Cutaneous lymphomas comprise a spectrum of diseases characterized by infiltration of the skin by malignant lymphocytes. The clinical manifestations of cutaneous lymphomas vary, and they can mimic benign dermatoses.

Cutaneous lymphomas comprise a heterogeneous spectrum of B- and T-cell lymphoproliferative disorders. More than 10 discrete clinicopathologic entities have been identified in both the revised European-American classification of lymphoma (REAL) and the European Organization for Research and Treatment of Cancer (EORTC) classification for primary cutaneous lymphomas.[1,2] The thoughtful review written by Dr. Abd-el-Baki and colleagues highlights the clinical manifestations of the most common subtypes and includes histopathologic, immunologic, and molecular characterization of these processes.

The recently published EORTC classification is based on 626 patients with a primary cutaneous lymphoma registered with the Dutch Cutaneous Lymphoma Working Group between 1986 and 1994. The frequencies and 5-year disease-related survival rates for the cutaneous T-cell lymphomas were: mycosis fungoides (44%; 87%), pagetoid reticulosis (<1%; 100%), granulomatous slack skin (<1%; -), CD30-positive large T-cell lymphoma (9%; 90%), CD30-negative large T-cell lymphoma (5%; 15%), Szary syndrome (2%; 11%), pleomorphic small/medium T-cell lymphoma (3%; 62%), and subcutaneous panniculitis-like T-cell lymphoma (<1%; 0%).[2] The frequencies and 5-year disease-related survival rates for the cutaneous B-cell lymphomas were: primary cutaneous follicular center cell lymphoma (13%; 97%), immunocytoma (marginal zone B-cell lymphoma [2%; 100%]), large B-cell lymphoma of the leg ((3%; 58%), intravascular large B-cell lymphoma (<1%; 50%), and primary cutaneous plasmacytoma (<1%; 100%).[2]

### Cutaneous T-Cell Lymphomas

The primary cutaneous T-cell lymphomas that typically run an indolent course include mycosis fungoides and its variants, CD30-positive cutaneous large T-cell lymphoma, and lymphomatoid papulosis.

Mycosis fungoides are most frequently treated with skin-targeting approaches, including the topical application of chemotherapy (nitrogen mustard [mechlorethamine] or carmustine [BCNU]), photochemotherapy (psoralen plus ultraviolet A [PUVA]), or radiotherapy.[3] The use of chemotherapy, interferon, retinoids, photophoresis, and recombinant toxins outside of the setting of a clinical trial is reserved for patients with refractory disease and for systemic manifestations. Although prolonged palliation can be achieved, few patients are cured.

Primary cutaneous CD30-positive large T-cell lymphomas are treated with radiotherapy when solitary or when localized skin lesions are present. Combination chemotherapy is utilized for patients who have generalized skin involvement or extracutaneous dissemination.[4]

Lymphomatoid papulosis often requires no therapeutic intervention. If generalized lesions are present or recur frequently, PUVA, topical chemotherapy, or low-dose methotrexate can provide palliation.[5]


The optimal therapy for Szary syndrome has not been clearly defined. Treatment with chemotherapy, interferon, retinoids, photophoresis, and/or radiation often provides short-term palliation.[3]

Primary CD30-negative cutaneous large T-cell lymphoma is treated with combination chemotherapy, with radiation therapy added for patients with solitary lesions or localized disease.[6]

Pleomorphic small/medium-sized cutaneous T-cell lymphoma, when localized, is treated with
radiation. Generalized disease can be palliated with chemotherapy and/or interferon.[7]
The treatment of subcutaneous panniculitis-like T-cell lymphoma remains inadequate. This disease
has an extremely poor prognosis despite the use of aggressive combination chemotherapy
regimens.[8]

**Cutaneous B-Cell Lymphomas**

Cutaneous B-cell lymphomas with an indolent course include primary cutaneous follicular center cell
lymphoma and immunocytoma. Localized primary cutaneous follicular center cell lymphoma is
treated with radiation, whereas more generalized skin involvement may be palliated with
chemotherapy, steroids, interferon and/or anti-CD20 monoclonal antibody-based therapy.[9]
Immunocytomas can be excised surgically and/or treated with radiation.[10]

Cutaneous B cell lymphomas with an intermediate to aggressive clinical behavior include large B-cell
lymphoma of the leg, intravascular large B-cell lymphoma, and plasmacytoma. Combination
chemotherapy is utilized for most patients with large B-cell lymphoma of the leg, with radiation
therapy added for solitary or localized presentations.[11] Intravascular large B-cell lymphomas are
approached with combination chemotherapy, unfortunately with modest success.[12] Primary
cutaneous plasmacytomas have an excellent prognosis after surgical excision or radiotherapy.

**Evaluation and Treatment Strategies Still in Evolution**

Strategies for evaluating and treating patients with cutaneous lymphomas continue to evolve. For
the majority of patients, these represent chronic diseases (> 80%) for which amelioration of
cosmetic consequences and symptoms (eg, pruritus) is critical.

Studies are underway through the Eastern Cooperative Group (ECOG) to determine treatment
strategies that result in better disease control. Cutaneous lymphomas have served as a paradigm for
the study of biological agents. Although significant palliation can be achieved with currently available
agents, the challenge is to develop therapies that provide prolonged control or cure.

**References:**


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