face, and hands. She had been resistant to a high number of therapeutic agents including hydroxychloroquine, chloroquine, methotrexate, isotretinoin, and cyclophosphamide for several years and had developed various side effects. Moreover, the long-term medication for SLE had not influenced her skin lesions in the past months. In December 2009, the patient received alitretinoin, which was administered orally with a daily dose of 30 mg. The patient showed continuous improvement of the discoid skin manifestations over a period of 5 months and finally a total clearance of most lesions. No serious adverse events were recorded during treatment with alitretinoin; however, the patient experienced recurring headache, which was successfully treated with nonsteroidal anti-inflammatory drugs. Therefore, alitretinoin was reduced to a daily dose of 10 mg.

The vitamin-A derivative, alitretinoin, has been approved for use in severe chronic hand eczema unresponsive to treatment with potent topical corticosteroids. This case suggests that alitretinoin could also be an effective alternative in the treatment of cutaneous manifestations in SLE; however, randomized controlled trials are needed to prove the efficacy and evaluate the safety of alitretinoin in this disease.

Discussion Points

- Treatment of therapy-resistant cutaneous manifestations in SLE
- Therapeutic guidelines in cutaneous lupus erythematosus

Learning Objectives

- Explain different possible skin manifestations in patients with SLE
- Discuss complications, differential diagnosis with allied diseases and treatment approaches
- Describe the spectrum of manifestations in cutaneous lupus, including: ACLE, CCLE, SCLE and ICLE
- Explain the therapeutic guidelines in cutaneous lupus
- Describe preventive strategies in cutaneous lupus including photoprotection
- Describe topical treatment options in cutaneous lupus
- Discuss common and experimental systemic treatment options in cutaneous lupus

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Workshop

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LUPUS NEPHRITIS

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Case 1: 35-year-old patient with lupus nephritis presents with anasarca, hypertension and renal insufficiency at her 18th week of pregnancy

Liz Lightstone, and Sandra Navarra

Clinical presentation A 35-year-old female was diagnosed lupus nephritis (LN) 7 years earlier with kidney biopsy showing LN Class IV-G activity 7, chronicity 3. She received methylprednisolone pulse and was thereafter maintained on prednisone, hydroxychloroquine (HCQ), mycophenolate mofetil (MMF)

and calcium plus vitamin D. Her condition was 'stable' despite erratic follow-up and poor adherence. Two months ago, while on prednisone 10 mg/day, she consulted at clinic because of anasarca and hypertension at Week 18 of her 4th pregnancy. Obstetric history included two spontaneous abortions at 2 months gestation in 2013, 2015 and a successful term pregnancy in 2018 with a healthy baby girl delivered by C-section. Laboratory tests showed: Hemoglobin 83 g/L, hematocrit 0.24, WBC 6.97, platelets 270, creatinine 2.29 mg/dL, ↓ C3 0.20 g/L, ↑anti-dsDNA 1152.12 IU/mL, urine albumin 4+, RBC 90–95/hpf with dysmorphic RBCs, WBC 10–12/hpf, hyaline and granular casts. Anticardiolipin, lupus anticoagulant, anti-Ro and anti-La were negative.

Clinical course Prednisone was increased to 40 mg/day and she was started on methyldopa, azathioprine, HCQ, calcium plus vitamin D, iron plus folate, and aspirin 80 mg/d. A week later, she was admitted due to cough, dyspnea, orthopnea and low-grade fever. Chest radiograph showed hazy densities on both lung fields. Laboratory tests showed: Hemoglobin 74 g/L, hematocrit 0.22, WBC 17.20, platelets 433. Urine albumin 4+, RBC 18–22/hpf, WBC 10–15/hpf, hyaline, granular and waxy casts, urine total protein 822.40 mg/dL, urine creatinine 151.08 mg/dL, urine protein-creatinine ratio 5.44; BUN 62.6 mg/dL, creatinine 2.90 mg/dL, total protein 5.01 mg/dL, albumin 2.66 g/dL, Na 132 mmol/L, K 5.69 mmol/L, phosphorus 6.07 mmol/L, and iCa 1.19 mmol/L; SARS-CoV2 test was negative. She was started on antibiotics and received red cell transfusions; regular hemodialysis was initiated due to pulmonary congestion and metabolic acidosis.

Discussion Points

- Distinguish pre-eclampsia from LN flare
- Management of severe LN flare with renal insufficiency during pregnancy including the role of plasma exchange

Case 2: 22-year-old female with refractory LN despite immunosuppressive regimens and rituximab

Liz Lightstone, Sandra Navarra

Clinical data A 22-year-old female presented with anti-phospholipid syndrome (APS) and LN at age 8. She had completed cyclophosphamide induction therapy, had been adherent to MMF (plus tacrolimus for 18 months), and received eight doses of rituximab. Renal histopathology (2008, 2016, 2019) shows persistent lupus nephritis ISN/RPS Class IV, varying activity and chronicity indices. She would attain partial renal remission for a few months, but never achieved complete remission. Blood pressure and renal function remain within normal range.

Discussion Point

- Updates in the management of LN

Case 3: 35-year-old male with LN presents with persistent hypokalemia

Liz Lightstone, Sandra Navarra

Clinical data A 35-year-old male with LN ISN/RPS Class IV – S, activity 11 and chronicity 4, presented with nephrotic-range proteinuria, hypertension, and impaired renal function. Renal ultrasound was normal without calcinosis. He received prednisone, MMF, HCQ, anti-hypertensives, and calcium plus vitamin D. He had persistently low levels of serum potassium (K) 2.4–3.8 meq/L, and serum bicarbonate (HCO₃) 11–20 mmol/L despite K and HCO₃ supplementation. Urine pH was 6.0, urine sodium (Na) 80 mmol/L, urine K 20.63 mmol/L, urine chloride (Cl) 97.90 mmol/L. Serum anion gap was normal,

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 $8.4 \times 10^9/L$, platelet $10^9 \times 10^9/L$, BUN 52.1 mmol/L, creatinine 2.73 mg/dL (eGFR 22.3 ml/min) $\rightarrow$ 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 ureteropelvic ectasia 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

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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 (meningitis 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-