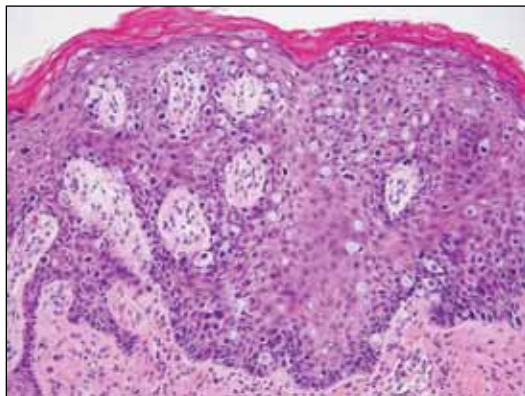


H&E, original magnification $\times 10$.



H&E, original magnification $\times 20$.

The best diagnosis is:

- a. benign acral nevus
- b. Bowen disease
- c. extramammary Paget disease
- d. mycosis fungoides
- e. superficial spreading melanoma in situ

PLEASE TURN TO PAGE 315 FOR DERMATOPATHOLOGY DIAGNOSIS DISCUSSION

Michael Zaleski, BS; Tammie Ferringer, MD

From Geisinger Medical Center, Danville, Pennsylvania.

The authors report no conflict of interest.

Correspondence: Tammie Ferringer, MD, Geisinger Medical Center, 100 N Academy Ave, Danville, PA 17821 (tferringer@geisinger.edu).

Extramammary Paget Disease

Extramammary Paget disease (EMPD) is an uncommon condition that usually presents in apocrine sweat gland-rich areas, most commonly the vulva followed by the perianal region. Lesions clinically present as erythematous, well-demarcated plaques that may become ulcerated, erosive, scaly, or eczematous. Extramammary Paget disease has a female predominance and usually occurs in the sixth to eighth decades of life.¹ Histologically, EMPD displays intraepidermal spread of large cells with plentiful amphophilic cytoplasm and large nuclei (Figure 1). These atypical cells may be seen “spit out” whole into the stratum corneum rather than keratinizing into parakeratotic cells (Figure 2). Frequently, the cytoplasm of these tumor cells is positive on mucicarmine staining, which indicates the presence of mucin, giving the cytoplasm a bluish gray color on hematoxylin and eosin–stained sections. Typically, EMPD cells can be found alone or in nests throughout the epithelium. The basal layer of the epithelium will appear crushed but not infiltrated by these atypical cells in some areas.² Extramammary Paget disease is epithelial membrane antigen and cytokeratin 7 positive, unlike other conditions in the differential diagnosis such as benign acral nevus, Bowen disease, mycosis fungoides, and superficial spreading melanoma in situ, with the rare exception of cytokeratin 7 positivity in Bowen disease.³

Benign acral nevi, similar to melanoma in situ, can have melanocytes scattered above the basal

layer, but they usually appear in the lower half of the epidermis without cytologic atypia.⁴ When present, these pagetoid cells are most often limited to the center of a well-delineated lesion. The compact thick stratum corneum characteristic of acral skin also is helpful in distinguishing a benign acral nevus from EMPD, which does not involve acral sites (Figure 3).²

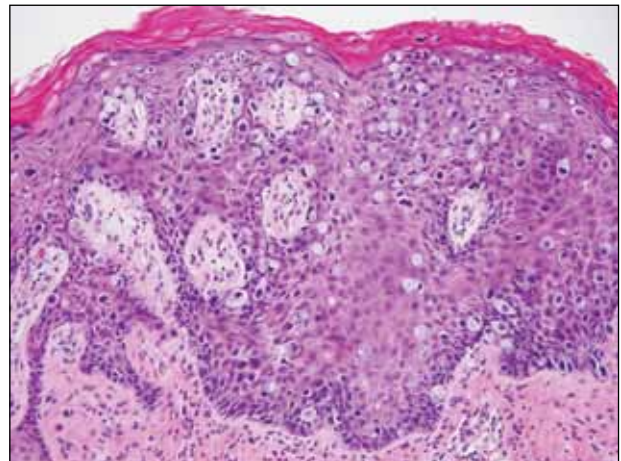


Figure 2. Atypical cells of extramammary Paget disease “spit out” whole into the stratum corneum rather than keratinizing into parakeratotic cells (H&E, original magnification $\times 20$).

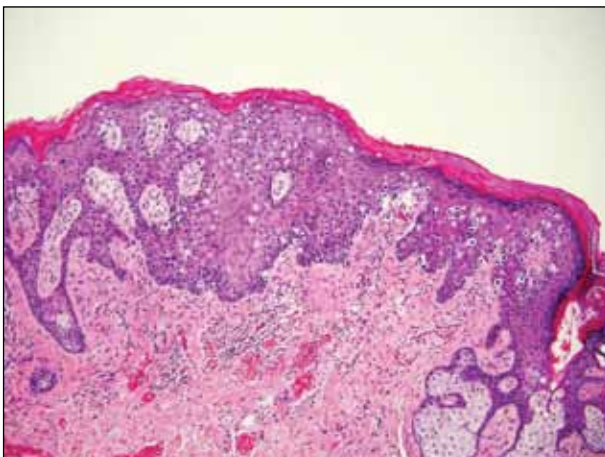


Figure 1. Intraepidermal spread of large cells with plentiful amphophilic cytoplasm in extramammary Paget disease (H&E, original magnification $\times 10$).

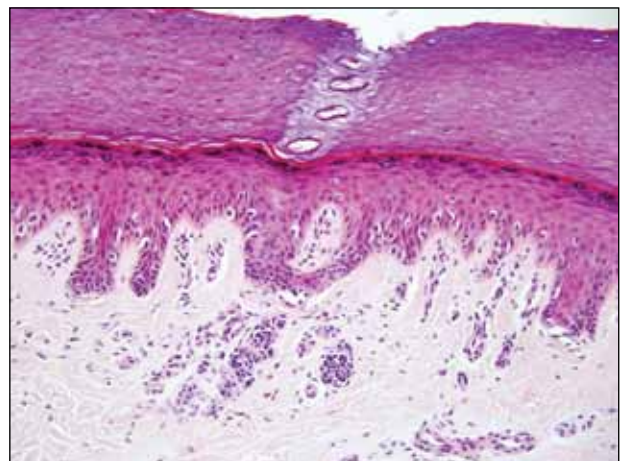


Figure 3. Cytologically normal melanocytes in the lower half of the epidermis in a benign acral nevus with a thick stratum corneum that is indicative of acral skin (H&E, original magnification $\times 20$).

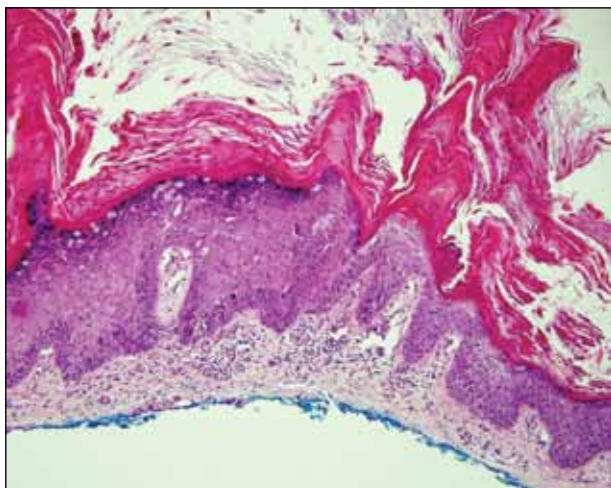


Figure 4. Parakeratosis and pagetoid spread of cells adjacent to areas of full-thickness epidermal keratinocyte atypia in Bowen disease (H&E, original magnification $\times 10$).

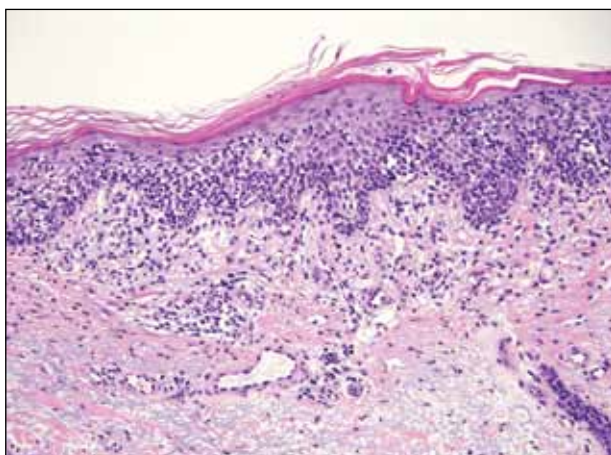


Figure 5. Atypical lymphocytes in the epidermis of mycosis fungoides display nuclei that are large, dark, and have irregular nuclear contours with perinuclear clearing (H&E, original magnification $\times 20$).

Bowen disease (squamous cell carcinoma in situ) may have pagetoid spread (or buckshot scatter) through the epidermis similar to EMPD and melanoma in situ. However, in Bowen disease the malignant cells are keratinocytes that keratinize and become incorporated into the stratum corneum as parakeratotic nuclei rather than intact “spit out” cells, as seen in melanoma in situ and EMPD. Usually the pagetoid spread is only focal in Bowen disease with other areas of more characteristic full-thickness keratinocyte atypia (Figure 4).²

Mycosis fungoides displays atypical lymphocytes with large dark nuclei and minimal to no cytoplasm

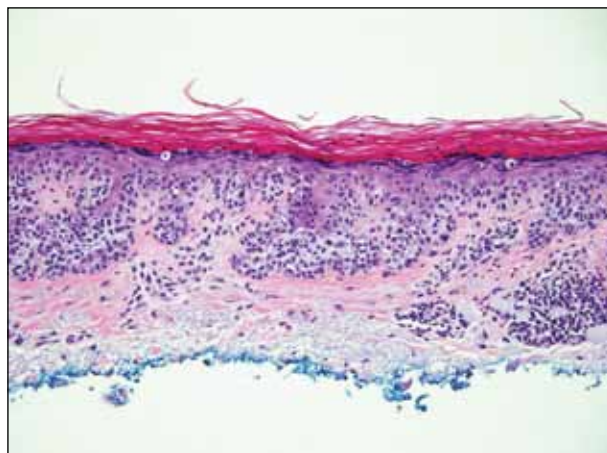


Figure 6. Nests of atypical melanocytes predominately involving the basal layer in superficial spreading melanoma in situ (H&E, original magnification $\times 20$).

scattered throughout the epidermis. The atypical cells have irregular nuclear contours and often a clear perinuclear space (Figure 5). These cells tend to line up along the dermoepidermal junction and form intraepidermal clusters known as Pautrier microabscesses. Papillary dermal fibroplasia also is usually present in mycosis fungoides.²

Similar to EMPD, superficial spreading melanoma in situ shows single or nested atypical cells scattered throughout all levels of the epithelium and may be “spit out” whole into the stratum corneum rather than keratinizing into parakeratotic cells. However, in melanoma, nests of atypical melanocytes predominate and involve the basal layer (Figure 6), whereas clusters of cells in EMPD typically are located superficial to the basal layer. The cells of melanoma also lack the amphophilic mucinous cytoplasm of EMPD.¹

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