2017 IPPF Patient Conference
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PEMPHIGUS & PEMPHIGOID
Below the Belt
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Below the Beltline

- Penile Disease
- Vaginal Disease
- Anal Disease
- Clinical Recognition
- Systemic Treatment
- Local Management
Studies on penile, anal, and pemphigus of the female genital tract have been published.

Broad principles of management and care are applicable to pemphigoid.
PENILE PEMPHIGUS
Penile pemphigus

- Only reported in pemphigus vulgaris not in pemphigus foliaceous
- Few Reports in literature
- A study of 12 patients
  \[Arch Dermtol\ 2001; 137: 756-758\]
- Symptoms are pain on erection, burning, oozing, adhering clothes
- Quality of life
- Cultures for virus & bacteria-repeatedly
Penile pemphigus

Treatment

- Systemic therapy: Will clear but slow
- Topical therapy: Expedite healing
  - Enhance comfort
- Washcloth or gauze soaked in Burrow’s solution and Hibiclens. Only one layer. Avoid multiple layers.
- Must touch skin 7-10 minutes
- Gently pat dry- Apply topical corticosteroid cream
- Initial 3/day. Healing occur 1/day
- Avoid showers
PENILE PEMPHIGOID
Penile Pemphigoid

- Early recognition important
- Avoid biopsy
- Early therapy prevents progression to scarring
- Topical therapy as important as systemic therapy (same principals as PV)
- Scarring irreversible
- Consequences of scarring significant
PEMPHIGUS OF THE FEMALE GENITAL TRACT
Vaginal pemphigus

- Symptoms: Pain on coitus, bleeding, foul smelling, pain and burning on micturition, adherence to clothes.

- Quality of life

- Treatment: - Systemic
  - Topical care
  - Sublesional injection
Vaginal pemphigus

TOPICAL CARE

- Based on extent
  - Widespread disease: Bathtub
  - Vaginal disease: Sitz bath
  - Clean Patt dry- Steroid cream
  - 3/day initially- 1/day improvement

- Micturition: - burning and pain
  - pour water- can long span

- Oozing lesions- Telfa dressing
Vaginal pemphigus

Sublesional corticosteroid injections

- Repeated cultures (bacteria & virus) Negative
- Indications
  1. No response to aggressive topical therapy 6-8 weeks
  2. Multiple sites
  3. Affecting ADL or QOL
  4. Pain on micturition
  5. Lack of healing- in spite systemic therapy
Vaginal pemphigus
Sublesional corticosteroid injections

- Prior to injection
  - Clean with Hebiclens
  - Local anesthesia - Soak gauze with Xylocaine
  - Avoid spray anesthesia
- Dissolve triamcinolone acetonide in 2% Xylocaine with epinephrine
- Inject sublesionally – Keep beveled edge of needle towards epidermis

  - Immediate – Blanching noticed - Injection successful
    - Not intralesional
  - Avoid intradermal subcutaneous injections
PEMPHIGOID OF THE FEMALE GENITAL TRACT
Pemphigoid of the Female Genital Tract

MAIN CONCERNS

- Early recognition essential
- Diagnosis by biopsy
- Institute local and systemic therapy as soon as diagnosis (same as PV)
- Scarring irreversible
- Consequences of scarring catastrophic
Anal Disease
Anal Pemphigus

- Systemic Therapy - Response is systemic
  - Antibody decrease
  - Anti-inflammatory

- Local Management
  Anal Hygiene
  Diet and Stools
  Topical Care
  Intralesional Injections
Anal Pemphigus

MANAGEMENT OF DIET & STOOLS

- Hard Stools
  - Create new lesions
  - Prevent healing
  - Expand & exacerbate existing lesions

- Focus on
  - Diet
  - Laxatives
  - Avoidance of constipation causing drugs
Anal Pemphigus

DIET

-Balance between oral & anal lesions

-American diet poor in fiber

-Diet 2 features
  (i) Consistency
  (ii) Fiber content

- Consume 10gm of fiber per 1000/kcal daily
  - total 25-30gm fiber – use tables
Anal Pemphigus

LAXATIVES

- Bulk – Psyllium, methycellulose (preferred)
- Stool Softeners - Docusate sodium
  - Saline laxative MOM
  - Stimulant – Biscodyl, senna, castor oil, lactulose
- Recommendation based on patient health, other medications
- Change from one to other
Anal Pemphigus

TOPICAL CARE

- Wide spread disease - soak tub bath
  (Domeburro Powder, Herbiclens)

- Limited / Localized disease - Sitz Bath Soak gently

- Apply topical corticosteroid cream
  - 3 / day when severe
  - Reduce 1 / day when improved

- Oozing wet lesions use Telfa to prevent adhering
Anal Pemphigus

ANAL HYGIENE

- Avoid regular toilet paper – trauma and contamination
- Use moist paper without aggressive wiping
- Bidet with central fountain – adjust pressure – avoid trauma
- Avoid fecal contamination of open lesions.
- After cleaning gently compress soft gauze to dry.
Anal Pemphigus

Sublesional corticosteroid injections

- Repeated cultures (Bacteria & Virus) Negative
- Indications
  - No response to aggressive topical therapy 6-8 weeks
  - Multiple sites
  - Affecting ADL or QOL
  - Pain on defecation
  - Lack of healing - in spite of systemic therapy
Anal Pemphigus

Sublesional corticosteroid injections

- Prior to injection
  - Clean with Hebiclens
  - Local anesthesia - Soak gauze with Xylocaine
  - Avoid spray anesthesia

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- Scarring irreversible
- Consequences of scarring significant
  - e.g. loss of sphincter control - diapers
Anal pemphigoid (erosions in anal mucosa)
Anal pemphigoid (scarring of skin & mucosa)
Penile, Anal & Vaginal Pemphigus

SUMMARY

- Generally occur with severe mucosal or generalized disease
- Patient participation, enthusiasm and level of interest, determine outcome
- Local management results in:
  - Reduction of systemic therapy dose
  - More comfort
  - Rapid recovery

- Important principles:
  - Avoid trauma
  - Repeatedly check for infections
- Avoid offending agents or facts that exacerbate lesions.
- Healing complete. No scarring. No sequelae.
More information can be found at our website for the Center for Blistering Diseases.

http://centerforblisteringdiseases.com
You are not a drop in the ocean.

You are the entire ocean, in a drop.

- Rumi -
THANK YOU