**Introduction**

The skin represents one of the major organs afflicted by lupus erythematosus (LE). LE was in fact first described as a skin disease in the mid and late 19th century (Talbot, 1993) and denominated for the characteristic mutilations seen in some subtypes of the disorder. Ever since in 1936 systemic manifestation without skin symptoms was appreciated as a disease entity, the subject has differentially been dealt with in the dermatological and rheumatological literature. Malar rash and discoid lesions are listed among the criteria defined by the American College of Rheumatology (ACR) for the diagnosis of systemic lupus erythematosus (SLE) (Tan et al., 1982). Characteristic and defined entities encompassing various forms of acute, subacute and chronic cutaneous LE can be opposed to less characteristic skin manifestations like rashes and symptoms of cutaneous vasculitis (Provost, 2004). Both groups of symptoms may however be present at any stage of the disease development. 20% of all SLE cases present with initial skin manifestations and 50–70% of all SLE patients will eventually show skin symptoms during the course of their disease (Costner et al., 2003).

**Classification**

Chronic cutaneous lupus erythematosus (CCLE) comprises different clinical entities with discoid lupus erythematosus (DLE) as the most common form (Kuhn et al., 2007, Rothfield et al., 2006). In contrast to acute cutaneous LE (ACLE) and subacute cutaneous LE (SCLE), lesions are long-lasting for up to decades with occasional spontaneous remissions. Disfiguring atrophy and scarring with emotional discomfort for patients as well as an increased risk to develop into squamous cell carcinoma in long-standing DLE pose distinct medical problems (Costner et al., 2003, Patel and Werth, 2002). In many cases, symptoms are restricted to the skin without major systemic inflammatory or autoimmune manifestations. This used to be undiscriminately called DLE until thirty years ago when it was clearly dis-
sected from other forms, especially from what is now referred to as SCLE (Gilliam, 1977, Gilliam, Sontheimer, 1981). The term DLE should nowadays be restricted to those subsets of CCLE with morphologically distinct plaques irrespective of systemic involvement.

In other, rather rare cases, DLE as well as further subsets of CCLE may present as the first or intercurrent manifestations of SCLE and SLE and will eventually result in systemic disease. Exact epidemiological data of the different CCLE subtypes as well as their relation to SLE and SCLE are not available partly due to the different perception over the last couple of decades and within dermatology and rheumatology. CCLE may be underestimated within rheumatological literature and in case of absent systemic manifestations not be amply diagnosed at all. CCLE lesions, especially DLE lesions are found in up to 20% of SCLE patients and may predate manifestation of systemic disease. Further support of the relation as well as distinction of DLE and SLE is provided by the finding that DLE is present in 15–30% of SLE patients at any time during their disease course and is the prominent feature in 5–10% of such patients (Parodi and Rebora, 1997; Tebbe et al., 1997, Rothfield et al., 2006). Classical DLE as termed at the time of first diagnosis will progress into SLE in 5–10% of cases (Parodi and Rebora, 1997; Tebbe et al., 1997). Such courses may account for different criteria of diagnosing SLE and varying medical check-up among different medical specialists. Generalized DLE as well as DLE associated with high autoantibody titers have a higher chance to develop into systemic disease. However, the overall incidence of DLE is estimated ten-fold higher than that of SLE. At the same time, the female to male ratio of 3:2 to 3:1 for DLE is quite distinct from 9:1 in SLE indicating separate entities. Similarly, the age of disease manifestation (20–40 years) is slightly higher in DLE than in SLE. The other CCLE subtypes show varying correlations to SLE and will be discussed below (Costner et al., 2003).

The issue of CCLE as a distinct and separate entity at the benign end of a spectrum of LE or a limited stage or concurrent manifestation within the chronological evolution of SLE has to be further evaluated. Especially, characteristic etiopathogenic factors as well as prognostic markers for CCLE remain to be elucidated. The final diagnosis of a distinct subset of LE can only be made following careful case history, clinical manifestations at the skin and other organs and laboratory findings encompassing the criteria defined by the American College of Rheumatology (ACR). Alternative classification criteria for cutaneous lesions have been suggested by the European Academy of Dermatology and Venereology (EADV) and are currently debated. Being more specific, however less sensitive than the ACR criteria, they have so far not found their way into the rheumatologic literature (Parodi and Rebora, 1997).

Pathogenesis

Various causes and resulting inflammatory and immunological processes have been identified as relevant for the induction of cutaneous LE (reviewed by Lee and Sinha, 2006, Pelle, 2006, Kuhn and Bijl, 2008). However, their particular role in individual clinical subsets of CCLE with respect to quantitative or qualitative differences has been barely addressed nor