and urine anion gap positive at 2.73 meq/L, confirming the diagnosis of an underlying distal renal tubular acidosis (RTA).

Discussion Points

- Clinical significance and management of RTA in LN

Case 4: 31-year-old patient with LN and APS presents with fever, deteriorating kidney function and an ovarian mass

Liz Lightstone, Sandra Navarra

Clinical data A 31-year-old female was diagnosed with LN and APS 8 years ago presenting with Raynaud's, mononeuritis multiplex, proteinuria, pancytopenia, and pericarditis. She was given methylprednisolone pulse, cyclophosphamide, and belimumab then maintained on hydroxychloroquine, MMF, aspirin, nifedipine, iron plus folate, and calcium plus vitamin D. She developed hematologic flare a year ago which responded well to methylprednisolone pulse and two doses of rituximab. Two months ago, she presented with intermittent fever, dysuria, and cough. Laboratory results were: Hemoglobin 97 g/L, WBC 8.4 $x10^9$/L, platelet 10^9 $x10^9$/L, BUN 52.1 mmol/L, creatinine 2.73 mg/dL (eGFR 22.3 ml/min) →3.26 mg/dL (eGFR 18.1 ml/min), urine protein 2+, RBC 40–50, pus >100; urine PCR by Gene Xpert® was positive for Mycobacterium tuberculosis (TB), no rifampicin resistance. Chest radiograph showed infiltrates suggestive of miliary TB. Abdominal ultrasound showed renal cysts with calcifications, mild left ureteropelvocaliectasia from a left adnexal cystic mass measuring 8.57 cm. Exploratory laparotomy with left salpingo-oophorectomy was performed; histopathology showed caseating granuloma. She was started on an anti-TB regimen.

Discussion Points

- Causes of renal insufficiency in a patient with LN
- Renal involvement in APS

Learning Objectives

- Describe management approach to LN
- Explain special considerations in the management of LN flare during pregnancy
- Discuss further management options in refractory LN
- Discuss clinical situations which significantly contribute to morbidity in LN

Workshop

20 PAEDIATRIC SLE

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Case 1: 9-year-old female with rash and oral ulcer

Alexandre Belot

Alisson is 9 years old and came to the outpatient unit with a rash and oral ulcer. Her past medical history highlighted two invasive infections (meningitidis and pneumonia). She had no muscular weakness, no lymphoproliferative disease and no fever.

Her laboratory exams revealed: WBC: 2.5 G/L, with PNM=1.2G/L, Hb = 10 g/dl, Platelet = 135G/L, CRP = 10 mg/L. CPK and aldolase were within normal values. Serology for Epstein-Barr Virus, parvovirus B19 and measles were negative.

Autoimmune check-up showed: ANA+ 1/1280, C3 and C4 normal, anti-dsDNA negative, anti SSA+ >8. The dosage of immunoglobulins was normal (including subclasses). She further developed severe systemic disease with lupus nephritis, white matter lesions at the cerebral MRI.

Complement CH50 was dramatically decreased and her C1q level was undetectable.

Discussion Points

- Explore complement deficiency in juvenile systemic lupus erythematosus (SLE)
- Discuss monogenic SLE

Case 2: 13-year-old female with skin rash and chilblain

Alexandre Belot

Jade is 13 years old and has been recently diagnosed with juvenile SLE. Her past medical history revealed a pervasive developmental disorder with features of autism and mental delay. Her parents are first cousins. Her first symptoms were skin rash, chilblain. Following first-line therapy with hydroxychloroquine and topical steroids, she developed a polyarthritis with hepatitis and leukopenia. Abdominal sonography was normal. Autoantibodies for autoimmune liver disease (dot hepatitis) were negative. Treatment with steroids and methotrexate was introduced and effective on the joints. Further genetic exploration revealed a biallelic mutation of TREX1. Later on, she was treated with a JAK inhibitor in addition to methotrexate resulting in a positive outcome.

Discussion Points

- Type I interferon in juvenile SLE
- Interferonopathies

Case 3: 14-year-old male with haematuria and renal colic

Alexandre Belot

Michael presented a macroscopic hematuria with renal colic at the age of 14 years old. Sonography revealed a left nephromegaly without evidence of urinary tract obstruction. A CT scan showed a left renal venous thrombosis. Initial work-up identified a triple positivity for lupus anticoagulant, anti-B2 Gp-I and anti-cardiolipin antibodies. Anticoagulant therapy was initiated. dsDNA and ANA were negative and complement was normal. Six months later, the hematuria had completely disappeared, according to a urine dipstick test, but proteinuria was still present with a protein:creatinine ratio of 120 mg/mmol in the urine. A new doppler sonography and CT scan showed the absence of perfusion in the left kidney.

Considering the proteinuria, a percutaneous biopsy was performed on the contralateral kidney and histology revealed Class V lupus nephritis. Notably, autoantibodies and complement were still normal. Rituximab and ACE inhibitors were introduced, and proteinuria rapidly disappeared.

Discussion Points

- How to explore APS in children
- Management of thrombosis in pediatric autoimmune diseases

Case 4: Anti-histone antibodies: does it always mean drug-induced lupus erythematosus?

Rolando Cimaz

A South-American 14-year-old girl presented with arthralgia, weakness and alopecia. As she was under antiepileptic treatment since she was 5 years old, on suspicion of drug-induced lupus erythematosus (DILE) anti-histone antibodies were dosed and showed positive results. She presented with mild anemia, leukopenia, hypocomplementemia, ANA, anti-

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/mmc; 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^2 x 2/day). But despite this therapy, she relapsed. So > rituximab 750 mg/m2/2 weeks. For SLE hydroxychloroquine was given.

Learning Objectives

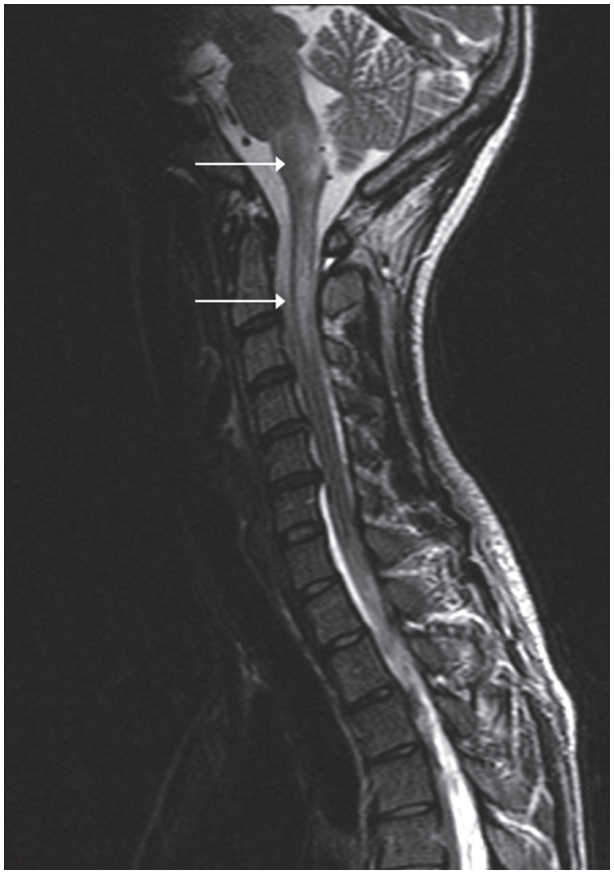
- Distinguish between idiopathic and drug-induced SLE
- Describe the treatment of APS in children
- Discuss treatment options for hematologic SLE

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

Workshop

21 NEUROPSYCHIATRIC SLE

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