Predicting the nature of follicular mucinosis: Still a sticky situation

Warren R. Heymann, MD
Marlton, New Jersey

In 1957, Hermann Pinkus presented 6 cases of alopecia characterized by the presence of mucin in the outer root sheath and sebaceous gland, calling the entity *alopecia mucinosa*.[1] Also known as follicular mucinosis (FM), the disorder is now considered as either primary FM (an idiopathic, benign form of the disease in children or young adults, with mostly facial lesions) or secondary FM (occurring in older patients and presenting in a more generalized distribution, with a more chronic course, and in association with other disorders, notably, cutaneous T-cell lymphoma [CTCL]).

In their discussion of the classification and diagnostic challenges of determining whether FM is idiopathic or due to mycosis fungoides (MF), Hooper et al assert that (1) an isolated patch in the head and neck region is much more likely to be idiopathic FM than MF, (2) monoclonality of T-cell gene rearrangement studies does not reliably distinguish FM from MF, and (3) none of the clinicopathologic features of FM or MF are without overlap. The standard assessment tools of T-cell gene rearrangement studies, flow cytometry, and immunohistochemistry must be utilized with close clinical follow-up in the diagnostic process because "no single diagnostic tool is sufficient in providing diagnostic certainty; rather, a collective evaluation of pathologic, molecular, and clinical criteria is required."[2]

Recent examples demonstrating why individual patient assessment is of paramount importance include (1) a 9-year-old boy presenting with an enlarging patch of alopecia on his scalp over 2 months and lichen spinulosus–like plaques on his trunk that were proved to be MF with FM[3] and (2) increasing reports of drug-induced FM, as exemplified by the cases due to infliximab, imatinib, oxcarbamazepine, captopril, and dextromorphan.[4]

FM in association with hematologic disorders other than MF has not been well defined. In this issue of the *Journal of the American Academy of Dermatology*, Geller et al describe clinicopathologic characteristics of FM in such patients. A total of 18 patients with FM and systemic hematologic malignancies without CTCL were identified; 9 of the malignancies occurred after hematopoietic stem cell transplantation. No cases of non–CTCL-associated FM (n = 46; 37 biopsy specimens) developed CTCL during a mean follow-up of 4.3 years. In the cases of CTCL associated with FM (n = 44; 31 biopsy specimens), MF was the most common subtype (n = 38), although other CTCLs were identified. FM in patients with non-CTCL hematologic malignancies differed clinically from MF-associated FM, presenting most frequently with erythematous papules, without plaques, without alopecia, and without histopathologically identified epidermal exocytosis (each of these features was statistically significant). The authors concluded that FM can present in patients with systemic hematologic malignancies, including those that develop after hematopoietic stem cell transplantation. A papular lesional morphology and histopathology may help to distinguish these cases from MF.[5]

In his seminal paper on alopecia mucinosa, Dr Pinkus opined, "Concerning the nature of these changes, only speculation can be offered."[1] More than 6 decades later, this is still apt. It is essential that dermatologists assess each patient with FM individually, looking at the patient's overall health and medication history, physical examination findings, histopathology, immunohistochemistry, and molecular study results, to determine the nature of that patient's FM.

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From the Division of Dermatology, Cooper Medical School of Rowan University, Marlton.
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Correspondence to: Warren R. Heymann, MD, 100 Brick Rd, Suite 306, Marlton, New Jersey 08053. E-mail: wrheymann@gmail.com.
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