RESIDENT ROUNDS: PART II

Mycosis Fungoides and Variants: Board Review

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TABLE 1.

Indolent Types of Cutaneous T-cell Lymphomas[1]				
Disease	**Clinical**	**Epidemiology**	**Pathology and Immunohistochemistry[2]**	**Prognostic Factors[3,4]**
Classic Mycosis Fungoides (MF)[5,6]	Patches/plaques on non sun-exposed areas; may evolve into tumors; hypopigmented patches in children and skin of color	Age 55-60; African Americans [4,7]	"Haloed" cells, groupings of atypical lymphocytes in the epidermis; epidermotropism, Pautrier microabscesses; CD3+, CD4+, CD8-, CD30-	Poor prognosis: - Plaques > Patches - Older age -Male -Elevated LDH -African American Good prognosis: -Hypopigmented MF - MF associated with lymphomatoid papulosis -Poikilodermatous MF 5-year survival is stage dependent, with stage T1a having a 95-100% 5-year survival rate
Variants of MF				
- Folliculotropic MF	Acneiform lesions, comedomes, cysts, nodular prurigo-like lesions; head and eyebrow involvement; follicular papules	Males > Female, with age of onset later than males[8]	Perifollicular lymphocytic infiltrate with epidermal sparing; follicular destruction +/- follicular mucinosis, eosinophils; CD3+/CD4+/CD8- in most cases	5-year survival 77%
- Pagetoid reticulosis	Solitary or localized scaling patches or plaques, distal extremities.	All age groups	Epidermotropic atypical lymphocytes appearing as pagetoid cells, singly or in clusters, prominent epidermal hyperplasia; CD3+, CD4+/CD8- or CD4-/CD8+ T-Cell	More indolent course than classic MF, but can disseminate[9]
- Granulomatous slack skin[10]	Erythematous pendulous skin-intertriginous areas preceded by papules, plaques and patches	Middle aged male	Multinucleated giant cells and granulomatous infiltrate with loss of elastic fibers, prominent elastolysis, massive dermal and subcutaneous infiltrative patterns resembling sarcoidal, tuberculoid, periadnexal and granuloma annulare-like lesions; CD3+/CD4+/CD8-	Slowly progressive w/ 5-year survival 66%; associated with non-Hodgkin Lymphoma[11]
CD30+ lymphoproliferative disorders				
- Primary cutaneous anaplastic large cell lymphoma	Solitary or localized tumors or nodules. May regress spontaneously	Males>Females, 6th decade; rare in children	Reed-Sternberg like cells in dermis and subcutaneous fat.	5-year survival >90%, relapses common; extracutaneous manifestations indicate poor prognostic sign. Subset of patients with extensive limb disease (ELD) have worse prognosis[12]

Indolent Types of Cutaneous T-cell Lymphomas[1] *continued*				
Disease	**Clinical**	**Epidemiology**	**Pathology and Immunohistochemistry[2]**	**Prognostic Factors[3,4]**
- Lymphomatoid papulosis	Ulcerated nodules and papules on trunk and extremities	All ages, typically 4th decade of life	Type A: CD30+ Reed-Sternberg like cells (most common) Type B: CD3+/CD30- epidermotropic small lymphocytes (similar to MF) Type C: dermal CD30+ Reed-Sternberg like cells (similar to Anaplastic Large Cell Lymphoma) Type D: Markedly epidermotropic CD8+/CD30+ (mimic Primary cutaneous CD8+ aggressive epidermotropic CTCL) Type E: Angioinvasive CD30+ (mimics Extranodal NK/T-Cell Lymphoma, nasal type)	Spontaneously regressing; associated with lymphoproliferative disorders; 5-year survival 98%
Subcutaneous panniculitis- like T cell lymphoma (SPTCL)	Deep indurated nodules and plaques more commonly located on the lower extremities	Young adults	Lymphocyte atypia, necrosis, and lymphocyte "rimming" of individual adipocytes by CD8+ cytotoxic T cells; histologic differential is Lupus Panniculitis	T cells expressing α/β(βF1-positive) rather than γ/δ (βF1-negative) have 5-year survival > 80%; associated with fatal hemophagocytic syndrome

TABLE 2.

Aggressive Types of Cutaneous T-cell Lymphomas[1]				
	Clinical	**Epidemiology**	**Pathology and Immunohistochemistry**	**Prognostic Factors**
Sezary Syndrome (SS)[13]	Leukemic variant of MF characterized by: pruritic erythroderma, Sezary cells, generalized lymphadenopathy, +/- palmoplantar keratoderma; Sezary count ≥ 1000 cells/microliter	Males>Females, Caucasian predominance	Circulating Sezary cells (CD41 CD7; CD26), peripheral blood lymphocytes with CD4/CD8 ratio >10	Rapidly progressive clinical course and poor prognosis. Average 5-year survival is 42%
Adult T-Cell Leukemia/ Lymphoma[14]	Human T-Cell Leukemia Virus Type 1 (HTLV-1) is clonally integrated into neoplastic cells. Skin involvement in ~50% of patients, especially in chronic and smoldering subtypes. Skin involvement can resemble MF/SS. Hypercalcemia, organomegaly, osteolytic bone lesions.	Japan, Caribbean, Southeastern USA, Central Africa	Multilobed T-Cells; CD3+/CD4+/CD8-/CD25+ neoplastic cells in lymph nodes, blood and skin	Mean survival < 2 years
Extranodal NK/T-Cell Lymphoma, nasal type	Dermal or subcutaneous papules or nodules that can ulcerate, typically on the lower extremities or nose	USA, Korea	Epstein-Barr virus in tumor cells; dense infiltrate in dermis and subcutaneous fat sometimes involving the epidermis; angiodestruction and zonal necrosis; CD2+ CD3epsilon+ CD56+	Poor prognosis
Primary cutaneous gamma delta T-Cell Lymphoma[15]there are few clinicopathological studies. In addition, T-cell receptor (TCR	Plaques, ulcerated nodules and tumors on extremities. May or may not have SPTCL.	Rare	T-Cell Receptor gamma-delta+ Beta F1- CD3+ CD56+ CD4-	5-year survival 42.4% without SPTCL features and 80.0% with SPTCL features; associated with hemophagocytic syndrome.

*Survival data based on 5-year disease specific survival where available.

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DISCLOSURES

None of the authors have declared any relevant conflicts.

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