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ORIGINAL ARTICLE

Subacute Cutaneous Lupus Erythematosus: Clinical Characteristics, Disease Associations, Treatments, and Outcomes in a Series of 90 Patients at Mayo Clinic, 1996-2011

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Abstract

Objective: To characterize the clinical presentation, laboratory studies, disease associations, and treatments of subacute cutaneous lupus erythematosus (SCLE).

Patients and Methods: A retrospective review of 90 patients with SCLE at Mayo Clinic from January 1, 1996, through October 28, 2011, was performed.

Results: The mean patient age at diagnosis was 61 years; 64 patients (71%) were women, and 11 cases (12%) were drug induced (1996-2000, no drug-induced cases; 2001-2005, 2 cases; 2006-2011, 9 cases). Seventeen of 59 patients (29%) with available data were smokers at the time of diagnosis. The SCLE lesions were photodistributed in 75 patients (83%), and 52 (58%) had papulosquamous morphologic findings. Anti-Ro/SS-A positivity was present in 84 of 85 patients tested (99%), whereas 32 of the 85 patients (38%) tested positive for anti-La/SS-B. Associated autoimmune connective tissue diseases included Sjögren syndrome (n=13, 14%) and systemic lupus erythematosus (SLE) (n=8, 9%). Eighteen patients (20%) had at least 4 American College of Rheumatology criteria for SLE; 1 had lupus nephritis, and none had neurologic or notable hematologic sequelae. The most common therapy was hydroxychloroquine, with a complete response noted in 34 of 46 patients (74%) with available follow-up data.

Conclusion: Twenty-eight percent of patients with SCLE (n=25) had an associated autoimmune connective tissue disease, although the severe sequelae of SLE, such as nephritis, were rare. The frequency of drug-induced SCLE increased during the study. Most patients responded to treatment with hydroxychloroquine.

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Subacute cutaneous lupus erythematosus (SCLE) is classically a photosensitive, nonscarring, non–atrophy-producing, erythematous annular or papulosquamous eruption that is typically distributed on sun-exposed skin of the upper torso, back, neck, and arms.[1,2] It is often idiopathic but also can be caused by a wide range of various medications.[3,4] Through this study, we aimed to examine the clinical characteristics, etiologic factors, serologic and disease associations, treatments, and outcomes of SCLE.

PATIENTS AND METHODS

Patients with SCLE treated at the Rochester campus of Mayo Clinic from January 1, 1996, through October 28, 2011, were identified from the institutional medical index and text retrieval system retrospectively. Databases of Mayo Clinic in Arizona and Mayo Clinic in Florida were not included in the medical index and retrieval system used for this study and, thus, were not included in the final patient cohort. Tertiary referral patients and local patients were included in the study. All the study participants provided research consent for review of their medical records. This study was approved by the Mayo Clinic Institutional Review Board. All the patients included in this study had a final clinical diagnosis of SCLE rendered by a dermatologist on the basis of historical, clinical, and histopathologic information.

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The following information was abstracted from each patient's medical record: demographic characteristics; smoking history; clinical features of SCLE eruption, including morphologic characteristics and distribution; etiologic factors; histopathologic findings on skin biopsy as interpreted by dermatopathologists; serologic data; and associated disease history, treatments, outcomes, and follow-up data. A patient was determined to have drug-induced SCLE on the basis of the clinical impression of the dermatologist who treated the patient.

RESULTS

Clinical Characteristics

After an initial review of 712 possible patients identified from the Mayo Clinic retrieval system, 90 were determined to have a final diagnosis of SCLE after a thorough review of the medical records (Table 1). The other 622 patients were excluded for various reasons, including an alternative final clinical diagnosis, lack of skin biopsy results to corroborate the SCLE diagnosis, and lack of dermatology consultation to confirm the SCLE diagnosis. All 90 patients had a skin biopsy that showed changes consistent with SCLE. These patients had a mean age at disease onset of 61 years (range, 20-90 years), and 64 (71%) were women and 86 (96%) were of white race; 25 of 59 patients (42%) reported current or past tobacco use. The most common distribution of the lesions was in sun-exposed areas (n=75, 83%), with papulosquamous morphologic findings (n=52, 58%). Ten patients (11%) with SCLE had concomitant presence of an additional subtype of cutaneous lupus (discoid lupus, 5 patients; and acute cutaneous lupus [malar rash], 5 patients).

Etiologic Factors

Most cases (n=79, 88%) in this SCLE cohort were idiopathic. However, 11 cases (12%) were attributed to drug-related causes (Table 1). The most common drug was hydrochlorothiazide (n=6, 55%), followed by an angiotensin-converting enzyme inhibitor (n=3, 27%), with 1 case (9%) each for diltiazem and fluoxetine. Six of these 11 patients (55%) took the inciting drug for 1 year or less before SCLE symptom onset, 2 (18%) had taken the offending drug for 2 or more years before symptom onset, and 3 (27%) had an unknown duration of drug ingestion before SCLE developed.

Stratification of patients with drug-induced SCLE into approximately 5-year periods based on time of diagnosis demonstrated the following: 1996 to 2000, 22 idiopathic and 0 drug-induced cases; 2001 to 2005, 27 idiopathic and 2 drug-induced cases; and 2006 to 2011, 30 idiopathic and 9 drug-induced cases.

Of 8 patients who underwent removal of the offending drug from their systems, 5 had clearance of SCLE and 3 had persistence of the skin condition. The other 3 patients with drug-induced SCLE continued to take their suspected offending drug or it was unknown whether use of the offending drug was stopped.

Serologic and Other Disease Associations

The most common serologic associations in patients with SCLE were anti-Ro/SS-A (84 of 85, 99%) and antinuclear antibody (78 of 88, 89%). The next strongest associations were rheumatoid factor or cyclic citrullinated peptide (19 of 49, 39%) and anti-La/SS-B antibody (32 of 85, 38%). The most common associated autoimmune connective tissue diseases (ACTDs) were Sjögren/sicca syndrome, affecting 13 patients (14%), followed by systemic lupus erythematosus (SLE), affecting 8 patients (9%). Overall, 25 of the 90 patients (28%) had an associated ACTD. Table 1 summarizes these associations.

The American College of Rheumatology (ACR) uses 11 criteria points for SLE classification; a patient with 4 or more of these criteria is said to have SLE by definition.[5] Of 90 patients, 18 (20%) met the ACR criteria for the diagnosis of SLE, although only 8 of these 18 patients (44%) received a formal SLE diagnosis on rheumatologic evaluation. Of the 18 persons, only 1 was found to have notable systemic involvement with lupus nephritis, and the other patients with SLE or overlap syndromes had a more limited disease course, with no renal, neurologic, or notable hematologic sequelae.

Treatment, Responses, and Follow-up

Patients in this cohort received topical or oral therapy, or a combination of both.

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