Pemphigus vulgaris: a case report of a blistering masquerader

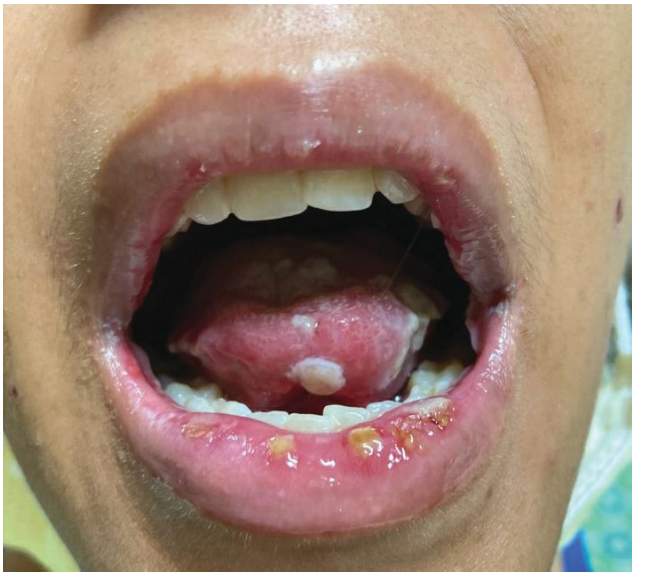
BY JESSICA PADNIEWSKI, DO, AND ERIN LUXENBERG, MD

Pemphigus vulgaris (PV) is a rare autoimmune blistering disorder mediated by anti-desmoglein-1 and anti-desmoglein-3 auto-antibodies, which results in acantholysis. PV typically presents in elderly patients with initial lesions in the oral cavity. Estimated incidence of PV ranges from 0.5-3.2/100,000. As with all bullous disorders, PV compromises the skin barrier and increases risk of infection and insensible fluid losses. The differential for blistering disorders is broad, including contact dermatitis, autoimmune disorders such as Bechet's or systemic lupus erythematosus, neoplastic disorders, infectious diseases such as HSV and mycoplasma, in addition to drug-related reactions to medications like NSAIDs or antibiotics. We present a case of PV which was initially thought to be disseminated rash due to HSV.

Case description

A 30-year-old female with history of papillary thyroid cancer s/p thyroidectomy presented for evaluation of painful oral lesions. She denied new medications or exposures but reported using NSAIDs intermittently for pain. She was initially diagnosed with mucositis related to NSAID use, instructed to avoid NSAIDs, and prescribed a prednisone taper. Symptoms overall improved until she took an aspirin and noted worsening oral lesions with new onset bilateral ear pain without any auditory deficit. She also reported losing about 15 pounds since onset of lesions. She denied any other symptoms.

On exam, multiple punched-out erosions with yellow granulation tissue scattered on upper and lower lips with desquamative gingivitis on the buccal mucosa were seen (Image 1). Preliminary lab work was remarkable for HSV-1 positivity and HCV-Ab positivity with negative PCR. Skin swabs and biopsy of ulcerated lesions were done. Oral PCR swab was positive for HSV and presumptive diagnosis of disseminated HSV was made. She was initiated on acyclovir 400 mg TID for 7 days with subsequent suppressive therapy with valaciclovir 500 mg daily. Despite this, her lesions persisted and progressed. Skin biopsy ultimately revealed intraepithelial vesiculations and acantholysis and negative direct immunofluorescence. Indirect immunofluorescence was positive with IgG to epithelium at 1:5,120 (H) with monkey esophagus and & 1:640 (H) with intact human skin which was concerning for a diagnosis of PV with mucocutaneous involvement.



Antibody testing returned strongly positive for both IgG-desmoglein1 antibodies and IgG-desmoglein3 antibodies, confirming the diagnosis of PV. The patient's skin lesions were refractory to a variety of topical treatments including clobetasol gel and ointment, fluticasone ointment, mupirocin, and systemic mycophenolate 1000mg BID and she was ultimately initiated on Rituximab infusions weekly x 4. She experienced a flare after receiving her COVID vaccination but has otherwise been stable and further improvement is expected.

Discussion

This patient's blistering rash was initially thought to be an eruption related to disseminated HSV infection. Several medications, environmental factors and infections have been identified as triggers to PV, including HSV infection (Brenner). This case exemplifies the importance of maintaining broad differentials and specifically maintaining a clinical suspicion for rare conditions. This is especially important when treatment dramatically differs such as in the case of disseminated HSV and PV. Current first-line treatment for PV includes steroid sparing immunosuppressive drugs such as Rituximab (Anandan). Additionally, this case exemplifies the importance of prudence with COVID-19 vaccinations in patients with underlying autoimmune disorders that may flare with vaccination. **MM**

Jessica Padniewski, DO, is a third-year resident in internal medicine, Internal Medicine Residency Program, Hennepin Healthcare. Erin Luxenberg, MD, is a staff dermatologist, Hennepin Healthcare, and assistant professor, University of Minnesota.

REFERENCES

Brenner S, Goldberg I. Drug-induced pemphigus. Clinical Dermatology. 2011 Jul-Aug;29(4):455-7. doi: 10.1016/j.clindermatol.2011.01.016. PMID: 21679874.

Anandan V, Jameela WA, Sowmiya R, Kumar MMS, Lavanya P. Rituximab: A Magic Bullet for Pemphigus. J Clin Diagn Res. 2017 Apr;11(4):WC01-WC06. doi: 10.7860/JCDR/2017/21868.9717. Epub 2017 Apr 1. PMID: 28571245; PMCID: PMC5449891.

American College of Rheumatology. ACR COVID-19 vaccine guidance recommends vaccination, addresses immunosuppressant drugs & patient concerns. Press release. Published February 11, 2021.