Rare variant of mycosis fungoides

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Cutaneous lymphomas represent a heterogeneous group of T-, NK- and B-cell neoplasms, with mycosis fungoides (MF) being the most common subtype.[1] Clinically, the disease is typified by gradual progression from patches (flat, scaly, various shades of red, variably pruritic) and plaques (indurated, often annular with central clearing), mostly on photoprotected sites, to tumors. Unusual clinical and histopathologic variants often coexist with typical patches or plaques. These clinical variants include follicular, syringotropic, vesicular, granulomatous, poikilodermic, hypo- and hyperpigmented, palmoplantar, hyperkeratotic, papillomatous, ichthyosiform, pigmented purpura-like, pustular and with mucosal involvement. Microscopically, patch-stage MF exhibits a sparse papillary dermal lymphocytic infiltrate with epidermotropism and atypical lymphocytes, either in collections or singly within the epidermis and these are probably the most specific finding in early MF. Plaques of MF, in contrast, are usually diagnostic, exhibiting all of the findings of patches, and more, including[1] deeper and denser dermal infiltrates,[2] cytologically atypical intraepidermal lymphocytes, [3] easily found Pautrier's microabscesses, and more prominent psoriasiform hyperplasia and papillary dermal fibrosis.[4] Immunohistochemical studies are essential to confirm the diagnosis.[2] In the following patient an unusual variant of MF was diagnosed after years of non-remitting disease.

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Case

A 20-year-old Arabian female presented to the Dermatology Clinic with a 3-year history of persistent, symptomless islands of prominent follicular papules on the trunk. Past medical history was irrelevant and family history was negative for similar conditions. In the past she received topical steroids and emollients with no improvement. On examination of the skin, the patient had islands of grouped prominent follicular papules with no change in the color of the skin (Figure 1). Systemic examination was normal. Complete blood count, liver function, renal function tests and chest x-ray were normal. A 4-mm punch biopsy specimen from one of the lesions on the trunk showed a follicular-centric infiltrate with small- to medium-sized lymphocytes in the follicular root sheath epithelium. In addition, there were areas of focal epidermal lymphocytic infiltrate. Immunocytochemistry showed lymphocytes with positive T-cell markers of CD45RO and CD3. Polymerase chain reaction (PCR) on the paraffin block revealed positive clonal rearrangement of the T-cell receptor gene so the diagnosis of follicular MF was made. The patient was treated with topical retinoic acid and was periodically evaluated in the clinic.

Discussion

MF is a peripheral non-Hodgkin T-cell lymphoma initially presenting in the skin.[3] It is classified as an indolent lymphoma by the EORTC.[4] However, most patients with patches or plaques never have disease

progression to the tumor or erythrodermic stages. Besides, erythroderma may intervene at any time, and its distinction from Sézary syndrome (SS) depends on the findings in peripheral blood and other clinical features. MF may present at any stage and unusual clinical and histopathologic variants often coexist with typical patches or plaques. The common denominator among all variants is the presence of diagnostic histologic findings.[2] Follicular MF is a rare variant of MF in which malignant lymphocytes selectively surround and infiltrate hair follicles in the absence of epidermal invasion or follicular mucin.[5] Clinically patients with follicular MF present with plaques, erythematous papules and comedo-like lesions.[6] Our patient is the youngest (22 years old) case of MF reported in the literature (age range 39-65).[5] It has been suggested that the relationship of follicular MF to ordinary MF mirrors the relationship of lichen planopilaris to lichen planus.[7] It has also been suggested that certain intercellular adhesion receptor systems may play a role in the folliculotropism seen in follicular MF. In other words, intercellular adhesion molecule-1 (ICAM-1) has a high affinity for lymphocyte function associated antigen-1 (LFA-1) positive cells, which are prominent in infiltrates of cutaneous T-cell lymphoma, as Gilliam et al reported.[5] The stimulus for the upregulation of ICAM-1 in the hair follicle in cases of follicular MF is still unknown. In fact, establishing or excluding a de novo diagnosis of cutaneous lymphoma is complicated and difficult. Diagnosis requires all available clinical, pathologic, immunohistochemical, and cytogenetic findings. Even after all available data have been obtained, a definitive, specific, and prognostically or therapeutically relevant diagnosis may remain elusive.

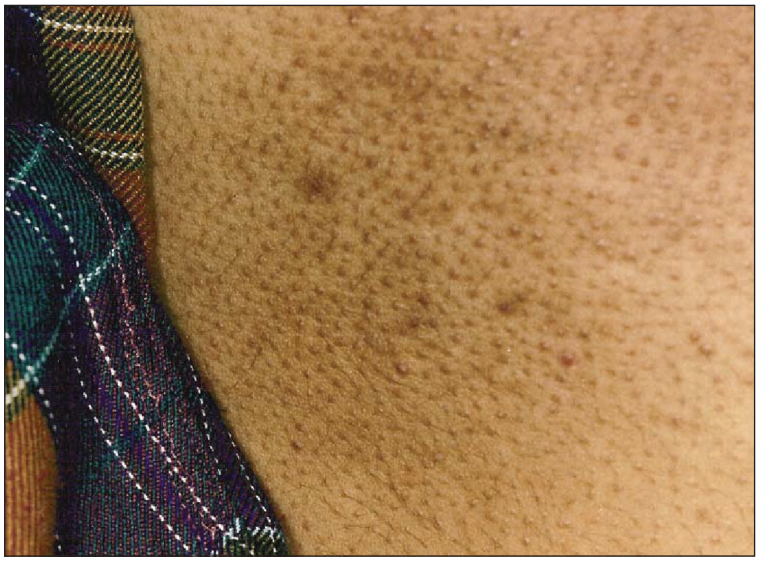

Figure 1: Prominent follicular papules on the trunk.

Immunohistochemical studies are essential for distinguishing B, T, natural killer (NK), and non-lymphoid cells and their subsets based on their immunophenotype. MF cells variably express T-cell markers (CD2, CD3, CD5, CD7, Leu-8, CD45RO), the hallmark being the T helper/inducer subset marker CD4. Therapy for CTCL is currently dependent on the clinical stage rather than on the specific subtype and it is oriented toward achieving palliation in advanced cases. However, radiotherapy is the single most reliable method for inducing complete clinical remission in MF.[8] Besides, it is essential to know that the unusual histology of follicular MF with its deeper follicular lymphocytic infiltration, may decrease the efficacy of poorer penetrating topical agents thus complicating treatment plans.[9]

References

1. Jaffe ES, Harris NL, Stein H, Vardiman JW (eds) World Health Organization Classification of Tumours. Pathology and Genetics of Tumours of Haematopoietic and Lymphoid Tissue. *IARC Press, Lyon,* 2001.
2. Fung M, Murphy M, Hoss D, Grant-Kels J. Practical evaluation and management of cutaneous lymphoma. *J Am Acad Dermatol.* 2004;46:325-57.
3 Isaacson PG, Norton AJ. Cutaneous lymphomas. In: Isaacson PG, Norton AJ, eds. Extranodal Lymphomas. Churchill Livingstone, London,1994:131-191.
4. Kazakov DV, Burg G, Kempf W. J Eur Acad Dermatol Venereol 2004;18:397-415.
5. Dbloom J, Severson J, Gaspari A, Scott G. Follicular mycosis fungoides: a case report and review of the literature. J Cut Pathol 2001;28:318-24.
6. Kim SY. Follicular mycosis tuogoides. Am J Dermatopathol 1985; 7: 300.
7. Goldenhcrsh MA, Zlotngorski A, Ruserunarut E. Follicular mycosis fungoides. Am J Dermatoparhol 1994; 16: 52.
8. Kaye FJ, Bunn PA, Steinberg SM, Stocker JL, Ihde DC, Fischmann AB, et al. A randomized trial comparing combination electron-beam radiation and chemotherapy with topical therapy in the initial treatment of mycosis fungoides. N Engl J Med 1989;321:1784-90.
9. Jackow- CM, Cather JC, Hearne V, Asano AT, Musser JM, Devic M. Association of crythrodermic cutaneous T-cell lymphoma, superantigen-positive staplrylococctts aureus, and oligoclonal T-cell receptor v{} gene expansion. Blood 1997;89:32.