Don’t be Vulgar: A Subtle Case of Pemphigus Vulgaris

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Background
- “Pemphigus” a group of autoimmune mucocutaneous blistering disorders, significant morbidity & mortality
- Acantholysis in epithelium of mucous membranes or skin
- Pemphigus vulgaris (PV) most common
- Pemphigus foliaceus
- First signs of PV observed on oral mucosa in 80% of cases¹

Case Description
HPI:
- 29-year-old female
- 6 mo. h/o intermittent oral ulcerations & bleeding
- Lesions symptomatic over time, healing & reoccurring in ~ 2-week cycles.
- 8 lb weight loss over last month due to 1 pain with swallowing & eating, described as "her mouth on fire."

Physical:
BP: 101/63, HR: 93, RR: 16, T: 36.9 °C, O2SAT: 100%
General: Alert, no acute distress
CV: RRR Lungs: CTAB

Exam Findings

Figure 1. Patchy moderate erythema of the bilateral buccal mucosa & soft palate

Figure 2. Moderate sloughing of the right buccal mucosa (RBM)

Figure 3. Moderate white changes of the posterior left buccal mucosa

Figure 4. Well-defined 0.4 cm ulcer of the sublingual caruncle

Figure 5. 95% improvement of symptoms since initial visit

Differential & Work-up
Lymphoma, leukemia, HIV, syphilis, HSV, autoimmune disorder (Behcet’s)

BMP, ESR: WNL
CBC: WBC ↑ 10.30, Eos: ↑ 5.2%
CRP: 0.4 ANA: Negative
dsDNA Ab: 34 HIV: Nonreactive
RPR: Negative HSV: Negative

Punch Biopsy, RBM: Suprabasal acantholysis, consistent with PV

Immunofluorescence: Intercellular deposition of IgG & CD3 consistent with PV

Management
Consult: Oral Medicine & Dentistry
Dispo: ED Observation Unit pending results & PO challenge

Symptom Management⁶
- Topical lidocaine solution or gel
- Triamcinolone paste or fluocinonide gel
- Dexamethasone mouthwash

Outcome
- 3 weeks treatment w/ oral prednisone, mycophenolate mofetil, & dexamethasone mouth wash, patient is able to eat
- Adverse effects: rapid heartbeat & minor ankle swelling, resolved

Discussion
- PV can be life-threatening; prior to tx, 70-100% of patients died in ~ 1-5 years²³
- Incidence: 0.1 – 0.5/100,000 people/year
- 40 – 60 years old, female⁴⁵
- Oral systemic glucocorticoids are very effective treatment
- Nonsteroidal immunomodulatory agents in conjunction

Conclusion
- Data on treatment is limited; uncertain therapy efficacy⁶
- Widespread loss of epidermal barrier requires assessment for⁶:
  - Protein & fluid loss
  - Electrolyte imbalances
  - Dietary insufficiencies
  - Increased catabolism
  - Local and systemic infections

Take Home Points
- Treatment always indicated at onset, even for mild disease
- Prognosis dramatically improved with astute diagnosis & treatment; of importance for EM physicians to keep PV on differential

References