New validated diagnostic criteria for pyoderma gangrenosum

To the Editor: We read with interest the review on neutrophilic dermatoses by Ashchyan et al[1] and believe that it will be of significant value to the dermatologic community. To supplement their review, there are 2 additional viewpoints that we would like to highlight, specifically, regarding the diagnosis and treatment of pyoderma gangrenosum (PG).

Ashchyan et al[1] state that PG remains a "diagnosis of exclusion,"[2] which is a characterization that is difficult to justify, as it is impractical to have a medical diagnosis requiring that all other possible diagnoses be ruled out. In fact, the lack of clear diagnostic criteria for PG may be 1 reason why it has been reported that many cases initially diagnosed as PG can ultimately be reclassified as an alternative diagnosis.[3]

Pertinent to this topic, 2 PG diagnostic criteria have been recently published.[4,5] The new criteria were independently developed in parallel by separate groups following different approaches.[5] The first of the 2 studies utilized a score-based approach in which criteria weight was determined by observed prevalence among patients with PG.[4] The authors of the second study based their criteria on a Delphi exercise, which was then mathematically refined and validated.[5] It is hoped that these diagnostic models will be of benefit in the clinical and research settings. Both models attempt to deemphasize the need to exhaustively exclude other causes of ulceration and instead focus more on the pathologic features of PG. Of course, when PG is suspected, relevant causes of ulceration should still be excluded.

Second, Ashchyan et al also highlighted as a "key point" the fact that the criterion standard therapy for PG is systemic corticosteroids. Although corticosteroids and cyclosporine have been the best characterized agents in the literature, we would caution against designating any PG therapy as a criterion standard.[6] To date, there have been only 2 randomized controlled clinical trials in PG.[7,8] Although Ashchyan et al do describe the STOP-GAP trial in their discussion of treatments, the finding that the prednisolone and cyclosporine treatment arms had similar overall healing rates, namely, 47% at 6 months, was not addressed. In addition, the STOP-GAP study demonstrated that serious adverse reactions, such as infections, were more common in the prednisolone group.[7] On the basis of the available data, selection of a systemic immunosuppressant should be tailored to each individual patient according to medication adverse event profiles, PG severity, and medical comorbidities, especially in light of the fact that approximately 55% of PG occurs in association with underlying systemic disease.[9]

Emanual Maverakis, MD,[a] Stephanie T. Le, MD,[a] Jeffrey Callen, MD,[b] Uwe Wollina, MD,[c] Angelo Valerio Marzano, MD,[d] Daniel Wallach, MD,[e] Courtney Schadt, MD,[b] Yocasta C. Martinez-Alvarado, MD,[f] Michelle Y. Cheng, MD,[a] Chelsea Ma, MD,[a] Alexander Merleev, PhD,[a] Anthony Ormerod, MD,[g] Fiona Craig, MRCP,[g] Finja Jockenhofer, MD,[b] Joachim Dissemond, MD,[b] Katrin Salva, MD,[b] Hywel C. Williams, DSc,[i] and David Fiorentino, MD, PhD[j,k]

From the Department of Dermatology, University of California, Davis, Sacramento, California[a]; Division of Dermatology, Department of Medicine, University of Louisville, Louisville, Kentucky[b]; Department of Dermatology and Allergology, Academic Teaching Hospital Dresden, Dresden, Germany[c]; UOC di Dermatologia, IRCCS Fondazione Ca' Granda Ospedale Maggiore Policlinico, Milano—Dipartimento di Fisiopatologia Medico—Chirurgica e dei Trapianti, Università degli Studi di Milano, Milano, Italy[d]; Department of Dermatology, Paris Hospitals, Paris, France[e]; Department of Dermatology, Hospital Civil de Guadalajara, Fray Antonio Alcalde, University of Guadalajara, Jalisco, Mexico[f]; Department of Dermatology, School of Medicine and Dentistry, University of Aberdeen, Aberdeen, United Kingdom[g]; Department of Dermatology, Venereology and Allergology, University Hospital of Essen, Germany[b]; Centre of Evidence Based Dermatology, King's Meadow Campus, University of Nottingham, United Kingdom[i]; and Department of Dermatology[j] and Department of Internal Medicine, Division of Immunology and Rheumatology, Stanford University, Stanford, California[k]

Funding sources: None.

Conflicts of interest: None disclosed.

Reprints not available from the authors.

Correspondence to: Emanual Maverakis, MD, Department of Dermatology, University of California, Davis, 3301 C St, Ste 1400, Sacramento, CA 95816

E-mail: emaverakis@ucdavis.edu

REFERENCES

1. Ashchyan HJ, Nelson CA, Stephen S, James WD, Micheletti RG, Rosenbach M. Neutrophilic dermatoses. Pyoderma gangrenosum and other bowel and arthritis associated neutrophilic dermatoses. *J Am Acad Dermatol*. 2018;79:1009-1022.
2. Su WPD, Davis MDP, Weenig RH, Powell FC, Perry HO. Pyoderma gangrenosum: clinicopathologic correlation and proposed diagnostic criteria. *Int J Dermatol*. 2004;43(11):790-800.
3. Weenig RH, Davis MDP, Dahl PR, Su WPD. Skin ulcers misdiagnosed as pyoderma gangrenosum. *N Engl J Med*. 2002;347(18):1412-1418.
4. Jockenhöfer F, Wollina U, Salva KA, Benson S, Dissemond J. The PARACELSUS score: a novel diagnostic tool for pyoderma gangrenosum. *Br J Dermatol*. 2018. https://doi.org/10.1111/bjd.16401. Accessed August 10, 2018.
5. Maverakis E, Ma C, Shinkai K, et al. Diagnostic criteria of ulcerative pyoderma gangrenosum. *JAMA Dermatol*. 2018;154(4):461.
6. Partridge ACR, Bai JW, Rosen CF, Walsh SR, Gulliver WP, Fleming P. Effectiveness of systemic treatments for pyoderma gangrenosum: a systematic review of observational studies and clinical trials. *Br J Dermatol*. 2018;179:290-295.
7. Ormerod AD, Thomas KS, Craig FE, et al. Comparison of the two most commonly used treatments for pyoderma gangrenosum: results of the STOP GAP randomised controlled trial. *BMJ*. 2015;350:h2958.
8. Brooklyn TN, Dunnill MGS, Shetty A, et al. Infliximab for the treatment of pyoderma gangrenosum: a randomised, double blind, placebo controlled trial. *Gut*. 2006;55(4):505-509.
9. Binus AM, Qureshi AA, Li VW, Winterfield LS. Pyoderma gangrenosum: a retrospective review of patient characteristics, comorbidities and therapy in 103 patients. *Br J Dermatol*. 2011;165(6):1244-1250.

https://doi.org/10.1016/j.jaad.2018.08.068