



AUTOIMMUNE CONNECTIVE TISSUE DISEASES

CHILBLAIN LUPUS ERYTHEMATOSUS - CLINICAL AND HISTOLOGIC CHARACTERIZATION OF AN INSTITUTIONAL COHORT

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Chilblain lupus erythematosus (CHLE) is a rare form of chronic cutaneous lupus characterized by lesions in an acral distribution exacerbated by exposure to cold. There is a paucity of literature concerning patients with CHLE, with the last cohort reported over 15 years ago. Additionally, diagnostic criteria for CHLE are not well-defined.

We conducted a retrospective chart review of CHLE patients seen at the Cleveland Clinic from 1980-2015. Patients were included if they satisfied one “required” criteria (acral skin lesions induced by exposure to cold) and at least 1 of 2 “major” criteria: (1) documented histologic evidence of cutaneous lupus erythematosus (CLE), (2) met American College of Rheumatology criteria for SLE. Clinical features were collected and analyzed including sex, involved site(s), presence of SLE and/or Raynaud's, antibody profile, histologic features, treatment and response.

We identified 18 patients who met our criteria for a diagnosis of CHLE (16 women; 2 men). CHLE lesions were most commonly found on fingers (n=16 pts), hands (11), toes (10), feet (7), and ears (4). Most patients (11/18) had a diagnosis of SLE prior to CHLE onset and suffered from Raynaud's phenomenon (13/18). All patients were ANA positive; other common autoantibodies included dsDNA(10/15), SSA(7/16) and RNP(7/15). Twelve patients had biopsies of a CHLE lesion, and these typically displayed superficial and deep inflammation with variable interface change. Treatment strategies varied, but included both immunomodulatory medications and vasoactive medications. The majority of patients had improvement of their CHLE lesions with treatment.

CHLE is a rare subtype of CLE seen at our institution, and mainly occurred in women with SLE. Raynaud's phenomenon and hyperviscosity may be predisposing factors for this rare cutaneous lupus subtype. Treatment with immunomodulatory and/or vasoactive medications tends to be beneficial in affected patients.

