

4.3.9 Paget disease

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| Definition | <ul style="list-style-type: none">• <i>Mammary Paget disease</i>: Intraepidermal spread of an underlying, usually intraductal, carcinoma of the breast• <i>Extramammary Paget disease</i>: Intraepidermal spread of cutaneous adnexal carcinomas or of a metastatic carcinoma (colon, prostate, bladder) |
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Clinic	Erythematous plaque, sometimes eroded, present either on nipple or in axillae, groin or anogenital region for the extramammary variant. Often clinically mistaken for dermatitis or candidiasis in groin
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Histopathology

- Pagetoid (scattered, like pattern of shot gun) distribution of solitary or nested *epithelial tumor cells* in often acanthotic epidermis
- *Tumor cells typically with pale basophilic* cytoplasm, enlarged pleomorphic nuclei and prominent nucleoli
- Lymphocytic infiltrate in upper dermis

Additional studies

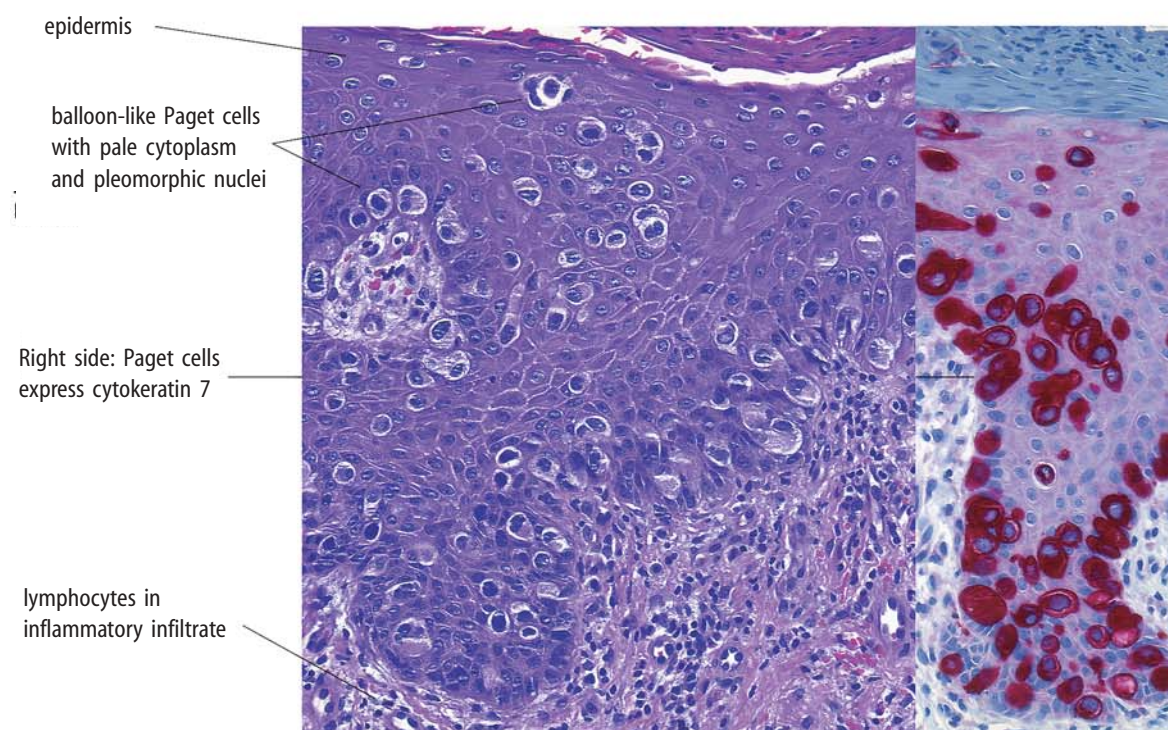
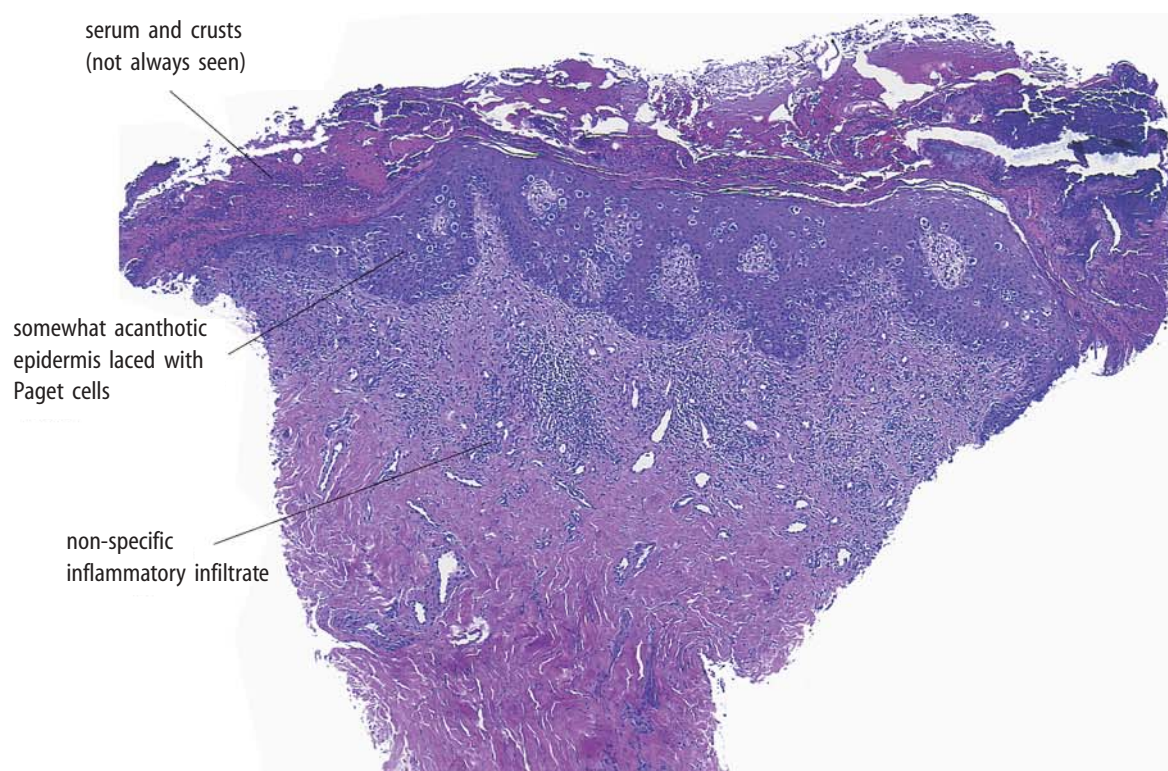
The tumor cells are usually PAS-positive but immunohistochemical studies are mandatory. The tumor cells react with CAM5.2, CK7, EMA and gross cystic disease fluid protein-15. HER2 (ErbB2) is a sensitive marker of tumor activity which can be expressed in all forms of Paget disease, not just those associated with breast carcinoma. Markers in extramammary Paget disease depend on underlying tumor (CDX2 for colon, PSA for prostate and uroplakin for bladder carcinomas)

Differential diagnoses

- **Melanoma (superficial spreading type)**
Epidermis features atypical, sometimes clear melanocytes both singly and in nests, usually more prominent in basal layer, and positive for S-100, HMB-45 or Melan A
- **Bowen disease and Bowenoid papulosis**
Acanthosis with full-thickness epidermal maturation defects, atypical mitoses, clumped nuclei
- **Pagetoid reticulosis**
Localized form of mycosis fungoides (Sec. 4.6.1) with atypical clear lymphocytes in pagetoid intraepidermal pattern
- **Periorbital sebaceous carcinoma**
Sebaceous carcinomas of the eyelid usually have replacement of epidermis by pale sebaceous cells with abundant cytoplasm

Comment

Most cases of extramammary Paget disease are primary apocrine adnexal carcinomas. In about 15% of patients an underlying carcinoma (colon, prostate, bladder) has metastasized to epidermis.



Dermatopathology

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2008, XIV, 299 p., Hardcover

ISBN: 978-3-7985-1839-1