Bullous systemic lupus erythematosus: phenotypic variability after seven years of follow up

P.Iranzo, E.Darwich, J.M.Mascaró, J.E. Herrero*, C.Herrero.

Department of Dermatology. Hospital clinic. Hospital de Mar* . Barcelona. Spain

Introduction: Bullous systemic lupus erythematosus (BSLE) is an acquired subepidermal autoimmune blistering disease arising in patients with systemic lupus erythematosus. The immunopathologic criteria of BSLE are similar to those of epidermolysis bullosa acquisita and autoantibodies to type VII collagen are also present. We describe the evolution of a patient with BSLE.

Method: Analysis of cutaneous and systemic manifestations, inmunopathologic examinations and laboratory findings of a patient with BSLE after 7 years follow-up. *In vitro* demonstration of serum pathogenicity using human skin cryosections.

Results: A 15-year-old girl with a 2 month history of asthenia, arthro-myalgias, hand swelling and photo-induced malar erythema, presented arciform erythematous plaques with peripheral tense blisters on the trunk, facial vesicles, oral erosions, fever, malaise and arthritis. Laboratory: ESR 77 mm/hr, hemolytic anemia (+ direct Coombs test), subclinical hypothyroidism, urine proteins: 4926 mg/24 h, ANA > 1:640 URF, anti-dsDNA > 200, antibodies to anti-Ro, La, RNP, and Sm negative and hypocomplementemia. Renal biopsy: Type IV nephritis, with extensive deposition of IgA, IgG, IgM, C1q, C3, and C4. Cutaneous biopsy: subepidermal blister, superficial perivascular dermatitis with neutrophils. Direct immunofluorescence: IgG, IgA, IgM deposition at the floor of the blister. Immunoblot: *145* KD band. *In vitro,* serum patient induced subepidermal split of human skin cryosections in presence of normal leukocytes.Treatment with prednisone 1mg/kg/day, chloroquine 150 mg/day and cyclophosphamide 750 mg/ m^2 monthly pulses, resulted in an immediate clearing of the cutaneous lesions and progressive decrease of proteinuria. Three years later, she presented with facial annular lesions, axilary granulomatous interstitial dermatitis, ANA + > 640 URF, anti-dsDNA, CLIFT +++, and positive antibodies anti-Sm, anti-U1RNP, and anti-Ro.

Conclusions: We report the phenotypic variability, immunological instability, the pathogenicity of patient's serum, long term evolution of BSLE and its non-reported association with granulomatous interstitial dermatitis.