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

Charlene M. Gaw, MPH[1], Valerie A. Dobiesz, MD, MPH[2]
[1]Mayo Clinic Alix School of Medicine, Rochester, MN, [2]Department of Emergency Medicine, Brigham and Women's Hospital, Boston, MA

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 cases[1]

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 ↑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

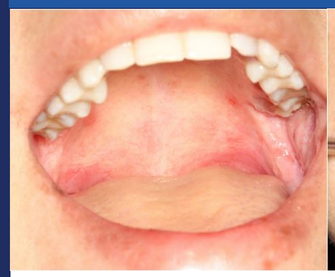

Figure 1. Patchy moderate erythema of the bilateral buccal mucosa & soft palate

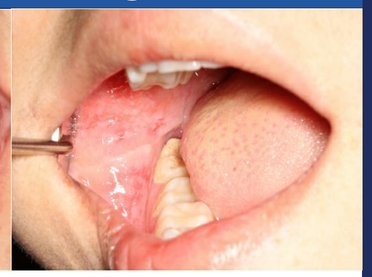

Figure 2. Moderate sloughing of the right buccal mucosa (RBM)

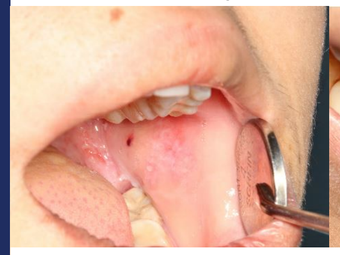

Figure 3. Moderate white changes of the posterior left buccal mucosa

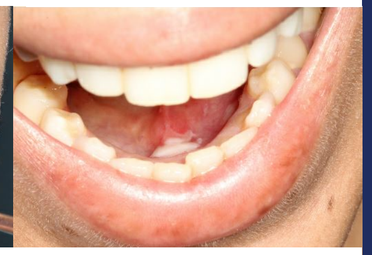

Figure 4. Well-defined 0.4 cm ulcer of the sublingual caruncle

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 Management[6]

- 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

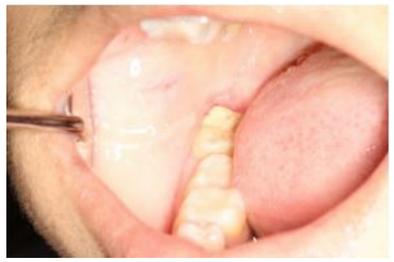

Figure 5. 95% improvement of symptoms since initial visit

Discussion

- PV can be life-threatening; prior to tx, 70-100% of patients died in ~ 1-5 years[2-3]
  - Incidence: 0.1 – 0.5/100,000 people/year
  - 40 – 60 years old, female[4-5]
- Oral systemic glucocorticoids are very effective treatment
  - Nonsteroidal immunomodulatory agents in conjunction

Conclusion

- Data on treatment is limited; uncertain therapy efficacy[6]
- Widespread loss of epidermal barrier requires assessment for[6]:
  - 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

1. Santoro, FA, Stoopler, ET, and Werth, V.P. Pemphigus. *Dent Clin North Am*. 2013; 57: 597–610
2. Bystryn J. and Rudolph, J.L. Pemphigus. *The Lancet.* 2005; 366(9479): 61-73, ISSN 0140-6736,
3. Sergei A. Grando (2012) Pemphigus autoimmunity: Hypotheses and realities, Autoimmunity, 45:1, 7-35, DOI: 10.3109/08916934.2011.606444
4. McMillan, R., Taylor, J., Shephard, M. et al. World Workshop on Oral Medicine VI: a systematic review of the treatment of mucocutaneous pemphigus vulgaris. *Oral Surg Oral Med Oral Pathol Oral Radiol*. 2015; 120: 132–142.e61
5. Stoopler, E.T. and Sollecito, T.P. Oral mucosal diseases: evaluation and management. *Med Clin North Am*. 2014; 98: 1323–1352
6. Martin, L.K., Werth, V.P., Villaneuva, E.V., et al. A systematic review of randomized controlled trials for pemphigus vulgaris and pemphigus foliaceus. *Journal of the Am Academy of Derm*, 2011; 64(5):903-8. ISSN 0190-9622,