17 MANAGEMENT OF NEUROPSYCHIATRIC SLE

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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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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