Complex Medical Disease

Cases

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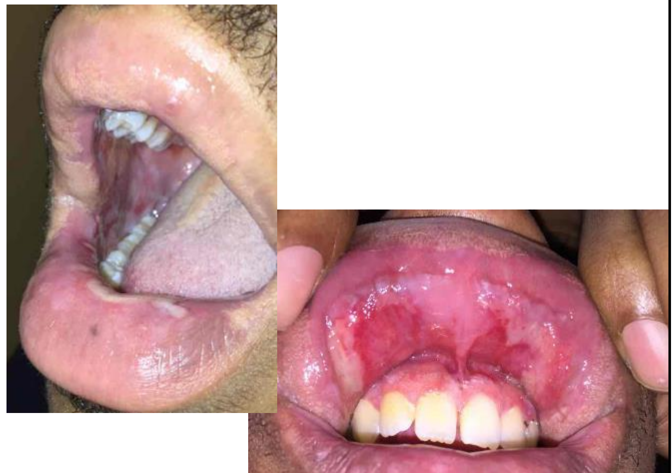
Case 1

- 28 year old male
- 8 year history of recurrent oral and cutaneous erosions, vesicles and bullae
- Initially presented with exclusively oral lesions, subsequent flares involved genital and ocular mucosa, and eventually skin
- No significant PMH, no new medications



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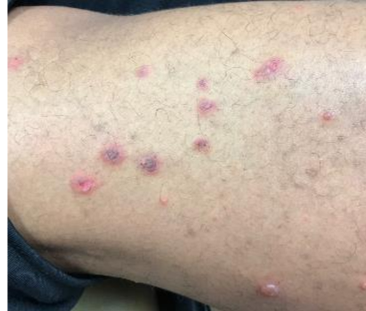
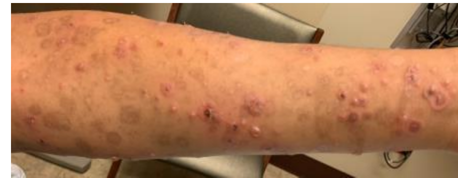
Case 1

- Most flares preceded by URI symptoms
- Testing at time of flares
  - Positive Mycoplasma IgM titers
  - Rhinovirus/enterovirus, adenovirus, seasonal coronavirus, and SARS-CoV-2 coronavirus
    - PCR testing on nasal swabs during active flares
  - HSV antibody titers, HSV PCR on active mucosal vesicles negative
  - HIV negative





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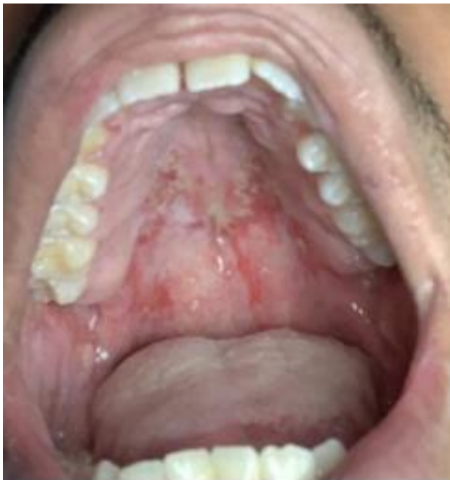
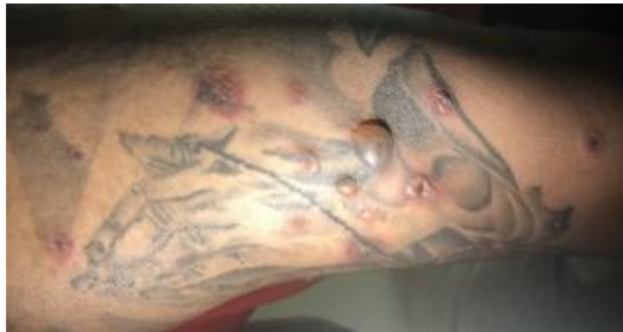
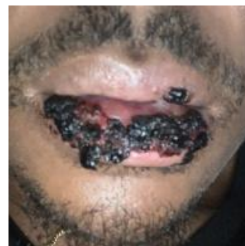
Case 1







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Case 1

- Multiple biopsies have demonstrated vacuolar interface dermatitis with epidermal necrosis
- Negative DIF, negative desmoglein and BPAg antibody titers

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Recurrent Reactive Infectious Mucocutaneous Eruption (RIME)

- Mucosal and cutaneous eruption that occurs following an infection (can be bacterial or viral)
  - Includes mycoplasma induced rash and mucositis (MIRM)
- Occurs mostly in children and adolescents
- Infection triggers and immune response against cutaneous and mucosal epithelium
- Most patients have 2-3 mucosal sites affected – erythema, erosions
- Skin lesions can be vesiculobullous or targetoid
- Histology demonstrates vacuolar change, interface dermatitis, epidermal necrosis
- Recurrent disease present in 9-38% of cases

Canavan TN et al. J Am Acad Dermatol. 2015 Feb;72(2):239-45

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Recurrent Reactive Infectious Mucocutaneous Eruption (RIME)

- Therapy include supportive care
- Antibiotics if appropriate
- Systemic corticosteroids (weak evidence)

- Other immunosuppressive/anti-inflammatory therapies
  - Cyclosporine, IVIg, anti-TNFα inhibitors

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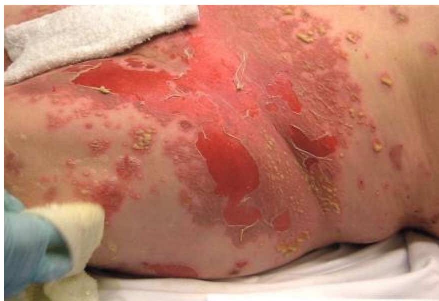
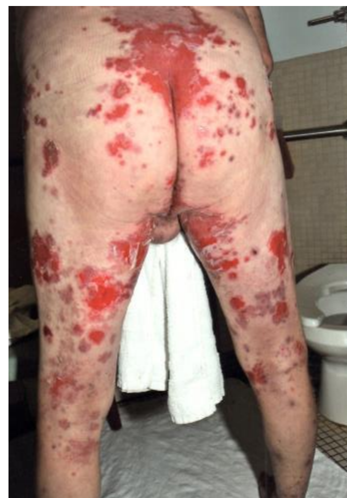
Case 2

- 65 year old male
- Acute onset of vesicles, pustule, erosions
- PMH COPD, emphysema, no new medications





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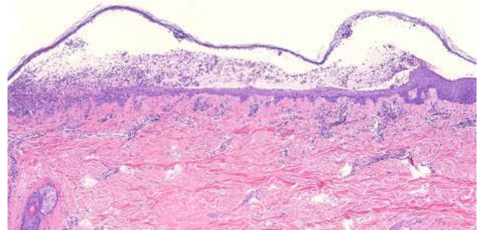
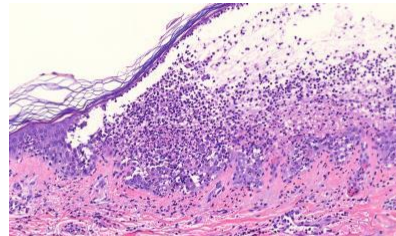
Case 2

- Histology – subepidermal pustule with acantholysis
- DIF – intracellular IgA, C3
- ELISA
  - Anti-Dsg1 – 38 (IgG)
  - Anti-Dsg3 - negative





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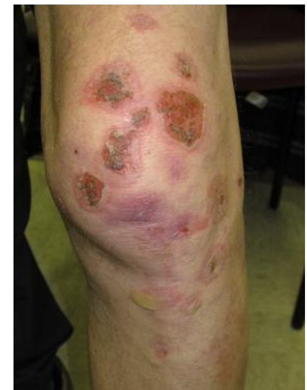
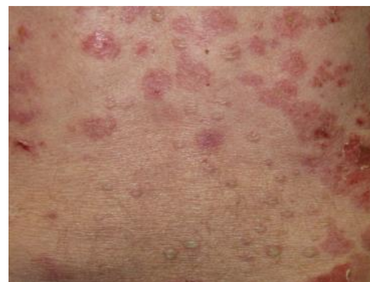
Case 2

- Good response to prednisone and dapsone, prednisone tapered off
- Well controlled on dapsone for about a year, then worsening of disease
- Repeat antibody evaluation
  - IIF IgG and IgA staining keratinocyte cell surface
  - ELISAs*
    - IgG against Dsg1, Dsc3
    - IgA against Dsg1
- Patient was diagnosed with metastatic lung CA





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* Performed by Takashi Hashimoto, MD, Kurume University Institute of Cutaneous Cell Biology

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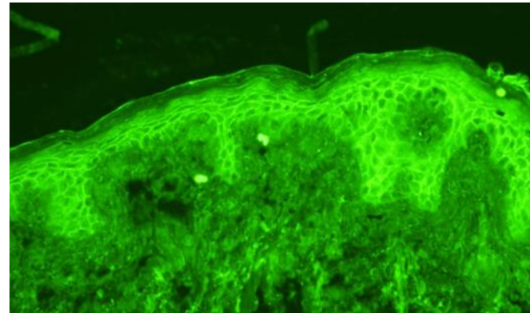

IgA IIF

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IgG/IgA pemphigus

- Rare subtype of pemphigus
- Features of IgA pemphigus, PV, and PF
- Cutaneous involvement common, oral involvement in 40%'
- Pustules present in 39%
- Neutrophilic and eosinophilic epidermal and dermal infiltrates
- Dsg1 and/or Dsg3 more commons targets (IgA and IgG most often directed against the same antigen), anti-Dsc antibodies rare
- 27% of patients with associated malignancy

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Criscito MC et al. J Am Acad Dermatol. 2021 Jul;85(1):237-240

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The End

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