Reply to: "New validated diagnostic criteria for pyoderma gangrenosum"

To the Editor: We would like to thank Maverakis et al for expressing interest in our article and for raising important discussion points.[1] In particular, we applaud them for drawing attention to the new diagnostic criteria for pyoderma gangrenosum (PG) that were published this year.[2,3] These criteria represent an important step forward in the diagnosis of PG.

Maverakis et al contend that PG should no longer be labeled a diagnosis of exclusion, as it is impractical to have a medical diagnosis that requires one to rule out all other possible diagnoses.[2] We respectfully disagree. From a semantic standpoint, we believe that the label "diagnosis of exclusion" is crucial to remind physicians to exclude mimickers, such as ulceration of vascular, infectious, inflammatory, and neoplastic etiologies, before rendering a diagnosis of PG.[3] For example, failing to exclude infection before initiating immunosuppressive therapy could have dire consequences for the patient and medicolegal ramifications for the physician. Indeed, in their article, Weenig et al demonstrated that PG is commonly misdiagnosed and concluded that a thorough evaluation is required in all patients suspected of having PG to rule out alternative diagnoses.[4]

The sole major diagnostic criterion for ulcerative PG yielded by the Delphi exercise was biopsy of ulcer edge demonstrating a neutrophilic infiltrate.[2] Biopsy is also valuable to evaluate for PG mimickers. Although we acknowledge that excluding every alternative diagnosis in every patient may not be practical, at a minimum we recommend that patients with a suspected diagnosis of ulcerative PG undergo a thorough history and physical examination, a skin biopsy with tissue culture, a complete blood count with differential, and age-appropriate malignancy screening, with further evaluation guided by the patient's age, comorbidities, and symptoms according to the clinical judgment of the treating physician.[5]

Maverakis et al appropriately question our "key point" that systemic corticosteroids are the therapeutic criterion standard for PG. This key point glosses over the nuance that we presented in the body of our article, and we appreciate the opportunity for clarification. The original diagnostic criteria for PG proposed by Su et al listed rapid response to systemic steroid treatment as the fourth minor criterion.[6] From a historical perspective, systemic corticosteroids have been used as the therapeutic criterion standard against which novel steroid-sparing therapies have been measured. In our discussion regarding the management of PG, we noted that cyclosporine and tumor necrosis factor-α inhibitors are now considered first-line therapies for PG along with systemic corticosteroids. We emphasized that individual patient characteristics should guide the physician in selecting first-line therapy.[3]

In conclusion, we thank Maverakis et al[2] for their letter and their contributions to the literature on PG. We look forward to reading more of their work and, we hope, collaborating to shed light on this challenging disease.

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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(6):1009-1022.
2. Maverakis E, Ma C, Shinkai K, et al. Diagnostic criteria of ulcerative pyoderma gangrenosum: a delphi consensus of international experts. *JAMA Dermatol*. 2018;154(4):461-466.
3. Jockenhofer F, Wollina U, Salva KA, Benson S, Dissemond J. The PARACELSUS score: a novel diagnostic tool for pyoderma gangrenosum. *Br J Dermatol*. doi: 10.1111/bjd.16401, Accessed December 8, 2018.
4. Weenig RH, Davis MD, Dahl PR, Su WP. Skin ulcers misdiagnosed as pyoderma gangrenosum. *N Engl J Med*. 2002;347(18):1412-1418.
5. Ashchyan HJ, Butler DC, Nelson CA, et al. The association of age with clinical presentation and comorbidities of pyoderma gangrenosum. *JAMA Dermatol*. 2018;154(4):409-413.
6. Su WP, Davis MD, Weenig RH, Powell FC, Perry HO. Pyoderma gangrenosum: clinicopathologic correlation and proposed diagnostic criteria. *Int J Dermatol*. 2004;43(11):790-800.

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