

Clinical characteristics of lupus erythematosus panniculitis/profundus: a retrospective review of 61 patients

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Background: Lupus erythematosus panniculitis/profundus (LEP) is a rare variant of cutaneous lupus erythematosus (CLE), characterized by involvement of the subcutaneous fat. LEP can coexist with systemic lupus erythematosus (SLE) or discoid lupus erythematosus (DLE), or occur in isolation. In this study, we present the largest cohort of LEP patients to date and aim to investigate the clinical presentation of LEP and its relationship to SLE and DLE.

Methods: We conducted a retrospective study of 5,717 patients diagnosed with CLE from January 2000 to October 2019 at Brigham and Women's Hospital, Massachusetts General Hospital, and New York University Langone Medical Center. We confirmed 61 cases of LEP.

Results: Of 61 subjects, 52 were female and 9 were male; 42.9% identified as non-Hispanic white, 23.2% as black, 17.9% as Hispanic, and 16.1% as Asian. The mean age of LEP symptom onset was 35.8 years of age. LEP involved 2 or more body locations in 59% of patients. The most common lesion locations were the face and proximal upper and lower extremities. ANA was positive in 67.8% of patients. A mean of 2.2 systemic therapies was required for disease quiescence, and the majority of patients required at least one immunosuppressive agent for disease control.

Thirteen subjects had a concurrent diagnosis of SLE, while 17 had DLE; 8 had both SLE and DLE. Of those with concurrent SLE, 17 were diagnosed with SLE prior to LEP onset, 3 were diagnosed with LEP prior to SLE (mean=10.1 and 8.7 years between diagnoses, respectively); 1 was diagnosed simultaneously. With regards to DLE, 13 were diagnosed with DLE prior to LEP and 2 developed DLE after LEP (mean=10 and 2.1 years between diagnoses, respectively); 9 patients were diagnosed simultaneously.

Mean time from initial presentation to diagnosis was 19.2 months (n=45). Of these patients, the 23 subjects with no history of SLE or DLE at time of LEP onset had a mean time to diagnosis of 29.8 months, whereas those with a history of SLE and/or DLE had a mean time to diagnosis of 8.8 months.

Conclusion: This is the largest study of LEP to date. While the co-occurrence of LEP and SLE has historically been considered to be 10-15%, this study, with its 34.4% co-occurrence, supports more recent data that demonstrates a higher association of LEP with

SLE of 33-45%. Furthermore, LEP is often recalcitrant to initial therapy, and patients often experience long diagnostic delays, particularly those without a history of SLE or DLE.