DERMATOPATHOLOGY

FOLLICULAR MYCOSIS FUNGOIDES PRESENTING WITH SPIKY/KERATOSIS PILARIS-LIKE LESIONS: AN INDOLENT CLINICOPATHOLOGIC VARIANT OF FOLLICULAR MYCOSIS FUNGOIDES

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Introduction: It is generally accepted that follicular mycosis fungoides (FMF) is a distinct entity with an intrinsically worse prognostic factor. However, recently reporting on a few series of patients presenting only with spiky and/or keratosis pilaris-like hyperkeratotic papules and running an indolent course would argue this concept.

Objectives: To better categorize the clinicopathologic features and course of a subset of FMF patients presenting with keratosis-pilaris-like lesions.

Materials and Methods: The clinical notes of patients with a diagnosis of FMF seen during the previous 10 years at the Department of Dermatology of the University of Turin (Italy) were reviewed to identify any cases that at presentation had only hyperkeratotic follicular lesions.

Results: Sixteen patients (10 male, 6 female) with a mean age of 56 years were enrolled. Patients presented with disseminated, slightly erythematous, hyperkeratotic, spiky or cone-shaped follicular papules which, histopathologically, showed hyperkeratotic columns protruding from follicular plugging in concert with selective infiltration of the infundibular epithelium by atypical, mostly CD4+, lymphocytes. T-cell clonality was demonstrated in 12/16 cases. The mean duration of the lesions before diagnosis was 36 months. FMF was not a clinical consideration in any of these patients initially. The course was mostly indolent, whilst progression to overt FMF was noted in 2 patients (median follow up: 56 months). Skin-targeted therapy was the first line therapy in most patients, whilst systemic treatment with bexarotene was administered first in two patients.

Conclusions: Our study would suggest that the course of patients presenting with spiky

and/or keratosis pilaris-like lesions is indolent and similar to that of patch stage MF. A bias in the recognition of this clinicopathologic presentation of FMF and/or its intrinsically less aggressive behavior might explain the discrepancy in the prognosis between this subset of patients and those with overt FMF lesions.