Incidence of cutaneous lupus erythematosus and risk of disease progression into systemic lupus erythematosus: a Swedish population-based, nationwide register-based open cohort study including 1,088 patients

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Background/Objectives: Population-based epidemiologic studies reporting the incidence of isolated cutaneous lupus erythematosus (CLE) are rare. The main aim of this population-based cohort study was to examine the incidence of CLE and its subsets in Sweden. A further aim was to investigate the co- morbidity and short-time risk in the development of systemic lupus erythematosus (SLE).
Methods: We conducted a population-based open cohort study. The cohort included all patients diagnosed with CLE (defined according to the relevant International Classification of Disease (ICD) code; ICD-10: L93) in Sweden between 2005 and 2007. The cohort (n=1088 unique patients) was derived from the Swedish National Patient Register (NPR).
Results: The incidence of CLE in Sweden was 4.0/100,000. The female:male ratio was 3:1. The most common age of onset was 35-85 years (mean age 54 years). The most common reported subset was discoid lupus erythematosus (DLE) (80 %). Two hundred sixty (24 %) patients were already diagnosed with SLE at the time they were diagnosed with CLE. More than 10 % progressed to SLE during the first year after being diagnosed with CLE, with the risk of progression being highest for the subacute cutaneous lupus erythematosus (SCLE) subset. During the observation period (2005-2007), nearly an additional 20 % had disease progression to SLE.
Conclusions: This is the first nationwide epidemiologic study on CLE in Sweden. Our study showed that the incidence of CLE is about equal to that of SLE and that there was a higher short-term risk for progression to SLE than previously described for CLE. Other subsets than DLE and SCLE were rarely reported in our system and therefore an update of the ICD codes for this diagnostic group could increase reporting of these cases. Our study clarifies that CLE is not a rare disease and that close monitoring and follow-up are called for in this patient group because of the risk of developing SLE, especially in female patients.