5 Cutaneous Lymphoproliferative Tumours

5.1 Cutaneous T-cell lymphoma

A primary T-cell lymphoma of the skin.

5.1.1 Mycosis fungoides

(Fig. 104)

Mycosis fungoides in most cases first becomes evident in the patch stage, followed by evolution into plaque and tumour or nodule stages. In some cases, however, the plaque stage appears to develop de novo. Involvement of all cutaneous surfaces may be very extensive in the plaque and tumour stages. Erythroderma may occur during any of the three stages of mycosis fungoides.

**Patch stage.** The epidermis characteristically shows psoriasiform hyperplasia overlying a perivascular and diffuse lymphocytic infiltrate, sometimes relatively sparse, in the superficial dermis. The lymphocytes may be closely applied to the basal layer of the epidermis in a focally lichenoid pattern; fibrosis is present in the superficial stroma, where coarse bundles of collagen replace the normal delicate fibrils of the papillary dermis. Epidermotropism of atypical lymphocytes with hyperchromatic, irregular (cerebriform) nuclei is an important feature. These cells may be arranged singly or in small collections between the keratinocytes, sometimes associated with slight spongiosis, but only rarely with spongiotic vesicles. There is also a poikilodermatous form of patch-stage mycosis fungoides in which the epidermis is atrophic, with alternating hyper- and hypopigmentation and telangiectasia in the superficial dermis. Epidermotropism may be subtle in the patch stage, an early sign being the presence of lymphocytes in a beaded pattern between the epithelial cells of the basal layer.

**Plaque stage.** In the plaque stage there is a denser infiltrate of lymphocytes in the papillary and superficial reticular dermis, composed of atypical lymphoid cells with hyperchromatic nuclei, markedly convoluted nuclear membranes (cerebriform nuclei), plasma cells, larger cells with pale cytoplasm, vesicular nuclei and prominent nucleoli resembling Reed-Sternberg cells, and
eosinophils. Epidermotropism is more pronounced with for­
formation of collections of atypical lymphoid cells (Pautrier
microabscesses). Lymphocytic infiltration of the epithelium of
adnexal structures is often present, sometimes with associated
excessive mucin deposition in and degeneration of follicles
(follicular mucinosis).

**Tumour stage.** In this stage there is a deep and dense dermal
infiltrate composed of sheets of lymphoid cells with infiltration
of adnexal structures. The cell population is more mono­
morphous, composed predominantly of atypical, pleomorphic
lymphoid cells including large immunoblast-like cells. Epider­
motropism is absent or sparse, and the papillary dermis also is
often spared.

**Variants**

**Spongiotic mycosis fungoides.** In some cases spongiosis with
vesicle formation may co-exist with epidermotropism of atypical
lymphoid cells in non-spongiotic areas.

**Hypereosinophilic mycosis fungoides.** Although eosinophils are
frequently present, especially in the plaque stage of mycosis
fungoides, in some cases they are much more plentiful, forming
dense groups and sheets, outnumbering the lymphoid cells in
some areas.

**Granulomatous mycosis fungoides.** Some cases of mycosis
fungoides are characterized by a prominent granulomatous com­
ponent in the infiltrate, composed of epithelioid histiocytes in a
diffuse or more localized pattern, resembling sarcoidosis or
tuberculous leprosy.

**Granulomatous slack skin** (Fig. 105). This rare condition may
be a variant of granulomatous mycosis fungoides in which there
is extensive elastophagocytosis by macrophages and giant cells
leading to the formation of loose folds of skin, especially in the
groin and axillae.

**5.1.2 Sezary syndrome**

*Cutaneous T-cell lymphoma in association with circulating neo­
plastic T-lymphocytes in the peripheral blood.*