LUPUS TREATMENT IN THE NEXT DECADE: THE NEXT CUTANEOUS LUPUS

Learning Objectives

Discuss biologic targets for SLE drug development

Explain recent clinical trial results

REFERENCES


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 polyarthritus, 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, polyarthritus, oedema, multiple purpural-red streaks, and active erythematous plaques on her face, proximal lower extremities and abdomen; some were ulcerated.

Laboratory tests RBC 3.8 (x1012/L), hemoglobin 11.8 g/dl, total WBC 2.3 (x109/L), platelets 62 (x109/L), ESR 8 mm, CRP 0.8 mg/dL, serum ferritin 1,487 ng/mL, ALAT 70 IU/mL, ASAT 26 IU/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 -dsDNA, anti-UlRNP, anti-Ro and anti-La all (+). C3 75 mg/dL, C4 10, Coombs test (+), pro-calcitonin 0.29 ng/ml (<0.5). VDRL and viral serologies were negative. A cutaneous ulcer culture showed Proteus mirabilis. Her SLEDAI was 19. 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 erythematosus 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.

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,