

DERMATOPATHOLOGY UNIT

UPP | Department of Dermatology

UPMC Dermatopathology "Case of the Month" Presentations

UPP - Department of Dermatology, Dermatopathology Unit

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Case Authors: *Joanne Simpson MD, MPH, Larissa Geskin MD, Drazen M. Jukic, MD, PhD*

JUNE 2005 CASE OF THE MONTH

CLINICAL FINDINGS

Clinical History

56-year-old white male with a 10 month history of persistent generalized pruritic erythematous plaques, concentrated on mid abdomen, chest, and back. The patient was seen first nine months after the onset of this eruption in the general dermatology clinic. A skin biopsy was taken at that time to rule out Grover's disease versus Pityriasis Lichenoides et Varioliform Acuta (PLEVA) versus bites versus dermal hypersensitivity and was inconclusive, but suggestive of Grover's disease. Despite treatment with topical steroids, his lesions persisted. The patient was referred to our Contact Dermatitis clinic to be evaluated for allergic dermatitis, however no identifiable allergens were detected. Several weeks following patch testing, the eruption became more pruritic. The patient returned for evaluation and a repeat skin biopsy was performed to again rule out Grover's versus lymphomatoid papulosis..

Physical Exam

On examination, the patient had multiple erythematous papules, some with overlying excoriations, on his legs, chest, abdomen, and back. [See: FIGURE 1].

Histopathology

The initial biopsy demonstrated a moderately dense dermal infiltrate of mostly small but occasional large lymphoid cells admixed with histiocytes, neutrophils and few eosinophils with varying degrees of acantholysis and focal dyskeratosis. [Figure 2]

The repeat biopsy showed epidermal hyperplasia seen akin to seborrheic keratosis with foci of spongiosis and dyskeratosis, which by itself would be suggestive of Grover' s disease. However, there were numerous CD30+ large lymphocytes (also CD7, CD3, and CD5 positive and CD20 negative) seen in the dermis and epidermis. There was also prominent margination of neutrophils in the blood vessels with plump endothelium and eosinophils in the background. The changes, taken together with re-reviewed previous eosinophils in the background. The changes, taken together with re-reviewed previous case were deemed most suggestive of lymphomatoid papulosis (type 3A4). [Figure 3]

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Figures & Images

1. Click on the *Figure number* you wish to review.
2. Click on the *image* to enlarge

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Fig 1

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Fig 2

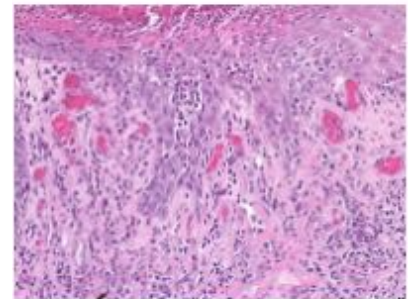
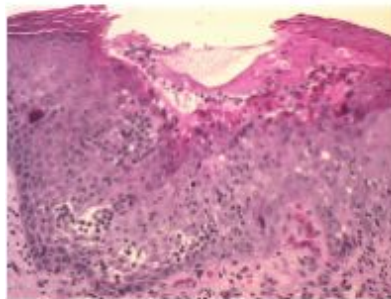


Fig 3

Fig 4

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DISCUSSION & DIAGNOSIS

Diagnosis

Lymphomatoid papulosis, Type 3A4

Discussion

Lymphomatoid papulosis (LyP) is an indolent chronic, recurrent lymphoproliferative disorder that lies in a spectrum of CD30-positive low-grade spectrum cutaneous T-cell lymphomas. The hallmark of these disorders is the presence of highly atypical tumor lymphocytes expressing CD30 (BerH2) antigen and similar T-cell receptor clonal rearrangement (1).

Clinically, LyP presents as recurrent crops of papulonodular or plaque like lesions, often with hemorrhagic crusting or ulceration most commonly seen on the trunk and extremities. Although its clinical appearance is benign and its prognosis is favorable, the histologic and cytologic features of LyP demonstrate features of malignancy. LyP has a variable histologic presentation. The most common form of LyP, LyP type A, is characterized by a wedge-shaped dermal infiltrate comprised of anaplastic lymphoid cells in a background of small lymphocytes, often with neutrophils, eosinophils, or both. Type B LyP is typified by lichenoid dermal infiltrate of small lymphocytes with epidermotropic infiltration of the epidermis in a manner that resembles mycosis fungoides. LyP Type C, on the other hand, demonstrates diffuse infiltration of the dermis by sheets of tumor cells and sparse neutrophils (2). Immunophenotypically, each of these forms of LyP demonstrates clonal, activated helper T cells with positive expression of CD3, CD4, CD5, and CD30 markers. These cells stain negatively for CD8, CD15, and EMA (3).

Grover's disease, also known as transient acantholytic dermatosis, demonstrates discrete pruritic, hyperkeratotic papules and papulovesicles seen mainly on the back and thighs of middle-aged to elderly males. Histologically, the hallmark of Grover's disease is the presence of focal acantholysis. Four variants of Grover's disease exist, Darier's disease-like (acantholytic dyskeratosis), Hailey-Hailey disease like (suprabasal cleft with significant acantholysis), pemphigus vulgaris-like (suprabasal acantholysis), pemphigus foliaceus-like (superficial acantholysis). Immunofluorescence and immunohistochemical studies have not demonstrated consistent patterns of positivity in this disease (2).

Grover's disease and lymphomatoid papulosis can often clinically resemble one another, however, these two entities are usually distinctive histologically. This case represents an unusual presentation of persistent, recurrent, pruritic papules in a 56 year-old man—a clinical presentation suggestive of Grover's disease, but with histologic findings of both Grover's disease and lymphomatoid papulosis, type A. To date, there are only 2 reports in the literature of a similar histologic pattern, in which there is significant epidermal atypia in association with CD30+ dermal infiltrate (4,5). Both of these reports included cases in which squamous cell carcinoma atypical lymphocytic proliferation with features of CD30+ lymphoma and LyP. Our patient's histopathologic findings of epidermal acantholysis superimposed on a wedge-shaped dermal infiltrate of atypical CD 30-lymphocytes may represent an atypical variant of lymphomatoid papulosis, in which the epidermal component resembles Grover's disease.

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