Case 3: 19-year-old female (AM) with 1-month history of malar rash, polyarthritis, fatigue, fever

David Isenberg

Patient AM presented aged 25 years with 6 month history of Raynaud's and a 2 week history of malar rash, polyarthritis, fatigue and fever. On admission, her laboratory tests revealed ANA–1:640 and a positive anti dsDNA antibody. On Day 3, she had complained of proximal weakness and CK >3,000 iu/L. She was due to have a muscle biopsy on Day 7 but presented with severe abdominal pain and her amylase was >3,000 IU/L, she was presumed to have pancreatitis. Her condition deteriorated alarmingly between Days 8 and 10, when she developed adult respiratory distress syndrome and died.

Discussion Point

- What did the post-mortem show?

Case 4: 41-year-old patient (LC) returns from holiday in the Dominican Republic of Congo and presents with ↑+ proteinuria and fever

David Isenberg

Patient LC had an Indian father and West Indian mother. At age 18 years she presented with arthritis and hair loss. Laboratory results revealed: postive ANA 1:2560; dsDNA, anti-RNP, ab and LAC. She was treated with steroids and azathioprine.

Aged 23 years, she went to see her GP, presenting with a cold blue fingers; 5 hours later she collapsed; several of her fingers were gangrenous. She was given prostacyclin, steroids, and cyclophosphamide and improved.

Age 31 years she presented with heavy proteinuria, biopsy revealed Class IV nephritis and she was given IV cyclophosphamide. For the following 8 years (age 32–40 years) she was managed on steroids and azathioprine, before stopping treatment. Aged 40 years, she experienced renal relapse which improved with prednisolone and azathioprine. Aged 41 years, she went on holiday to the Democratic Republic of the Congo, and returned with 4+ proteinuria, fever, and she felt unwell. She had a further renal biopsy.

Discussion Point

- What did the biopsy reveal?

Learning objectives

- Describe the causes of infections in SLE
- Discuss prevention of infections in SLE
- Explain management of difficult infections in SLE

03 FAMILY PLANNING AND SLE: FROM CONCEPTION THROUGH CHILD-BEARING

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Case 1: Counselling before pregnancy

Angela Tincani

A 28-year-old lady (Monica) with previous lupus nephritis makes contact to plan a pregnancy.

Previous history: 2011 (aged 33 years): Class IV lupus glomerulonephritis: NIH protocol induction treatment: monthly intravenous cyclophosphamide for 6 months (cumulative dose 8 grams) and 1 mg/kg/day prednisone tapered to 5 mg/day in 6 months. Following maintenance treatment with mycophenolate mofetil (MMF) 1 g twice a day.

She is currently taking low dose steroids and MMF as maintenance treatment. With these drugs her nephritis has been in complete remission for 2 years.

The management of the lady before pregnancy included: stop MMF and switch to azathioprine 100 mg/day (~2 mg/kg/day) and monitor disease activity for at least 3–6 months with monthly evaluation of renal parameters and bimonthly anti-dsDNA, C3, C4; hydroxychloroquine 200 mg every other day with 400 mg; low-dose aspirin; folic acid and Vitamin D. At positive pregnancy test: stop ramipril; start low molecular weight heparin (prophylactic dose) and calcium.

Discussion points

- The effects of pregnancy on disease: Risk stratification based on SLE activity and presence of concomitant risk factors (arterial hypertension, obesity, smoking, etc)
- The effects of the disease on pregnancy outcome: A complete serological profile is needed to assess the risk for the fetus (anti–Ro/SSA antibodies, antiphospholipid antibodies).
- Drugs that need to be withdrawn prior to conception due to teratogenicity and drugs to be added as preventative measures.

Case 2: SLE flare in pregnancy

Angela Tincani

A 34-year-old pregnant woman (Cinzia) with systemic lupus erythematosus (SLE) is referred to the Pregnancy Clinic for Systemic Autoimmune Diseases because of a severe mucocutaneous flare occurred in the second trimester of pregnancy.

Previous history 1990 (age 17 years), diagnosis of SLE (in a different institution) based on polyarthritis, skin rash, fever, anti-nuclear antibodies and anti-dsDNA at high titre; autoimmune Thyroiditis with hypothyroidism; she was treated with high dose oral steroids, azathioprine; hydroxychloroquine stopped after 3 months because of acute skin rash (after intense sun exposure).

Maintenance treatment Methylprednisolone 4 mg/day; azathioprine 50 mg/day (~1 mg/kg/day); levothyroxine.

The patient had withdrawn her lupus medications at the beginning of pregnancy and started low-dose aspirin following her gynecologist's advice and did not consult her rheumatologist during pregnancy.

She was referred to our pregnancy clinic in May 2007 (age 34 years, at 18 weeks gestation) because of a severe mucocutaneous flare consisting of diffuse skin rash, chilblain lupus at the hands and feet, multiple oral aphthous with erosions of the palate. Very active SLE serology was also recorded.

She was closely monitored during the 2nd and 3rd trimesters and treated with methylprednisolone pulses followed by medium-high dose prednisone and azathioprine (for topical dermatological treamtent). Her disease was managed satisfactorily. She delivered a female baby at term.

Discussion points

- Decreasing the risk of lupus flares in pregnancy
- Treating a severe mucocutaneous lupus flare in the second and third trimester

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- Managing the risks and benefits of corticosteroids during pregnancy

Case 3: Anticoagulants during pregnancy

Angela Tincani

A 27-year-old female (Elena) came to our pregnancy clinic in 2005 because she wanted to plan a pregnancy. Her SLE was in complete longstanding remission, she had normal blood pressure, normal body weight (50 kg, BMI=20) and she had quit smoking one year earlier.

Previous history: Her SLE was diagnosed in 1996 (age 18 years) in another institution based on glomerulonephritis, polyarthritis, skin rash, anti-nuclear antibodies and anti-dsDNA at high titer. She had been treated with high dose oral steroids, cyclophosphamide, methotrexate, azathioprine, and hydroxychloroquine. She had been diagnosed with deep venous thrombosis in the right lower limb in 2000 (age 22 years) when she was found to be triple antiphospholipid antibody positive and started in warfarin.

Maintenance treatment: Prednisone 5 mg every other day; hydroxychloroquine 200 mg/day and warfarin INR target 2–3. In 2004 she underwent a voluntary abortion at 8 weeks for fetal exposure to oral anticoagulation. She was told that the fetus might suffer from malformations.

We advised the patient to maintain warfarin and stop it as soon as the pregnancy test was positive (perform an early test, at the very first day of menstrual delay) when low molecular weight heparin should be started. The treatment during pregnancy included enoxaparin (4000 UI twice a day), low-dose aspirin, prednisone 5 mg every other day, hydroxychloroquine and folic acid.

Elena delivered a healthy baby girl at 38 weeks of gestation. Heparin was restarted 12 hours after delivery and the treatment was switched back to warfarin one week later. The possibility of breast feeding was discussed.

Three months later she asked for contraception: According to the risk profile and the patients desire progesterone-releasing intrauterine device was chosen.

Discussion points

- How to administer anticoagulant treatment during pregnancy and puerperium
- Risks and benefits of breast feeding
- Problems related to contraception in SLE patients

Case 4: Unplanned pregnancy

Ricard Cervera

A 28-year-old African woman with a 15-year history of SLE was admitted at the Hospital Clinic of Barcelona at Week 38 of pregnancy because of high blood pressure.

She had been diagnosed of lupus nephritis at the age of 14 years and suffered several flares since. When she get married at the age of 26 year, she wanted to become pregnant and was referred to a preconception counselling clinic in order to be advised regarding pregnancy in SLE. The attending physician provided information regarding the need to wait until lupus nephritis was under control and no potential teratogenic drugs were used. However, a positive test for pregnancy was detected just 2 weeks later.

Discussion points

- How to deal with an unplanned pregnancy
- Managing high blood pressure during pregnancy

Learning objectives

- Discuss principles and strategies for preconception counselling in patients with systemic lupus erythematous
- Recognise clinical and laboratory features which help assess pregnancy complications in SLE patients
- Describe strategies for the management of pregnancy complications in SLE patients

04 CHALLENGES IN LUPUS NEPHRITIS

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Case 1: 27-year-old female with a 5-year history of SLE

Richard Furie

A 27-year-old female with a 5-year history of systemic lupus erythematosus (SLE) was admitted to the hospital because of confusion and fever. Past manifestations of SLE included polyarthritis, rash, recurrent episodes of pericarditis, and anaemia (but no nephritis). A flare 2 months prior to admission, consisting of pericarditis, fever, hypocomplementaemia, and a 2-fold rise in anti-DNA antibodies, was successfully treated with prednisone 40 mg/day; prednisone was subsequently tapered. At the time of admission, medicines included hydroxychloroquine 400 mg/day, prednisone 15 mg/day, calcium, and a vitamin.

The patient was given broad-spectrum antibiotics for the treatment of sepsis and/or bacterial meningitis. Methylprednisolone 60 mg/day was also administered. However, the patient's mental status worsened, and she became comatose. All cultures were sterile. Her creatinine, which was 0.6 mg/dL at baseline, rose 3-fold.

The impression was that of a flare of SLE complicated by anaemia, thrombocytopaenia, nephritis and CNS disease. 'Pulse' steroids were administered for 3 days without subsequent improvement. Intravenous gamma-globulin failed to improve the thrombocytopenia, and her creatinine continued to rise.

Discussion points

- Diagnosis and treatment of thrombotic microangiopathy (TMA)
- Proposed modifications to the classification of lupus nephritis

Learning objectives

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

Case 2: 16-year-old male with active SLE, trace protein and hematuria

Dimitrios Boumpas

A 16-year-old male (60 kg) presents with active SLE (SLEDAI 10). He has active serology with low C3 and C4, anti-DNA is positive at low titer. Normal Cr, albumin and HCT. Urinalysis shows trace protein (300 mg/dL) with 510 RBCs in the urine. He was treated with hydroxychloroquine and prednisone 20 mg/day and was referred to you 4 weeks later. His SLEDAI is now 4 (rash, serology) and urinalysis shows trace protein and hematuria.

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