Mycosis Fungoides (Cutaneous T-Cell Lymphoma)

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Mycosis fungoides is the most common form of cutaneous T-cell lymphoma. In the western population there are around 0.3 cases of Sezary syndrome per 100,000 people. Sézary disease is more common in males with a ratio of 2:1, and the mean age of diagnosis is between 55 and 60 years of age.

Clinical Description

The curious name "mycosis fungoides" refers to the initial clinical description of mushroom-like tumors evolving from a desquamating rash. Four clinical stages are seen:

1. Patch stage-persistent, pruritic, red, pink, or brown patches, with or without scale
2. Plaque stage-persistent, pruritic, red, pink, or brown plaques
3. Tumor stage-persistent red, brown, or violet papules, nodules, and/or tumors (d' emblee type refers to sudden appearance of tumors without previous patches or plaques)
4. Sezary syndrome-exfoliative erythroderma with numerous, bloodborne Sezary cells (convoluted T lymphocytes) [1–6]

Etiology

The cause of mycosis fungoides is still unknown.

Theories include chronic, low-grade contact dermatitis and/or retrovirus (HIV III, HTL V I) infection. (HIV III, also called HTLV I, is different from the AIDS virus, which is HlV I, and is also called HTLV III). The result is a malignant clone of helper T cells [7–10].

Histopathology

Cutaneous T-cell lymphoma shows a lichenoid (band-like) lymphocytic infiltrate with Pautrier microabscesses consisting of collections of atypical cerebriform or hyperconvoluted T lymphocytes in the epidermis, with no or at most minimal spongiosis and a mixed lymphohistiocytic perivascular dermal infiltrate, with variable eosinophils and plasma cells. The lack of spongiosis is one clue to distinguishing cutaneous T-cell lymphoma from eczematous diseases.

Differential Diagnosis

The differential diagnosis includes the following:

1. Patch stage-eczema, tinea, pityriasis rosea, pityriasis lichenoides chronica, secondary syphilis, other papulosquamous disorders
2. Plaque stage-psoriasis, parapsoriasis en plaques (large plaque parapsoriasis may represent a precursor lesion to mycosis fungoides)
3. Tumor stage-squamous cell carcinoma (usually single, not multiple), other lymphomas (cutaneous nodules of Hodgkin's disease and leukemic infiltrates), postscabetic nodules, Kaposi's sarcoma
4. Sezary syndrome-other causes of exfoliative erythroderma, including psoriasis, generalized eczema, drug eruptions, tinea, erythema multiforme (toxic epidermal necrolysis)

R.A. Norman, E.M. Young, Jr, *Atlas of Geriatric Dermatology*,
DOI 10.1007/978-1-4471-4579-0_47, © Springer-Verlag London 2014

Therapy

Treatment includes the following:

1. Electron beam therapy, orthovoltage radiotherapy
2. Topical chemotherapy-topical nitrogen mustard (mechlorethamine), topical carmustine (BCNU, bischloroethyl nitrosurea)
3. Systemic chemotherapy-methotrexate, cyclosporine
4. Photochemotherapy–PUVA (psoralen plus UV A light)
5. Extracorporeal photochemotherapy-plasmapheresis and PUVA
6. Vorinostat is a second-line drug for cutaneous T-cell lymphoma. Treatments are often used in combination with phototherapy and chemotherapy

No single treatment type has revealed clear-cut benefits in comparison to others, and treatment for all cases remains problematic.

Prognosis

The prognosis is poor, with a gradual but inexorable progression from patch to plaque to tumor stage, which may take over 20 years. Patients often succumb to other diseases, infections, or complications of therapy for mycosis fungoides. Patients with Sézary disease have a median survival of 5 years.

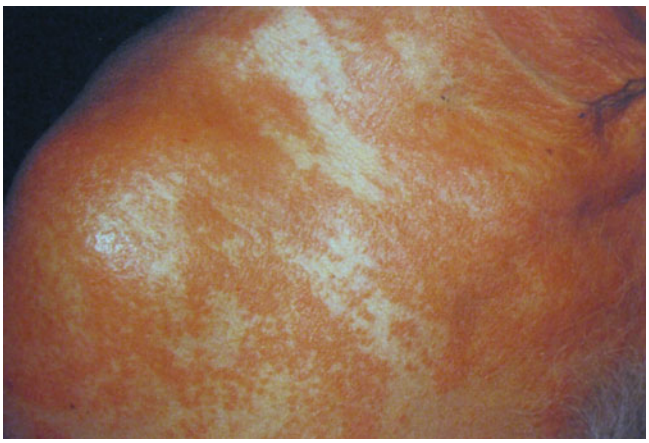


Fig. 47.1 Patch stage of mycosis fungoides. This may be treated as "eczema" for 10–20 years before the correct diagnosis is made by biopsy

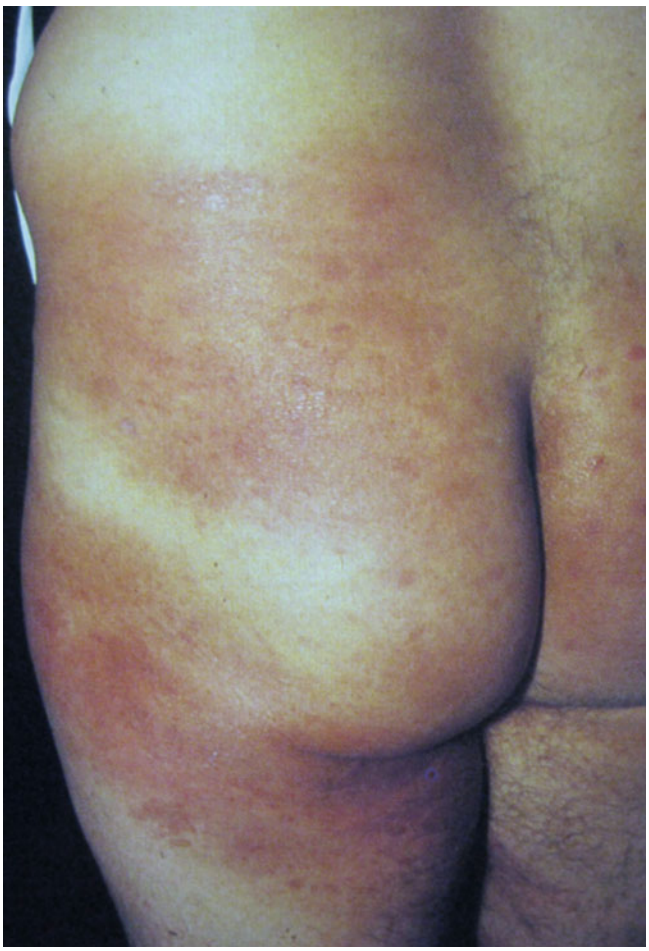
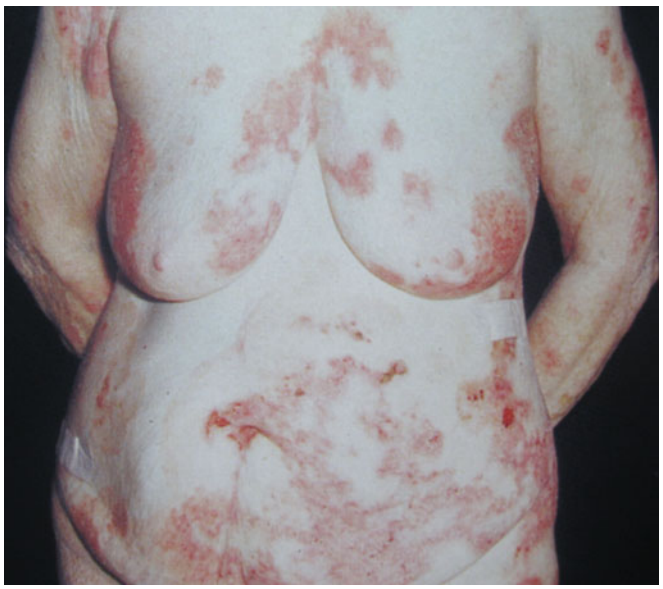
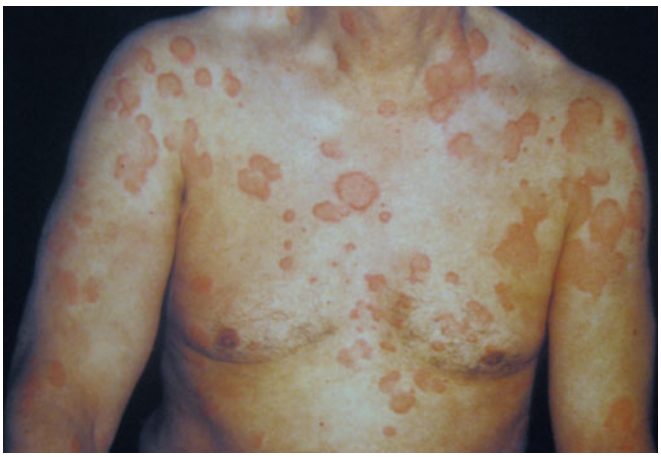
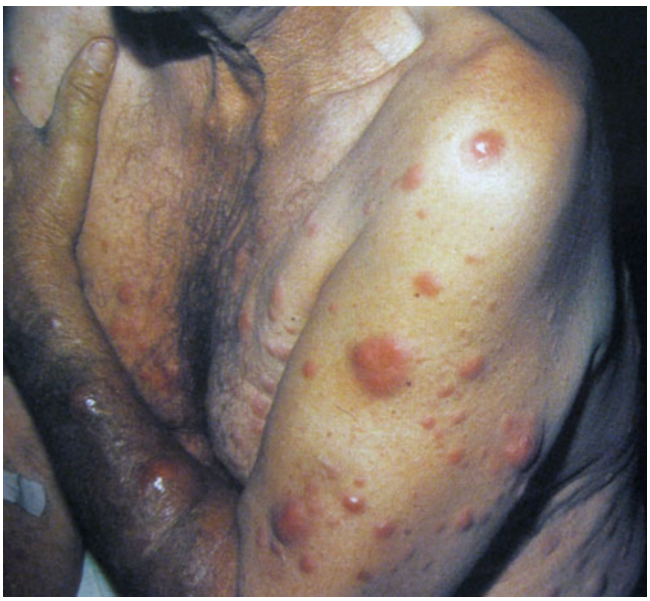


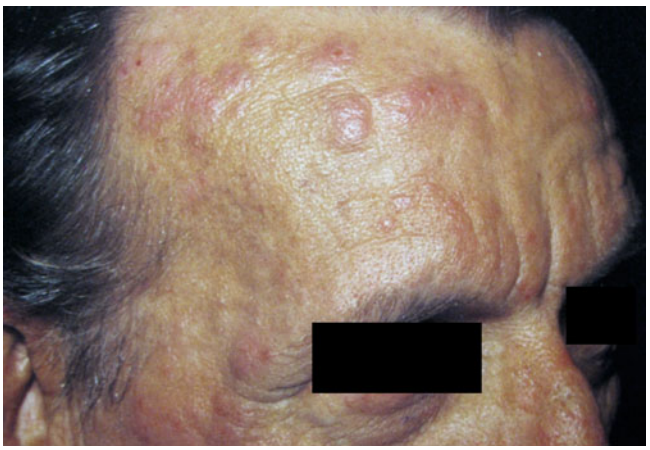
Fig. 47.2 Large plaque parapsoriasis of the thighs and buttocks shows histologic changes of mycosis fungoides

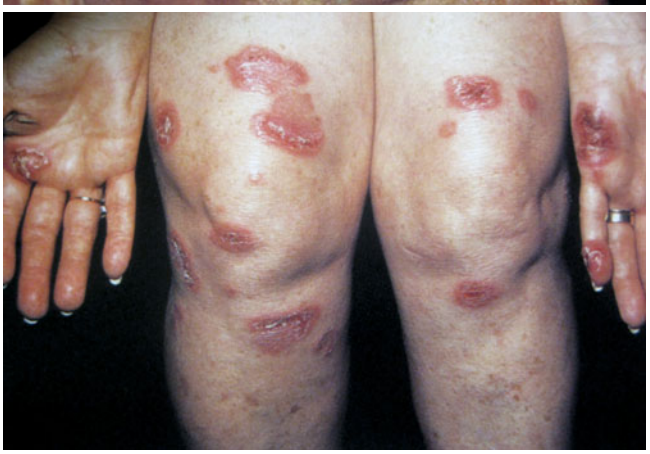




Figs. 47.3 and 47.4 Widespread plaque stage of mycosis fungoides







Figs. 47.5, 47.6, and 47.7 Tumor or nodular stage of mycosis fungoides

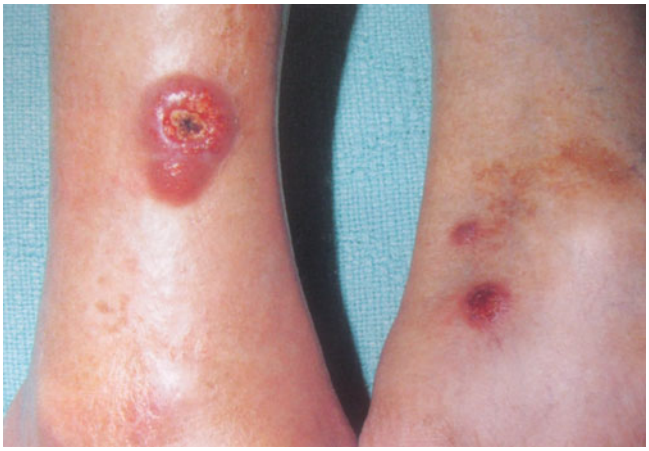


Fig. 47.8 Mycosis fungoides presenting as ulcerated nodules on the legs

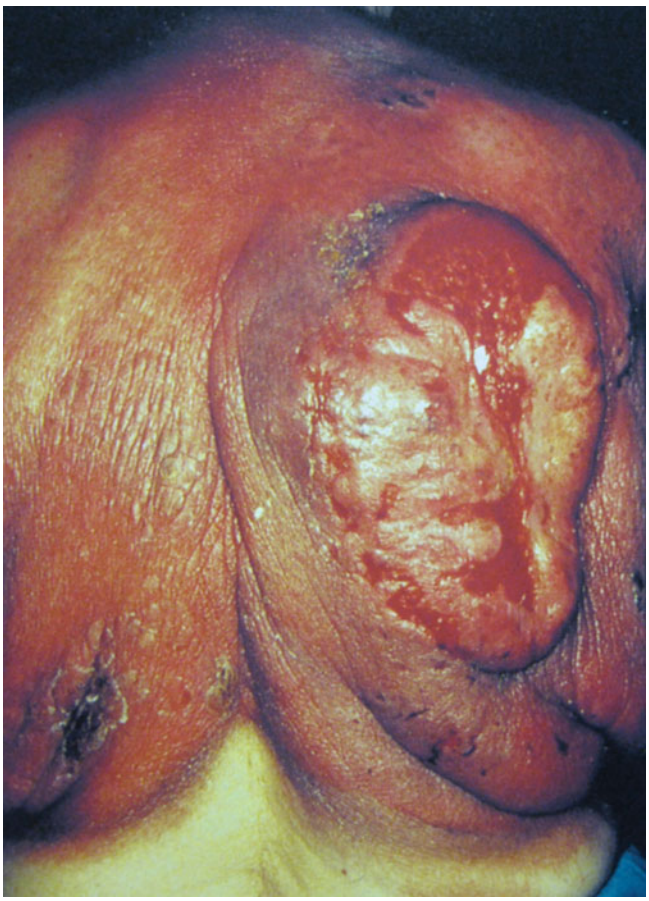


Fig. 47.9 Gigantic fungating tumor of mycosis fungoides. Differential diagnosis includes dermatofibrosarcoma protuberans

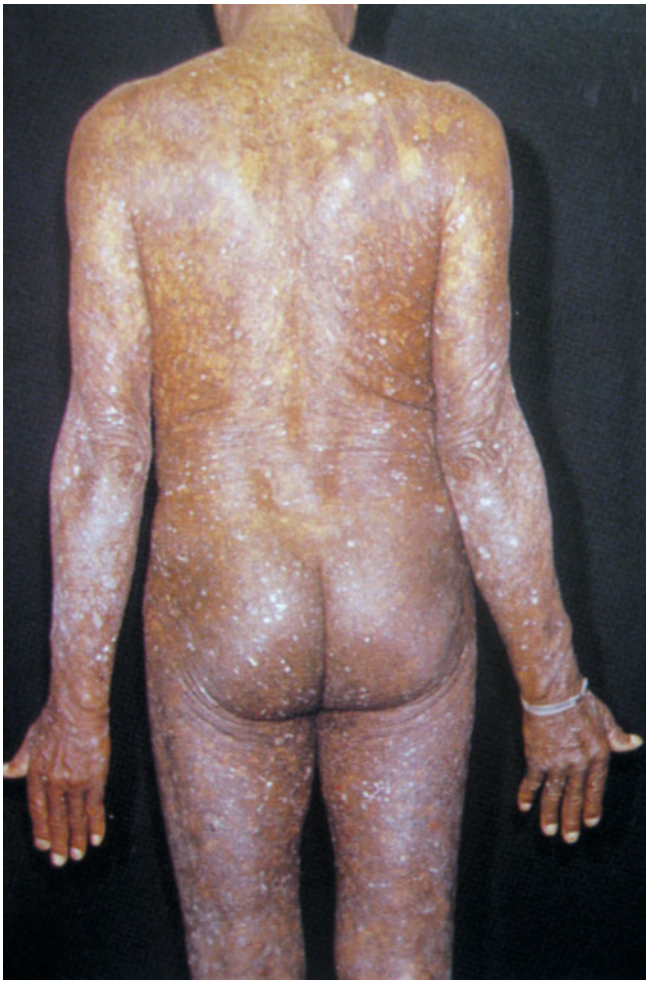


Fig. 47.10 Sezary syndrome is the blood borne erythrodermic form of mycosis *fungoides*

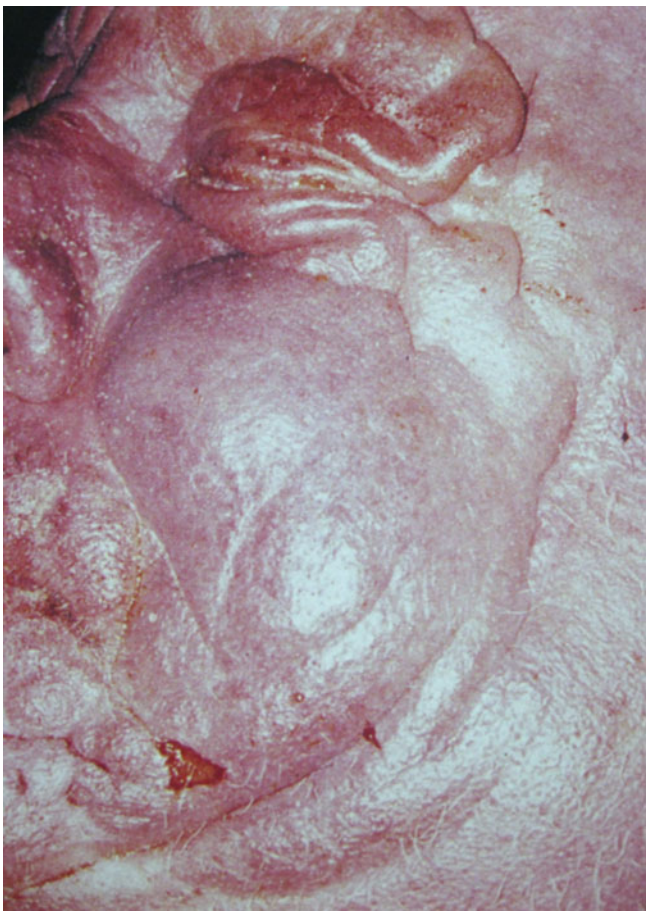
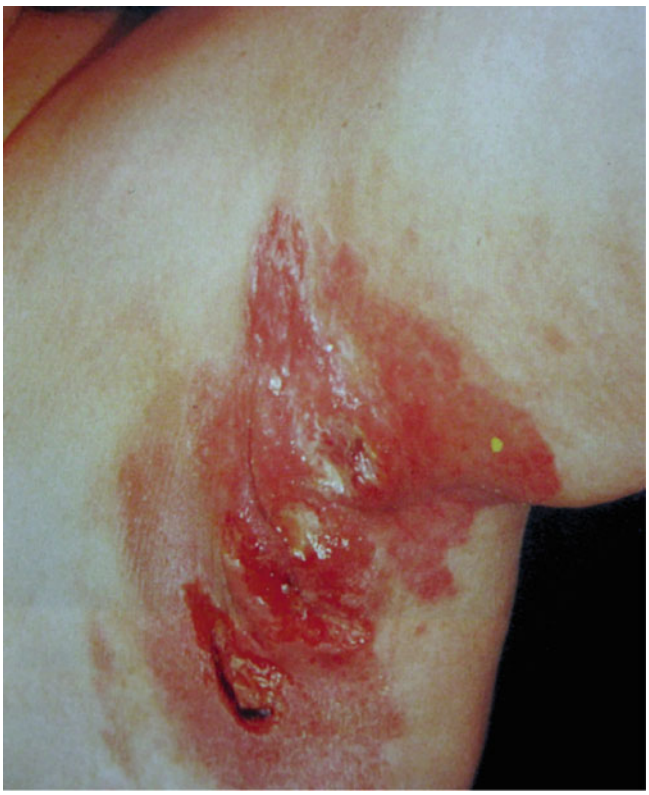
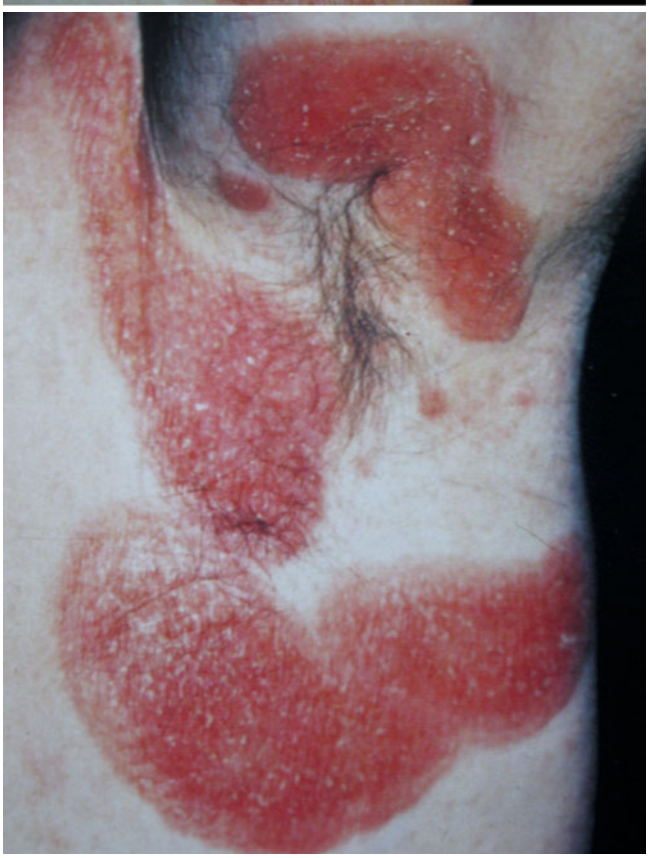
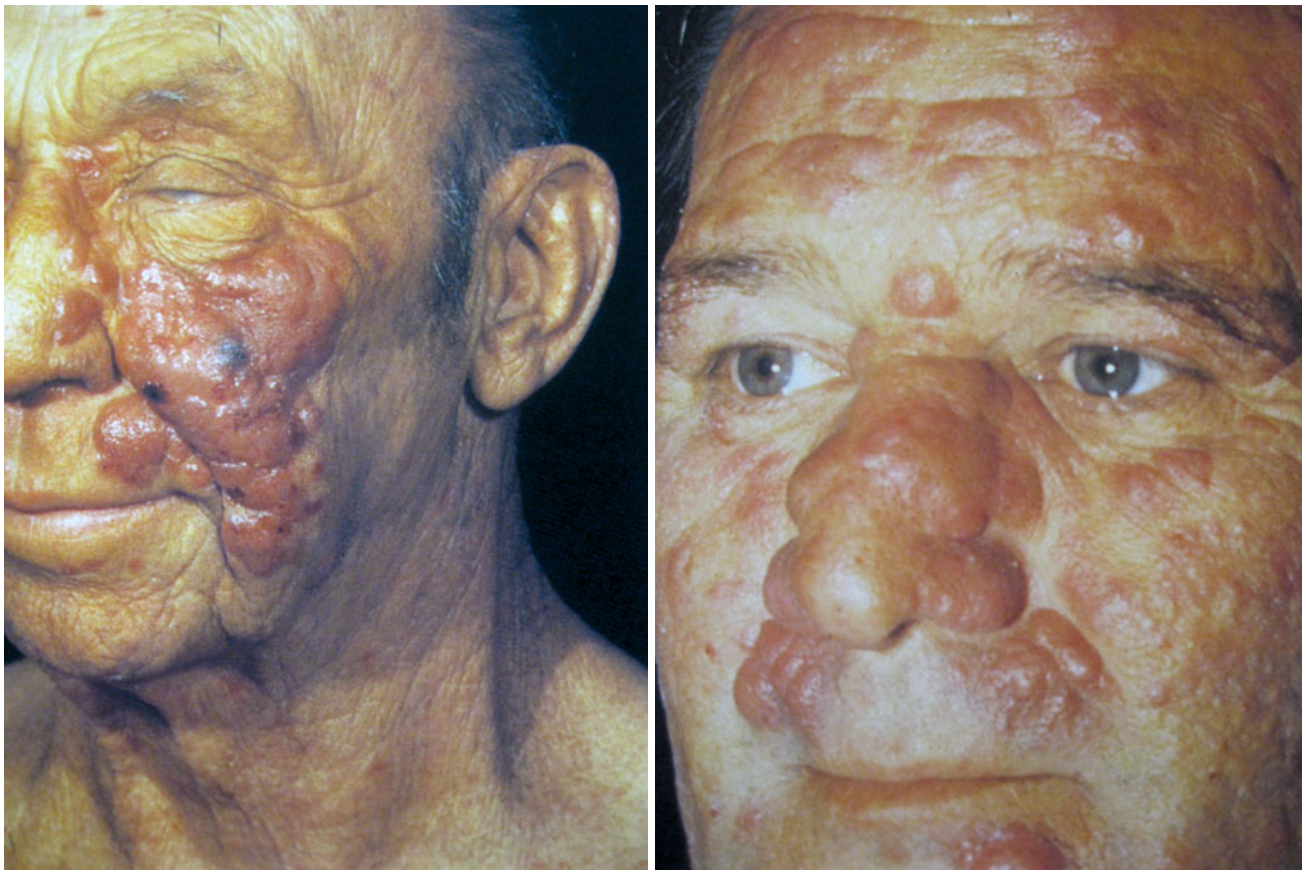


Fig. 47.11 Leonine (lion-like) face results from diffuse cutaneous infiltration by lymphoma in a patient with Sezary syndrome (Courtesy of Dr. R. Kanas)





Figs. 47.12 and 47.13 Mycosis fungoides of the axilla



Figs. 47.14 and 47.15 Mycosis fungoides of the face

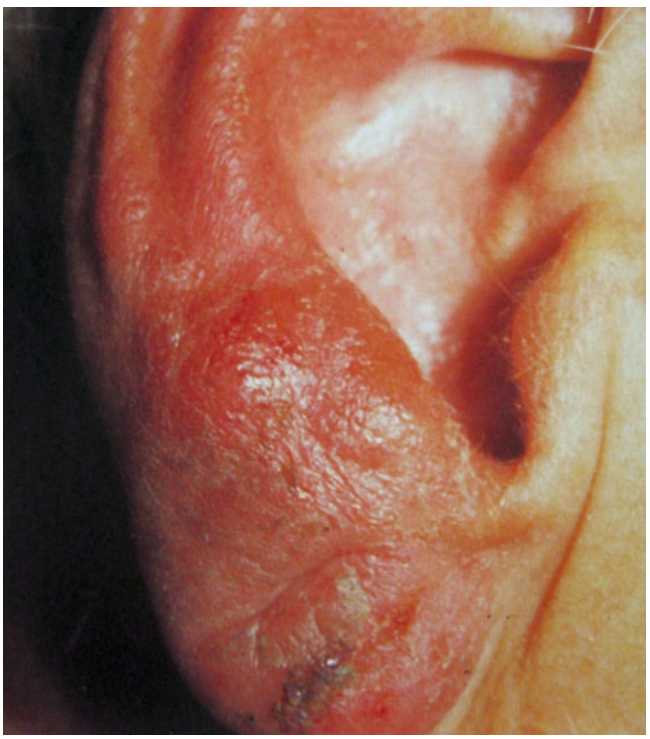


Fig. 47.16 Infiltration of the earlobe by mycosis fungoides is fairly common

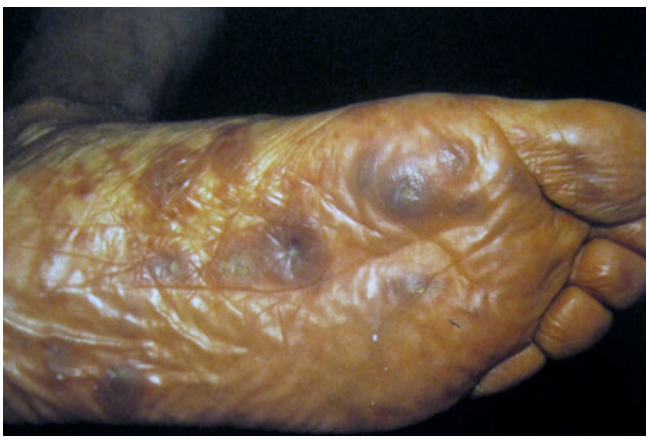


Fig. 47.17 Mycosis fungoides of the sole

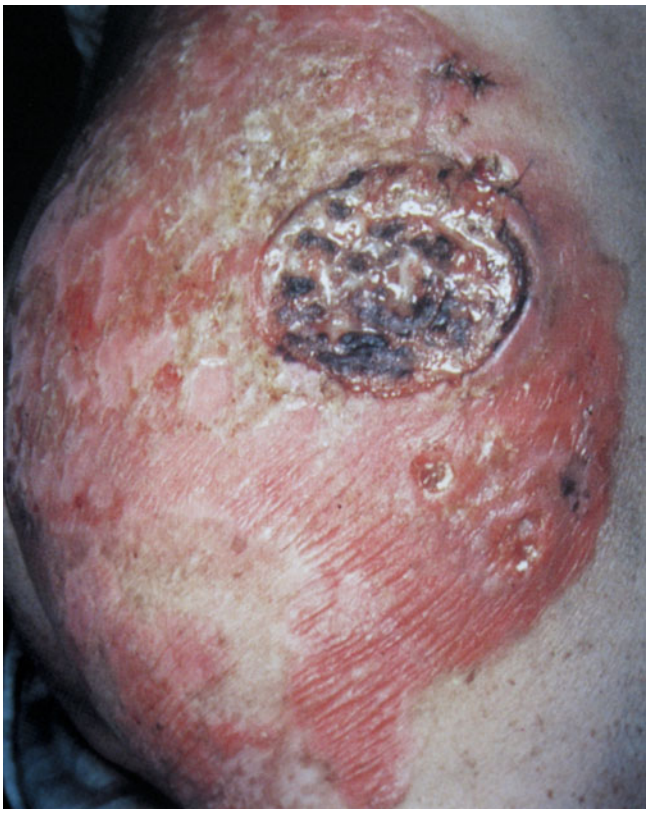


Fig. 47.18 D'ernblee type of mycosis fungoides-sudden appearance of tumors

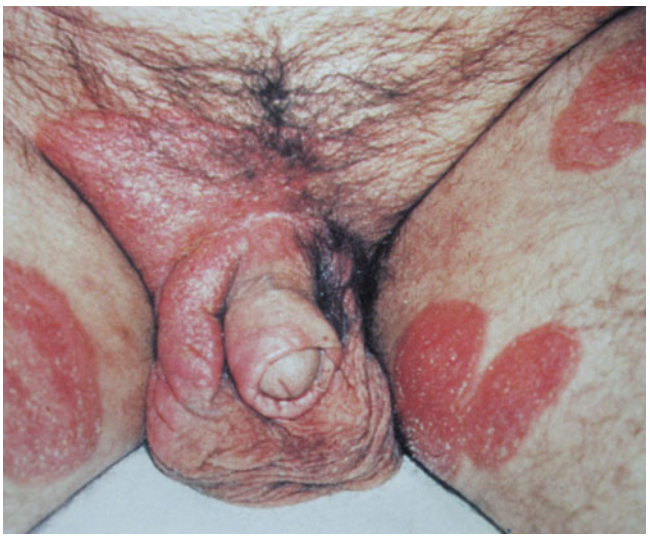


Fig. 47.19 Mycosis fungoides of the groin was mistakenly treated for years as tinea cruris (ringworm)

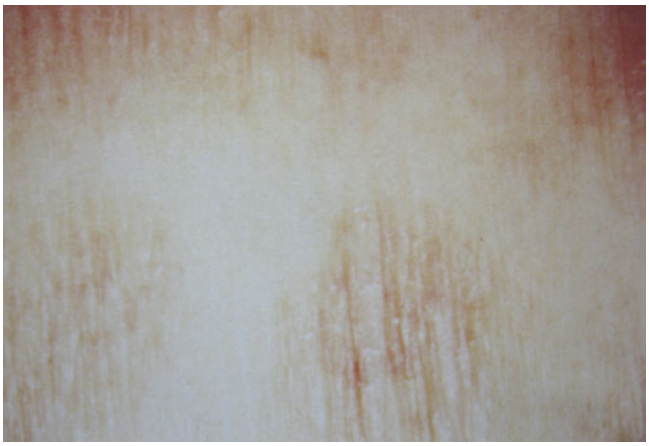


Fig. 47.20 Poikiloderma atrophicans vasculare. Atrophic, erythematous patches resemble those of eczema, tinea, and psoriasis. This form of parapsoriasis occasionally progresses to mycosis fungoides

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