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17 LUPUS TREATMENT IN THE NEXT DECADE: THE NEXT DECADE IS UPON US

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There is little question that our colleagues in rheumatology who treat patients with rheumatoid or psoriatic arthritis have outdone us 'lupologists' in bringing new therapies to the community. Biologics have truly been transformative for those patients with inflammatory arthritis. We are now starting to see the same successes in lupus. As we celebrate the 10th anniversary of the FDA approval of belimumab, the lupus community recently witnessed the approval of two drugs, belimumab and voclosporin, for the treatment of lupus nephritis. In addition, several Phase 2 studies yielded favorable results and will be progressing to Phase 3.[1–5] Drug development activity is currently unprecedented, and there is no doubt that research advances will improve outcomes and ensure brighter futures for our patients with systemic lupus.

Learning Objectives

- Describe unmet needs in SLE treatment
- Discuss biologic targets for SLE drug development
- Explain recent clinical trial results

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Workshop

18 CUTANEOUS LUPUS

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Case 1: 35-year-old Mestizo female
Bernardo Pons-Estel

A 35-year-old Mestizo female was diagnosed with systemic lupus erythematosus (SLE) in 2005 based on polyarthritis, malar rash, photosensitivity, mucosal ulcerations, positive ANA and anti-dsDNA, and low complement. She was treated with prednisone 20–30 mg/day and hydroxychloroquine (HCQ) 400 mg/day. In June 2010, lupus pneumonitis was diagnosed.

In July 2018, she was first admitted to our hospital. She was cushingoid and had fever, fatigue, malar rash, oral ulcers, alopecia, polyarthritis, oedema, multiple purplish-red streaks, and active erythematous, palpable and painful subcutaneous indurated nodules/plaques located on her face, proximal lower extremities and abdomen; some were ulcerated.

Laboratory tests RBC 3.8 ($x10^{12}$/L), hemoglobin 11.8 g/dl, WBC 2,3 ($x10^{9}$/L), platelets 62 ($x10^{9}$/L), ESR 8 mm, CRP 0.8 mg/L, serum ferritin 1,487 ng/ml, ALAT 70 UI/ml, ASAT 26 UI/ml, GGT 64 U/L, BUN 43 mg/dl, serum creatinine 1.54 mg/dl, GFR 67 mL/min, cholesterol 186 mg/dL, triglycerides 149 mg/dl, proteinuria 210 mg/24 h, ANA 1/320, speckled pattern, anti-Sm (+) and anti-dsDNA, anti-U1RNP, anti-Ro and anti-La all (-). C3 75 mg/dl, C4 10, Coombs test (-), procalcitonin 0.29 ng/ml (<0.5). VDRL and viral serologies were negative. A cutaneous ulcer culture showed proteus mirabilis. Her SLEDAI was19. Three skin biopsies of indurated lesions all showed lobular panniculitis.

She was treated with IV methylprednisolone (500 mg/day/3 days), and high-dose intravenous immunoglobulin, and discharged with mycophenolate mofetil 1 g/day, HCQ 400 mg/day, prednisone 20 mg/day and TMS-SMX 860/160 BID for 5 days. A week later she was re-admitted with fever, pancytopenia, hepatosplenomegaly, lymphadenopathy and several painful, indurated erythematous lesions. Her SLEDAI was 13. A bone marrow aspiration and biopsy was interpreted as a macrophage activated syndrome in the context of SLE exacerbation and treated with IV dexamethasone, colony stimulating factor, and rituximab 1000 mg. Despite treatment, she remained severely ill with fever, asthenia, petechiae and purpura on her abdomen and thighs, and pancytopenia. Due to disease severity, treatment with etoposide was indicated. Finally, the patient presented an acute episode of respiratory distress followed by death.

Discussion Points

- Interpreting the different skin manifestations in a patient with SLE
- Analyzing complications and differential diagnoses with other associated diseases

Case 2: 28-year-old Caucasian female
Annegret Kuhn

A 28-year-old Caucasian female was diagnosed with systemic lupus erythematosus (SLE) in 1996 and presented with severe, erythematous, scarring discoid lesions on the scalp,

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

19 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 meqs/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,