Proving Pyoderma, Diagnosis and Treatment of Suspected Pyoderma Gangrenosum

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Case Presentation

- A 49 y/o F with PMH SVT, GERD, HLD, Hashimoto's thyroiditis, psoriatic arthritis, autoimmune hives, IDA, and LLE wound presents with concern for infection due to an indolently growing and highly painful wound to the LLE onset 2 months
- Her wound began as a small red dot that evolved into a blister and growing ulcer – cultures of this wound showed normal flora
- Despite several courses on antibiotics and oral steroids patient did not have regression of wound – although she does report slowed growth on high dose prednisone
- On physical exam patient has a 10 x 6 cm discoid lesion to left lower extremity just superior to the medial malleolus with erythema and cribiform granulation tissue, necrotic edges, and surrounding induration

Background

- Pyoderma gangrenosum is a rare, chronic, non-infectious, neutrophilic dermatosis in which a painful nodule or pustule breaks down to form a progressively enlarging ulcer with a raised, violaceous, undermined border.
- Estimated incidence is around 3-10/1,000,000 population per year
- 50% of PG is associate with Inflammatory Bowel Disease, and they are frequently associated with autoimmune conditions and solid or hematologic malignancies
- These wounds often exhibit pathergy – an exaggerated skin injury after even minor trauma like bug bites or bumps/bruises

Differential diagnosis for suspected PG

Common causes of cutaneous ulceration include infection (bacterial, granulomatous, fungal), vascular/diabetic ulceration, malignancy, insect bites, calciphylaxis, drug induced ulceration

Workup

Workup should rule out other causes of cutaneous ulceration and screen for potential underlying conditions

- Wound culture with fungal and acid-fast bacilli cultures
- Blood counts and inflammatory markers
- ABIs, evaluate for venous insufficiency
- Vasculitis and Anti-phospholipid antibody screen: ANCA and VDRL
  - PG patients are often pANCA (+) especially if in IBD, but cANCA suggests Wegeners
- Hepatitis screen
- SPEP – paraproteinemia association
- Biopsy: can reveal malignancy, mycobacterium, fungi, etc

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Diagnostic criteria

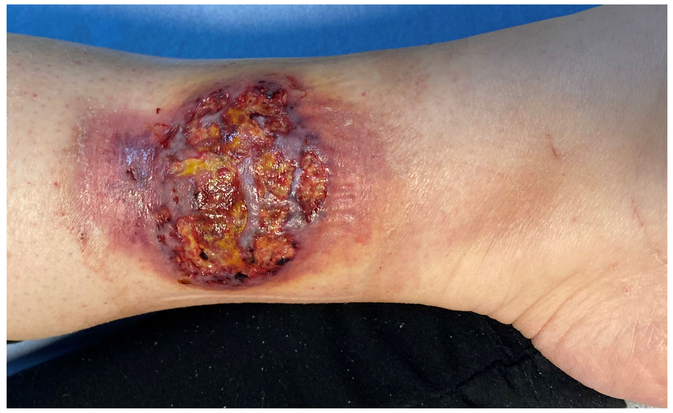
Major Criteria

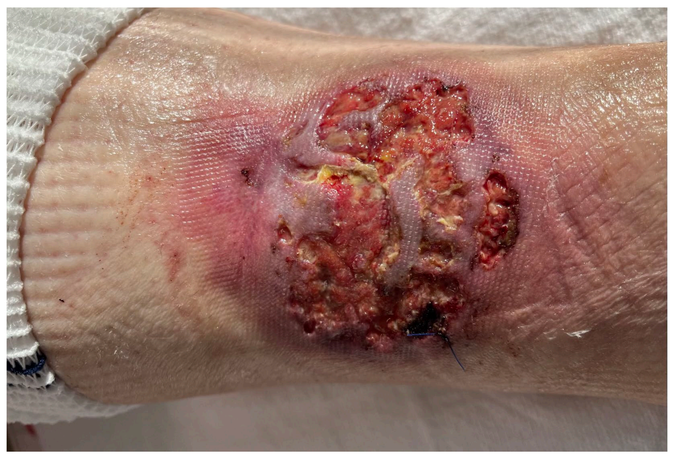
- Rapid progression of painful, necrolytic cutaneous ulcer with irregular, violaceous, and undermined border
- Other causes of cutaneous ulceration excluded

Minor Criteria

- History suggestive of pathergy or cribiform scarring
- Systemic diseases associated with PG – esp. IBD, inflammatory arthritis, hematologic diseases, and malignancy
- Histopathologic findings (sterile dermal neutrophilia, +/- mixed inflammation, +/- lymphocytic vasculitis
- Treatment response (rapid response to systemic steroid treatment)

Before and After





Treatment

- Control any contributing underlying disease
- Wound care!
- For small lesions: Potent topical steroid ointment, Tacrolimus ointment, intralesional steroid injections into the ulcer edge, or oral anti-inflammatory antibiotics such as doxycycline or minocycline
- For larger or more numerous ulcers: Systemic treatment needed. Oral prednisone for several weeks or longer, intermittent intravenous solumedrol for 3–5 days, or Cyclosporine, biologic agents, mycophenolate, methotrexate, and other immunosuppressants

Conclusion

This patient presented to the ED from her dermatologist's office for concern of superimposed infection and increased pain. Previous labs as part of a workup for PG showed negative bacterial, fungal, and acid-fast cultures along with negative hepatitis and vasculitis screens.

Initial out-patient biopsy was not characteristic of PG, however repeat biopsy inpatient showed benign skin with dermal abscesses, no malignancy identified, and no evidence of micro-organism involvement. These findings, in conjunction with the clinical presentation were consistent with PG.

Initial antibiotic therapy yielded little improvement, however patient showed marked improvement after initiation of high dose IV steroids (1g solumedrol daily). Cultures in the hospital again showed normal flora. Fecal calprotectin was elevated suggesting an underlying IBD in addition to the patient's known auto-immune history.

References

- Schmieder SJ, Krishnamurthy K. Pyoderma Gangrenosum. [Updated 2023 Jul 4]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK482223/
- Ruocco, E., Sangiuliano, S., Gravina, A., Miranda, A. and Nicoletti, G. (2009), Pyoderma gangrenosum: an updated review. Journal of the European Academy of Dermatology and Venereology, 23: 1008-1017. https://doi.org/10.1111/j.1468-3083.2009.03199.x
- George C, Deroide F, Rustin M. Pyoderma gangrenosum - a guide to diagnosis and management . Clin Med (Lond). 2019 May;19(3):224-228. doi: 10.7861/clinmedicine.19-3-224. PMID: 31092515; PMCID: PMC6542232.