes of lipothymia and dizziness, reported arterial hypertension diagnosed 3 years before that was successfully treated. In the last year, she had been taking oral contraceptive pill. The physical examination was normal, but livedo reticularis was noted. Her blood tests showed: thrombocytopenia 75,500/mm^3, prolongation of aPTT, hypergammaglobulinemia (25%); positive ANA, anti Ro/SS-A, low titer anti DNA; mild reduction C3 and C4. She tested positive for lupus anticoagulant, anticardiolipin and anti-beta2-GPI. Renal arteries ultrasound was normal; echocardiogram showed mild mitral insufficiency with thickened leaflets, and ENT consultation was without pathological findings. At brain MRI, multiple hyperintense foci in white matter compatible with ischemic lesions were present in T2.

Her diagnosis was probable antiphospholipid syndrome (APS) with lupus-like disease. She was managed with sun protection 50+, she stopped smoking and oral contraceptive, and started low-dose aspirin (LDA), whilst continuing anti-hypertensive therapy.

She returned to clinic in 2005, 12 gestational weeks pregnant, she was already on LDA and folic acid. The treatment was adjusted with the addition of prednisone 5 mg/day and enoxaparin at prophylactic dose. Unfortunately, intrauterine death occurred at 14 gestational weeks; placenta histology showed multiple infarctions. The diagnosis of APS with lupus-like disease was made. She was discharged with hydroxychloroquine (HCQ) 200 mg/day, LDA and prednisone 5 mg/day.

One year later, the patient returned to the clinic 6 gestational weeks pregnant. Low molecular weight heparin at prophylactic dose was added to the treatment and prednisone was increased at 10 mg/day.

In December 2005 at 30 gestational weeks, urgent caesarean section was performed for pre-eclampsia (proteinuria 5310 mg/24 h) evolved in HELLP syndrome.

In the following years she was treated with HCQ, low dose prednisone (5 mg/day), LDA, anti-hypertensive therapy and supplementation with vitamin D, folic acid and iron. Proteinuria progressively decreased (<500 mg/24 hours) and GFR reduced to 55 ml/min. She was persistently anemic (Hb 10 g) due to metrorrhagia and remained thrombocytopenic (60,000/mm^3). The baby was healthy and grew regularly.

In 2013 (aged 35-years-old) she was doing well, and she returned to the pregnancy clinic because she wanted another baby. After a joint consultation with the gynecologist and rheumatologist, she was discouraged to start a new pregnancy because of her history. In 2015 malar rash was observed together with evolution of her serology. The diagnosis of SLE with APS was made and she was started on belimumab.

Case 2: A 31-year-old woman

RI is a Caucasian female, that consulted us in 1995 at 31 years old.

She reported a pregnancy complicated by preeclampsia at 28 gestational weeks, 6 months before; a female baby of 850 g was born and, unfortunately, died after one day. Subsequent investigations revealed the presence of IgG anticardiolipin antibodies at high titre and lupus anticoagulant. At the time of our consultation, she reported frequent vision changes in the days before her monthly periods. She also reported photosensitivity since her 20s. Physical examination was normal with a normal blood pressure. Blood tests revealed low

titre ANA and triple antiphospholipid antibody positivity. Treatment with low dose aspirin (LDA) was started.

In 1996 she came to see us at 6 weeks of gestation, she was already on LDA and folic acid. Prednisone 5 mg was added. The pregnancy was carried on without complications. At 38 gestational weeks. Caesarean section was performed, because of PROM, and a female baby was born with a birthweight of 3100 g.

The patient, living in another town, came for a follow-up visit in 1998. She reported some episodes of dizziness and diplopia. Physical examination and blood pressure were normal. The serology was repeated and resulted unchanged. Hydroxychloroquine and LDA were prescribed.

One year later (1999), the patient presented at the beginning of a second pregnancy. The clinical situation was substantially unchanged, we decided to continue HCQ and LDA with the addition of 5 mg prednisone and folic acid. At 17 gestational weeks, HCQ was withdrawn because of an itching diffuse rush. At 35.3 gestational weeks, blood pressure started to rise, and a Caesarean section was performed at 36 gestational weeks. A healthy female baby was born, with a birthweight of 2510 g.

Five days after delivery, the mother developed slurred speech and right hemiplegia suddenly occurred while she was taking unfractionated heparin 0.2 ml twice a day. Brain CT scan revealed and ischemic area in lentiform nucleus, caudate nucleus and left posterior limb of the internal capsule.

Learning Objectives

- Explain the importance of counselling in order to improve the pregnancy outcome
- Describe risk factors for recurrence in patient with obstetric APS, including triple antibody positivity as an important risk factor driving treatment adjustments
- Discuss why it could be necessary discourage a patient to start a new pregnancy
- Describe the possible effective treatments of obstetric APS
- Explain the importance of focusing on the post-partum period and to the possible occurrence of serious complications linked to the presence of antiphospholipid antibodies

19 MANAGEMENT OF PREGNANCY IN SLE AND APS

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10.1136/lupus-2022-la.19

Since systemic lupus erythematosus (SLE) primarily affects women of childbearing age, therefore pregnancy assumes great importance for these patients. Accordingly, proper pre-conception counselling is highly relevant to management and can set the stage for a more favourable outcome.

Women of childbearing age with SLE should be advised of medications safe to continue during pregnancy and told that disease should be in remission or at least not acutely active. Hydroxychloroquine is strongly recommended for all women during pregnancy despite a concern raised regarding a very slight increase in malformations, which was not confirmed in a subsequent study. Low dose aspirin to prevent pre-eclampsia should be highly reinforced. The issues related to pregnancy outcome for the maternal-fetal dyad comprise three components, maternal, placental and fetal with the latter clearly influenced by the two former. With the completion of a large prospective US based study,[1] factors associated with poor pregnancy outcomes in patients who are generally stable at the time of conception have emerged and include being Hispanic or non-white, taking blood pressure medications, low platelet counts, active disease even if stable, and the presence of a lupus anticoagulant. In the absence of these factors, pre-eclampsia still occurs at a greater frequency than in the otherwise healthy population, with estimates of about 9%, as does small for gestational age at about 10%. In patients with no risk factors at baseline, the adverse pregnancy outcome rate is about 8% and fetal/neonatal mortality 3.9%. In patients who are either lupus anticoagulant (LAC) positive, or LAC negative but non-white and treated with anti-hypertensives, the adverse pregnancy outcome rate approaches 60%; fetal/neonatal mortality at 22%. In general, patients with prior kidney disease but in remission do well without an increased risk of flare with *de novo* kidney disease being quite uncommon with risk under 3%. Severe flares in patients stable at conception approach less than 3% at any time during pregnancy and postpartum, and approximately 10% intrapartum and 25% postpartum for mild/moderate flares (most not requiring intervention).[2]

Promising biomarkers, which may identify women early in pregnancy at risk for poor outcomes, include angiogenic factors and alternative and terminal complement activation factors. Turning to neonatal lupus, newer research supports the contribution of Type I interferon in the pathogenesis. A prospective study supports the use of hydroxychloroquine to decrease the recurrence of congenital heart block in half.[3] Data suggest that low titre anti-SSA/Ro antibodies do not confer risk of fetal cardiac injury but defining low and high titres in commercial laboratories is not well established since many do not provide a broad range of values. New approaches to surveillance are being addressed leveraging home heart rate and rhythm monitoring by the mothers.[4] The NIH has launched a recent US and Canada based study to evaluate whether the titre of anti-SSA/Ro 60 or 52 antibodies confers higher risk and if home Doppler monitoring can identify a transition period of conduction slowing that is reversible with dexamethasone and IVIG if administered within 12 hours of a Doppler abnormality confirmed by echocardiogram. Overall, the landscape for women with lupus contemplating pregnancy is favourable after achievement of stability or remission prior to conception. Prevention of reversible cardiac damage in those women with SLE and anti-SSA/Ro antibodies may be on the horizon.

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Case 1: A 36-year-old-white female

A 36-year-old white female who has occasional dry eyes, no dry mouth, and no other symptoms was tested for anti-SSA/Ro antibodies, given the dry eyes and a family history of