

## **BOWEN'S DISEASE AND PAGET'S DISEASE: PREMALIGNANT LESIONS OF THE ANAL MARGIN**

Bowen's – intraepithelial SQUAMOUS CELL CARCINOMA

Paget's – intraepithelial ADENOCARCINOMA

These entities are different in regards to: Histopathology, age/sex distribution, risk of invasive cancer, and risk of associated nonepithelial malignancies

### PERIANAL BOWEN'S DISEASE

-1912 John T. Bowen, 2 patients with atypical epithelial proliferation

-intraepithelial squamous (carcinoma in situ)

-most commonly in 48-59 year olds and more in females

-Relation to invasive disease:

-6-10% go on to develop invasive squamous cell carcinoma

-30% of these cases will develop metastases

-Relation to associated malignancy:

-Earlier idea (Graham & Helwig): 75% have, or will develop, internal (lung, GI, thymoma) or cutaneous malignancies and 40% develop this lesions within 7 years

-Reyman – case of 600 patients, did not corroborate Bowen's as a marker for internal malignancy – low as 4.7% - this is now the adopted opinion in most texts

### Clinical Features:

-Erythematous, scaly, irregular, crusted raised plaque with well defined margins

-Foci of ulceration indicate invasion

-Symptoms include itching, burning more than pain and spotty bleeding

-Can also be diagnosed incidentally (on hemorrhoidectomy specimens) - ~40%

-Confirmed on biopsy; Bowenoid Cells (multinucleated giant cells with vacuolization, hyperchromic nuclei, negative PAS stain)

-Disordered epidermal hyperplasia/thickness with hyperkeratosis

### Treatment:

-Should do colonoscopy although more important in Paget's

-Wide local excision

-Frozen section can aid in achieving free margins, lesion mapping as in Paget's

-Closure of defect: primary, rotation flaps, STSG

-Some evidence for good response to topical 5-FU and dinitrochlorobenzene

### Results

-Minimal local recurrence with wide local excision

-Cleveland Clinic: 12 patients treated by excision with negative margins – no recurrence

-But long-term follow up is imperative (1 small study showed 20% recurrence in 15 years)

## PERIANAL PAGET'S

- 1874 Sir James Paget described disease entity in the nipple of females
- 1893 Darier and Couillard described perianal Paget's
- Extramammary Paget's (wherever apocrine glands are): axilla, labia majora, penis, scrotum, groin, perineum, buttock, perianal
- Mean onset 59-65
- Rare, only 200 cases in literature
- In general, Paget's has a higher incidence of underlying malignancy and higher risk of progression to invasive cancer
- Relation to invasive disease:
  - Progress into invasive carcinoma in 40% in untreated lesions
- Relation to associated malignancy:
  - Associated with an underlying malignancy in approximately 70%
  - Grotsky: underlying carcinoma in an apocrine gland in 36-50% of cases
  - Helwig: most common internal carcinomas: 22% rectal, 11% anal, breast

## Clinical Features:

- Symptoms: ulceration, discharge, intractable pruritis for many months
- Well demarcated erythematous to gray, elevated, scaly plaque like lesion
- Commonly a delay in diagnosis
- Confirmed on biopsy: Paget Cells (large, pale vacuolated cells with hyperchromatic eccentric nuclei in the epidermis)
- Cells highlight with PAS (periodic acid-Schiff) stain; high mucin content of cells (opposite to Bowen's)

## Treatment:

- Evaluate for underlying/coexistent malignancies (scope)
- Treatment depends if there is an underlying invasive carcinoma
- In absence of invasive carcinoma -> wide local excision
- Microscopic disease can extend in epidermis beyond apparent disease so:
  - "Lesion Mapping" - biopsy anal verge, dentate line, quadrant biopsies of perineum, frozen section
  - If underlying invasive carcinoma is present -> wide excision, possible APR (if rectal adeno)
- Nonoperative methods: topical bleomycin and 5-FU, oral retinoids

## Prognosis

- Worse if underlying carcinoma of apocrine glands or underlying ca of rectum
- 25% with invasive disease have metastases at time of diagnosis (lymph nodes, liver, bone, lung, brain)
- Local recurrence is common, can occur 10 years after excision
- Annual sigmoidoscopy, biopsy of any new lesion, random biopsies at edges of STSG
- Colonoscopy every 3 years

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