

dsDNA and LAC positivity. The antiepileptic therapy was initially modified and then, as no more crises were present, interrupted. However, anemia, leukopenia, hypocomplementemia, ANA and anti-dsDNA persisted, and the diagnosis of idiopathic systemic lupus erythematosus (SLE) was made. After treatment with hydroxychloroquine and low dose prednisone the girl clinically improved and her laboratory results normalized. This case report is suggestive of the complexity in differentiating SLE and DILE and underlines the importance of a long and careful follow-up.

#### Discussion Points

- Medications that can trigger lupus symptomatology
- Triggers of lupus or of lupus-like disease (autoimmunity in general) with TNF inhibitors used for arthritis
- Risks of prescribing such medications (i.e. anti-TNF) in patients with inflammatory arthritis and pre-existing autoantibodies or a family history of autoimmune disease

#### Case 5: Bleeding and thrombosis in juvenile systemic lupus erythematosus

##### Rolando Cimaz

A young girl with immune thrombocytopenic purpura was also found to have antiphospholipid antibody syndrome. This case describes the complexity of therapeutic management linked to the risk of bleeding and thrombosis.

Irene is 15-year-old. In recent hours she has experienced intermittent claudication and lower right limb pain. Her physical exam reveals swelling and tenderness of right calf, pain to compression and mobilization of right foot. Laboratory tests reveal WBC 21.610 (N 77%); Hb 13.1 g/dl; PLT 91000/mm<sup>3</sup>; aPTT 48.4 sec, PT 99%; fibrinogen 236 mg/dl; D-Dimer 0.59 mg/L. Ultrasound revealed thrombosis.

Past medical history showed that 8 months before she had suffered from severe metrorrhagia. Laboratory tests had shown: PLT 7000, Hb 10.2 g/dl, Coombs direct test +, PT 1.18, aPTT 76 sec; IgM e IgG anticardiolipin +. Therapy consisted in intravenous immunoglobulin (two infusions) and then oral steroids. Five relapses occurred, and laboratory showed: ANA+ (1:160), ENA -, anticardiolipin -, C3 85, C4 7.5, LAC +. Repeat laboratory test showed ANA+, ENA-, anticardiolipin +, anti-b2 glycoprotein -, LAC +.

#### Discussion Points

- For thrombosis Heparin 6000U/x2/day. For how long? And for thrombocytopenia? Oral steroids (2 mg/kg/day); Mycophenolate mofetil (750 mg/m<sup>2</sup> x 2/day). But despite this therapy, she relapsed. So > rituximab 750 mg/m<sup>2</sup>/2 weeks. For SLE hydroxychloroquine was given.

#### Learning Objectives

- Distinguish between idiopathic and drug-induced SLE
- Describe the treatment of APS in children
- Discuss treatment options for hematologic SLE

## Workshop

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### NEUROPSYCHIATRIC SLE

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#### Case 1: A 43-year-old woman with focal cerebral involvement in SLE

##### Ricard Cervera

A 43-year-old woman, with a 1-year history of systemic lupus erythematosus (SLE) was admitted to the Hospital because of paresthesia in left arm and speech difficulties. Diagnosis of SLE had been made at the age of 42 years based on discoid lupus lesions, photosensitivity, arthritis, oral ulcers and detection of antinuclear, anti-dsDNA and anti-Ro/SS-A antibodies. Other anti-ENA and antiphospholipid antibodies (lupus anticoagulant and anticardiolipin antibodies) were negative at diagnosis. Relevant past history included: (i) Diagnosis of hypothyroidism due to autoimmune thyroiditis at the age of 22, treated with levothyroxine; (ii) two pregnancies at the ages of 29 (spontaneous abortion at Week 8) and 31 years (normal pregnancy). Lupus nephritis was diagnosed nine months later, and kidney biopsy disclosed a Class IV-S-A/C (Activity Index: 6/24; Chronicity Index:1/12). Methotrexate was discontinued and lupus nephritis induction of response therapy was started at Day Hospital with three daily pulses of methylprednisolone (1 g each) and low-dose pulse cyclophosphamide ('Euro-lupus' regimen). She was advised to continue on hydroxychloroquine (200 mg/day) and 30 mg/day of prednisone.

One week after the administration of the first pulse of cyclophosphamide (500 mg), the patient presented with numbness and paresthesia in left arm and hand as well as speech



**Abstract 21 Figure 1** T2 magnetic resonance sagittal slices showing a high intensity signal from the ponto-medullary junction to the cervical (A) and thoracic (B) medulla

difficulties and was admitted at the Emergency Room. At physical examination, dysarthria and paresis of left upper limb was confirmed. Blood pressure was 160/95 mm Hg.

#### Discussion Point

- Recognising and treating focal cerebral involvement in SLE

#### Learning Objectives

- Recognise and describe clinical, laboratory and imaging features, which help assess focal cerebral involvement in patients with SLE.
- Explain the principles and strategies for the management of cerebral manifestations in SLE.
- Demonstrate clinical awareness of potential severe cerebral complications in SLE

#### Case 2: A woman with recurrent myelitis and SLE

##### Thomas Huizinga

A woman was diagnosed with systemic lupus erythematosus (SLE) at the age of 28 years old based on arthritis, skin erythema, mouth ulcers, sunlight hypersensitivity, ANF +, anti dsDNA + and anticardiolipin antibodies. At the age of 34 she developed papillitis of both eyes with vision loss. Her MRI revealed no abnormalities and she was treated with methylprednisone 500 mg for 5 days. At the age of 35 she developed a recurrence, no neuritis bulbaris was observed and SLE was thought to be the most likely cause so she was treated with cyclophosphamide 750 mg/kg for 6 months and prednisone 60 mg for 4 weeks and then 10 mg/kg lowering every 4 weeks. After 6 months she had completely improved. At the age of 37 she gradually developed, over a number of weeks, problems urinating and sensory disturbances left thigh and right leg, physical exam revealed hypesthesia in her left leg, and feelings of a different temperature in legs compared to arms.

MRI revealed transverse myelitis at C6-Th2, compatible with SLE and she was retreated with cyclophosphamide and prednisone (figure 1). Subsequently, she did quite well for a number of years under subsequent treatment with azathioprine and cyclosporin for joint and skin involvement. At the age of 52 she developed hypesthesia of the thorax and MRI revealed a myelitis over a long span Th2-Th10 while receiving cyclosporin. A laboratory test for aquaporin 4 antibodies was negative. Combination treatment with prednisone and rituximab followed by belimumab was given, to which she responded quite well. After 2 years she developed less strength in both legs, sensibility disturbance in both legs and specifically C9 and Th7 dermatome, new miction problems and inability to defecate. MRI revealed new lesions at TH4-TH8 while a sharp reduction of previous lesions was observed; aquaporin antibodies tested positive. Based on the recurrence, rituximab maintenance therapy was given every half year. In 2020 she started to suffer from recurrent infections and her IgG levels were 5.6 mg/ml after which intravenous IgG suppletion was started and the infectious problems disappeared.

#### Discussion Point

- Diagnosis, management and recurrence of myelitis in SLE

#### Learning Objectives

- Describe the clinical presentation of myelitis and its recurrence
- Explain treatment options of myelitis
- Discuss the role of aquaporin-4 antibodies in myelitis

## Workshop

### 22 REPRODUCTIVE HEALTH IN SLE

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#### Case 1: 23 year-old-female seeking contraception

##### Lisa Sammaritano

A 23-year-old woman with systemic lupus erythematosus (SLE) who recently relocated wishes to establish rheumatology care. She was diagnosed with SLE aged 14 years with idiopathic thrombocytopenic purpura; she was treated with high dose steroid with good response and tapered off steroid within 6 months. She was well until age 20 when she presented with malar rash, inflammatory arthritis of PIPs, MCPs and knees, fatigue, and petechia following a vacation in the Caribbean. She was diagnosed with SLE with positive ANA and double stranded DNA. Her hemoglobin was 6.0 gm/dl with positive Coombs and low haptoglobin, her platelet count was 12,000. C3 and C4 were low. She was treated with intravenous 'pulse' methylprednisolone followed by high dose prednisone, azathioprine and hydroxychloroquine with good response.

She currently feels well without complaints other than mild morning stiffness in her hands and occasional malar rash. Current medications are azathioprine 125 mg daily, hydroxychloroquine 300 mg daily, and prednisone 5 mg daily. Physical exam at the visit is unremarkable and routine labs including urinalysis are normal. She has been using barrier contraception (condoms) but asks about other options. She is very worried that any hormone therapy will cause her lupus to flare again. How do you assess her, and what do you recommend?

a. Serologies show negative anti-dsDNA, normal C3C4, and negative antiphospholipid antibodies (aPL) including lupus anticoagulant (LAC), anticardiolipin (aCL) and anti-beta 2 Glycoprotein I (ab2GPI).

b. Serologies show negative anti-dsDNA, normal C3C4, and positive LAC, positive aCL IgG 68 and positive ab2GPI IgG 45

#### Discussion Point

- The best way to assess patients with SLE for safe and effective contraception, in the setting of negative and positive aPL

#### Case 2: 33-year-old seeking assisted reproductive technology

A 33-year-old woman with SLE presents for routine follow-up. You have followed her since her diagnosis at age 27 years when she presented with oral ulcers, photosensitive rash, arthritis, pleuritis, positive ANA and positive dsDNA antibody. She started hydroxychloroquine with control of her symptoms after a brief steroid taper. At age 31 she developed nephritis, Class IV and V on renal biopsy. She was treated with intravenous 'pulse' methylprednisolone, rituximab (two doses of 1 gm each) and mycophenolate 2000 mg daily. She continues on the mycophenolate 2000 mg daily, enalapril 20 mg daily and prednisone 5 mg daily. Her serum creatinine has been 1.2 mg/dl, and her dsDNA antibody is low positive with mildly low C3 and normal C4 (markedly improved from her initial