New validated diagnostic criteria for pyoderma gangrenosum

Emanuel Maverakis, MD, Stephanie T. Le, MD, Jeffrey Callen, MD, Uwe Wollina, MD, Angelo Valerio Marzano, MD, Daniel Wallach, MD, Courtney Schadt, MD, Yocasta C. Martinez-Alvardao, MD, Michelle Y. Cheng, MD, Chelsea Ma, MD, Alexander Merleev, PhD, Anthony Ormerod, MD, Fiona Craig, MRCP, Finja Jockenhofer, MD, Joachim Dissemond, MD, Katrin Salva, MD, Hywel C. Williams, DSc, David Fiorentino, MD, PhD

PII: S0190-9622(18)33084-6
DOI: https://doi.org/10.1016/j.jaad.2018.08.068
Reference: YMJD 13017

To appear in: Journal of the American Academy of Dermatology

Received Date: 22 August 2018
Accepted Date: 24 August 2018


This is a PDF file of an unedited manuscript that has been accepted for publication. As a service to our customers we are providing this early version of the manuscript. The manuscript will undergo copyediting, typesetting, and review of the resulting proof before it is published in its final form. Please note that during the production process errors may be discovered which could affect the content, and all legal disclaimers that apply to the journal pertain.
Article Type: Letter: Notes and Comments

Title: New validated diagnostic criteria for pyoderma gangrenosum

Authors: Emanuel Maverakis, MD1; Stephanie T. Le, MD1; Jeffrey Callen, MD2; Uwe Wollina, MD3; Angelo Valerio Marzano, MD4; Daniel Wallach, MD5; Courtney Schadt, MD2; Yocasta C. Martinez-Alvarado, MD6; Michelle Y. Cheng, MD1; Chelsea Ma, MD1; Alexander Merleev, PhD1; Anthony Ormerod, MD7; Fiona Craig, MRCP7; Finja Jockenhofer, MD8; Joachim Dissemond, MD8; Katrin Salva, MD8; Hywel C. Williams, DSc6; David Fiorentino, MD, PhD10,11

Affiliations:
1Department of Dermatology, University of California, Davis, Sacramento, California
2Division of Dermatology, Department of Medicine, University of Louisville, Louisville, Kentucky
3Department of Dermatology and Allergology, Academic Teaching Hospital Dresden, Dresden, Germany
4UOC 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
5Department of Dermatology, Paris Hospitals, Paris, France
6Department of Dermatology, Hospital Civil de Guadalajara, Fray Antonio Alcalde, University of Guadalajara, Jalisco, Mexico
7Department of Dermatology, School of Medicine and Dentistry, University of Aberdeen, Aberdeen, United Kingdom
8Department of Dermatology, Venereology and Allergology, University Hospital of Essen, Germany
9Centre of Evidence Based Dermatology, King’s Meadow Campus, University of Nottingham, United Kingdom
10Division of Immunology and Rheumatology, Department of Dermatology, Stanford University, Stanford, California
11Division of Immunology and Rheumatology, Department of Internal Medicine, Stanford University, Stanford, California

Corresponding Author: Emanuel Maverakis, MD, Department of Dermatology, University of California, Davis, 3301 C St, Ste 1400, Sacramento, CA 95816 (emaverakis@ucdavis.edu).

Word Count: 399/500
Figure Count: 0
Table Count: 0
Authorship Disclosure: No relevant financial or nonfinancial relationships to disclose.
To the Editor: We read with interest the review on neutrophilic dermatoses by Ashchyan et al. and believe it will be of significant value to the dermatologic community. To supplement their review, there are two additional viewpoints that we would like to highlight, specifically regarding the diagnosis and treatment of pyoderma gangrenosum (PG).

Ashchyan et al. state that PG remains a “diagnosis of exclusion,” a definition that is difficult to justify, as it is impractical to have a medical diagnosis that requires one to rule out all other possible diagnoses. In fact, the lack of clear diagnostic criteria for PG may be one reason why it has been reported that many cases initially diagnosed as PG ultimately can be reclassified as an alternative diagnosis.

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

Secondly, Ashchyan et al. also highlighted as a "key point" that the criterion standard therapy for PG is systemic corticosteroids. Although corticosteroids and cyclosporine have been the most well characterized agents in the literature, we would caution against designating any PG therapy as a “criterion standard”. To date, there have only been two randomized controlled clinical trials in PG. While 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, 47% at six 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. Based on the available data, selection of a systemic immunosuppressant should be tailored to each individual patient based on 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.


