Lupus erythematosus (LE) is a chronic inflammatory disease that can be clinically divided into three major categories: chronic cutaneous LE (CCLE), subacute cutaneous LE (SCLE), and systemic LE (SLE). In general, LE represents a spectrum of disease with some overlap of these categories. Classification is based on clinical, histologic, serologic, and immunofluorescent (see Chap. 22) features. Consequently, histologic findings alone may not be sufficient for correct classification. In addition, these categories can be subdivided into numerous variants affecting different levels of the skin and subcutaneous tissue.

**Chronic Cutaneous Lupus Erythematosus**

**Discoid Lupus Erythematosus**

Discoid LE (DLE) is the most common form of LE. Clinically, the head and neck region is affected in most cases. On the face, there may be a butterfly distribution. However, in some cases, the trunk and upper extremities can be also involved. Lesions consist of erythematous scaly patches and plaques.

Histologically, in DLE the epidermis and dermis are affected, and the subcutaneous tissue is usually spared. However, patchy infiltrates may be present. Characteristic microscopic features are hyperkeratosis with follicular plugging, thinning, and flattening of the epithelium and hydropic degeneration of the basal layer (liquefaction degeneration) (Fig. 21.1). In addition, there are scattered apoptotic keratinocytes (Civatte bodies) in the basal layer or in the epithelium. Particularly in older lesions, thickening of the basement membrane becomes obvious in the periodic acid-Schiff stain. In the dermis, there is a lichenoid or patchy lymphocytic infiltrate with accentuation of the pilosebaceous follicles. There is interstitial mucin deposition and edema, and usually no eosinophils and neutrophils are present.

Hypertrophic lesions have acanthosis and hyperkeratosis of the epidermis (Weldon 2002). Direct immunofluorescence is an important test that should be performed in this subtype. Usually, there is deposition of IgG and IgM in 50%–90% of cases. The differential diagnosis includes drug reaction, dermatomyositis, graft-vs-host disease, and mycosis fungoides. Jessner’s lymphocytic infiltration of the skin usually does not involve the epidermis as hydropic degeneration of the basal layer and scattered apoptotic keratinocytes do not occur. Otherwise, the dermal infiltrate is identical to LE. Consequently, Jessner’s lymphocytic infiltration of the skin is regarded as a variant of LE by some authors. In a drug reaction there are frequently eosinophils that are not
seen in DLE. In graft-vs-host disease, there is usually not a prominent lymphocytic infiltrate involving the dermis. Early-stage dermatomyositis can be histologically difficult to distinguish from DLE because it can exhibit identical changes. Consequently, clinical features should be considered for diagnosis. In mycosis fungoides, lymphocytes frequently exhibit nuclear atypia.

**Subacute Cutaneous Lupus Erythematosus**

The term “subacute cutaneous lupus erythematosus” was coined in 1977 by Gilliam (Gilliam 1977) and further characterized by Sontheimer et al. (Sontheimer et al. 1979) in 1979. The authors introduced a previously undescribed variant of LE with skin lesions at the neck, shoulders, upper thorax, and arms. Clinically, recurrent annular polycyclic or papulosquamous lesions involve the upper trunk, extensor regions of the arms, and face and neck region.

The histopathologic features of SCLE are almost identical to those of DLE (Fig. 21.2). In individual cases, in particular in early lesions, differentiation by histopathologic features alone may be difficult or impossible. However, usually in SCLE there are more prominent changes at the dermoeipidermal interface such as hydropic degeneration of the basal epithelial layer. Forming clefts or vesicles, epidermal atrophy, and dermal edema may be also more prominent as in DLE, and there may be extravasation of erythrocytes and dermal fibrin deposits. Apoptotic keratinocytes (Civatte bodies) in the epidermis may be numerous. In addition, the lymphocytic infiltrate is more confined to the upper dermis and bandlike compared with DLE.

Compared with DLE, SCLE has less hyperkeratosis, atrophy of pilosebaceous units, follicular plugging, and thickening of the basal layer (Bangert et al. 1984, Jerdan et al. 1990). The lupus band test result is positive in approximately 60% of cases.