ISSN 2664-8075 (Print) & ISSN 2706-5774 (Online)
South Asian Research Journal of Applied Medical Sciences
Abbreviated Key Title: South Asian Res J App Med Sci

| Volume-7 | Issue-4 | Jul-Aug- 2025 | **DOI: https://doi.org/10.36346/sarjams.2025.v07i04.004**

Case Report

Evolution of Classic Pyoderma Gangrenosum: Case Report and Review of the Literature

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Article History
Received: 29.05.2025
Accepted: 08.07.2025
Published: 10.07.2025

Abstract: Pyoderma gangrenosum was first described in 1908 and is a rare neutrophilic disease not associated with infection. It is characterized by the appearance of painful skin ulcers with undermined and irregular edges, usually purplish in color and with peripheral erythema. It most commonly affects the lower extremities, although it can occur in other locations, including the genital, perineal, and perianal regions. (1) There is a relationship between this condition and inflammatory bowel disease, as well as other autoimmune diseases and even neoplasms. It is my understanding that the physician is able to understand and explain to family members the estimated time of healing and the importance of the phenomenon of pathergy in the disease. This is possible because knowing the evolution of the lesions allows the physician to understand the phenomenon of pathergy. We present the case of a patient with lesions that appear to be pyoderma gangrenosum on the head, showing how they have evolved until resolution.

Keywords: *Pyoderma Gangrenosum, Evolution, Neutrophilic, Systemic Disease, Inflammatory Bowel Disease*.

INTRODUCTION

The disease was first described in 1908 by Brocq, and it is considered rare. It seems that "pyoderma gangrenosum" might not be the most accurate term in this case, as it is not an infection [1].

In terms of epidemiology, it appears to affect middle-aged people, approximately 40 years old, with a relatively low number of cases reported per million people. It has been observed that this condition is more prevalent in women and less common in children [1].

It has been observed in the literature that a significant proportion of cases appear to be linked to systemic diseases, particularly chronic nonspecific ulcerative colitis, rheumatoid arthritis, hematological disorders, and neoplasms. A study of a group of patients in Korea found that those with pyoderma gangrenosum seemed to have a higher risk of mortality compared to the general population [1-4]. Therefore, it would be advisable to perform an adequate interview and physical examination to determine whether the patient has any findings that suggest the presence of a systemic disease associated with pyoderma gangrenosum.

While the pathogenesis of pyoderma gangrenosum is not fully understood, it is recognized as an autoinflammatory disorder. In this condition, there is dysfunction of the innate and adaptive immune systems, with a predominance of neutrophil activity and an exacerbated cytokine-mediated inflammatory response [1].

It is possible that keratinocytes may be a significant source of proinflammatory cytokines IL1α, IL1β, IL8, IL36, and tumor necrosis factor (TNF), which have been observed to promote neutrophil recruitment, particularly IL8. It is possible that macrophages and monocytes may contribute to the production of IL1α, IL1β, IL8, chemokine 5 with CC motif (CCL5), CXCL9, CXCL10, CXCL11, and TNF. It has been observed that of these cytokines, CCL5 and CXCL9 appear to attract T cells, which are found perivascularly at the periphery of the wound or around the annexial structures. It has been suggested that IL17A may play a role in regulating the release of various chemokines by other cells, which could support the migration of neutrophils and monocytes. Additionally, there is a possibility that IL17A may interact with TNF, though further research is needed to fully understand its role in this context [1-6].

It seems that the best-documented factor that can induce PG ulcerations is trauma, which is defined as a minimal inflammatory skin reaction [1].

It is my understanding that four subtypes are described: ulcerative (classic manifestation), bullous (atypical), pustular, and vegetative [1].

Its morphology begins with a papular or pustular lesion that progresses to one or more ulcers, sometimes coalescent, with a necrotic base, irregular purple edges, and a granulation tissue appearance, crater-like in appearance. It is important to note that ulcers often have a distinctive appearance, characterized by a undermined edge that is indicative of a dense dermal neutrophilic infiltrate. The deep location of these inflammatory cells contributes to the reddish-purple tint of the undermined edge. Additionally, a peripheral area of erythema, which is known in histology as perivascular lymphocytes, is often present. It most commonly affects the lower extremities, although it can occur in other locations, including the genital, perineal, and perianal regions. It is characterized by the rapid appearance of painful skin ulcers [1].

Historically, it was classified as a diagnosis of exclusion, but it is currently classified using the Delphi method by a panel of experts, requiring one major criterion and four minor criteria, with a sensitivity and specificity of 86% and 90%, respectively [7]. Another option to consider is the PARACELSUS diagnostic tool. In cases where a total score of 10 points or more is obtained, there is a high probability of pyoderma gangrenosum, which can help differentiate it from venous leg disease.

Table 3: Adapted from the Diagnostic Criteria of Ulcerative Pyoderma Gangrenosum: A Delphi Consensus of International Experts [7]

Delphi Consensus Criteria (2018)	
Histology	• **Biopsy of ulcer edge demonstrating neutrophilic infiltrate**
History	Exclusion of infection Pathergy History of inflammatory bowel disease or inflammatory arthritis History of papule, pustule, or vesicle ulcerating within 4 days of appearing
Clinical examination (or photographic evidence)	Peripheral erythema, undermining border, and tenderness at ulceration site Multiple ulcerations, at least 1 on an anterior lower leg Cribriform or “wrinkled paper” scar(s) at healed ulcer sites
Treatment	Decreased ulcer size within 1 month of initiating immunosuppressive medication(s).

Table 3: Adapted from the PARACELSUS score: a novel diagnostic tool for pyoderma gangrenosum [8]

Paracelsus Score (2019)	
major diagnostic criteria (3 puntos)	Progressing disease (rapidly) Assessment of relevant differential diagnoses Reddish-violaceous wound border
Minor criteria (2 puntos)	Amelioration by immunosuppressant drugs Characteristically irregular shape of ulceration Extreme pain > 4/10 on a visual analogue scale Localization of lesion at the site of the trauma
Criterios adicionales (1 punto)	Suppurative inflammation in histopathology Undermined wound borders Systemic disease associated

Table 3: Adapted from Pyoderma gangrenosum: clinicopathologic correlation and proposed diagnostic criteria [9]

Criteria by Su *et al*., (2004)	
Major Criteria	Rapidly developing painful, necrolytic ulcer with an irregular, violaceous, and undermined border Ruled orogressing disease (rapidly)
Minor Criteria	History suggestive of pathergy or cribriform scarring on examination Underlying systemic disease with known association to PG Histopathology consistent with PG (sterile neutrophilic dermal infiltrate, mixed infiltrates, lymphocytic vasculitis) Rapid response to systemic CStrauma

Treatment for pyoderma gangrenosum is based on systemic immunosuppression, pain control, local wound management, and treatment of associated comorbidities. The choice of treatment depends on a variety of factors, including the extent, severity, and location of the lesions, as well as the presence of underlying diseases.

It is generally accepted that systemic glucocorticoids (prednisone or equivalents) and cyclosporine are considered first-line therapies, with comparable efficacy according to clinical trials and systematic reviews. Typically, systemic corticosteroids are initiated at doses ranging from 0.5 to 1 milligrams per kilogram per day, with a subsequent adjustment based on the patient's clinical response. It is recommended that the dosage of cyclosporine be adjusted to 2–5 mg/kg/day, as it has been found to be an effective alternative or in combination with corticosteroids. This approach has been observed to reduce exposure to corticosteroids and their associated adverse effects [1-5].

In cases where these treatments are not effective or in cases where there are additional health concerns, such as inflammation, biological agents have become an option. These agents, including TNF-α inhibitors like infliximab and adalimumab, have shown promise in both idiopathic PG and PG associated with inflammatory bowel disease. In cases where other biologics targeting IL-1, IL-17, IL-23, or complement C5a might be considered, there is a possibility that they could be effective [2-10].

There are other immunosuppressive options that could be considered, including mycophenolate mofetil, dapsone, azathioprine, methotrexate, thalidomide, and cyclophosphamide. These are reserved for cases that do not respond to other treatments or for those with contraindications to first-line treatments [5-10].

Local management of lesions is essential, and it includes non-adhesive dressings, control of exudate, prevention of secondary infections, and analgesia. It is advisable to exercise caution and avoid overly aggressive surgical debridement, given the potential risk of pathergy [10].

CASE PRESENTATION

A 74-year-old woman was seen with a skin condition on the right side of her head that had been there for about 6 months. It had started as a small bump that had grown into an ulcer.

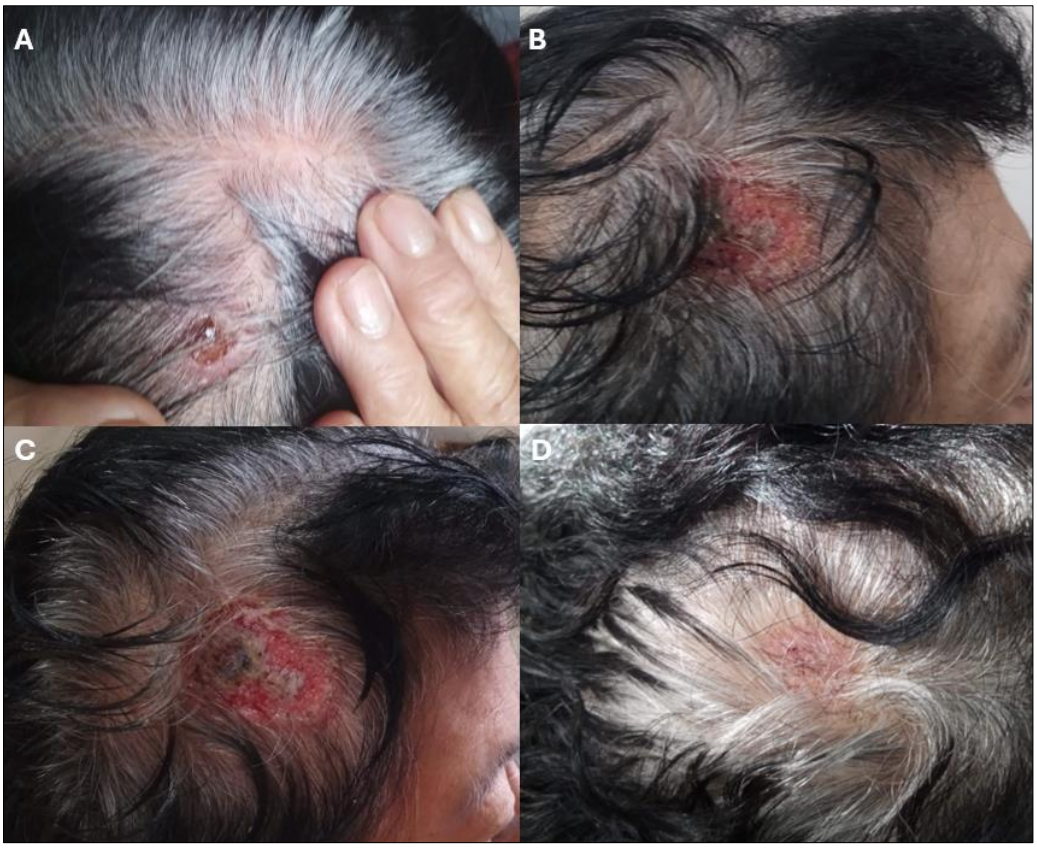
It is important to note that she had a history of poorly controlled systemic hypertension.

An ulcer secretion culture was performed outside the institution, and it was reported that Pseudomonas aeruginosa was present, which is susceptible to ceftazidime. She received treatment for this, but unfortunately, there was no improvement.

During the study protocol, a biopsy of the ulcer was performed, taking a sample from the edge of the ulcer, with histopathological study stained with hematoxylin and eosin. This study revealed a dense neutrophilic infiltrate at the dermis level, which led to the diagnosis of gangrenous skin. Antibody studies, imaging studies of the extent, and endoscopy were performed, but no abnormalities were detected.

The patient received treatment with prednisone 50 mg every 24 hours for 14 days, followed by weekly dose reductions. The patient's dermatosis showed signs of improvement two months later.

The patient remained hospitalized for a period of one month due to a number of infections associated with healthcare and uncontrolled hypertension. Her stay allowed us to document the rapid evolution of her condition after the start of systemic steroid treatment.



The evolution of the lesion is also documented. Image A is a photograph taken by the patient on day 4, image B is a photograph taken on day 7 during the medical examination, image C is on day 10, and image D is a photograph taken 3 months after the initial injury.

DISCUSSION

Pyoderma gangrenosum is a rare condition, with a higher incidence in reported cases of women aged 40–50 years. In this particular instance, the patient's age exceeded the stated age range, they did not have a history of autoimmune diseases, and neoplastic pathology was excluded.

It is my understanding that ulcerative pyoderma gangrenosum is associated with the pathergy phenomenon, which is an inflammatory skin reaction induced by microtrauma that leads to epidermal disruption. This, in turn, promotes a cascade of inflammatory cytokines that cause lesions that begin as papules or pustules that ulcerate, with a characteristic undermined and violaceous border. If I'm not mistaken, histopathology describes a neutrophilic infiltrate as reported in our case.

After careful consideration, the diagnosis was determined through a combination of clinical and anatomical correlation with classic pyoderma gangrenosum. It is important to acknowledge that, in the past, the diagnosis of pyoderma gangrenosum was often made through a process of elimination. However, advancements in medical technology have led to the development of new diagnostic tools that facilitate more precise diagnoses. Upon admission, the patient had a Paracelsus score of 14 points, and according to the Delphi method, the major criterion was present in the biopsy, with three additional criteria noted. Following a thorough histopathological study and a treatment plan that included steroids, she was discharged with a Paracelsus score of 19 points. According to the Delphi method, one major criterion and five points were identified.

Given its low incidence, pyoderma gangrenosum is not necessarily the first diagnosis considered when a culture is positive. In our patient, despite the administration of antibiotics guided by an antibiogram, there was only a slight improvement in the lesions. Therefore, when pyoderma gangrenosum is suspected, histopathological study supports the diagnosis. The criteria used to diagnose the disease are a useful tool, but they must be individualized in patients with superinfection, as in our case.

CONCLUSION

Pioderma gangrenosum is a non-infectious inflammatory disease characterized by pustules that rapidly progress to painful necrotic ulcers with undermined and violaceous edges, with neutrophilic infiltrate reported on biopsy. There are a variety of criteria that can be used for diagnosis, including the PARACELSU, DELPHI, or Su criteria.

Treatment options include systemic immunosuppression, recognizing that this could be the initial presentation of an intestinal inflammatory disease.

REFERENCES

1. Maverakis, E., Marzano, A. V., Le, S. T., Callen, J. P., Brüggen, M. C., Guenova, E., Dissemond, J., Shinkai, K., & Langan, S. M. (2020). Pyoderma gangrenosum. Nature reviews. Disease primers, 6(1), 81. https://doi.org/10.1038/s41572-020-0213-x
2. Maronese, C. A., Pimentel, M. A., Li, M. M., Genovese, G., Ortega-Loayza, A. G., & Marzano, A. V. (2022). Pyoderma Gangrenosum: An Updated Literature Review on Established and Emerging Pharmacological Treatments. American journal of clinical dermatology, 23(5), 615–634. https://doi.org/10.1007/s40257-022-00699-8
3. Plumptre, I., Knabel, D., & Tomecki, K. (2018). Pyoderma Gangrenosum: A Review for the Gastroenterologist. Inflammatory bowel diseases, 24(12), 2510–2517. https://doi.org/10.1093/ibd/izy174
4. Lee, S., Lee, J. Y., Ju, H. J., Lee, J. H., Koh, S. B., Bae, J. M., & Han, J. H. (2023). Association of All-Cause and Cause-Specific Mortality Risks With Pyoderma Gangrenosum. JAMA dermatology, 159(2), 151–159. https://doi.org/10.1001/jamadermatol.2022.5437
5. Łyko, M., Ryguła, A., Kowalski, M., Karska, J., & Jankowska-Konsur, A. (2024). The Pathophysiology and Treatment of Pyoderma Gangrenosum-Current Options and New Perspectives. International journal of molecular sciences, 25(4), 2440. https://doi.org/10.3390/ijms25042440
6. Becker, S. L., Vague, M., & Ortega-Loayza, A. G. (2025). Insights into the Pathogenesis of Pyoderma Gangrenosum. The Journal of investigative dermatology, 145(6), 1305–1322. https://doi.org/10.1016/j.jid.2024.09.023
7. Maverakis, E., Ma, C., Shinkai, K., Fiorentino, D., Callen, J. P., Wollina, U., Marzano, A. V., Wallach, D., Kim, K., Schadt, C., Ormerod, A., Fung, M. A., Steel, A., Patel, F., Qin, R., Craig, F., Williams, H. C., Powell, F., Merleev, A., & Cheng, M. Y. (2018). Diagnostic Criteria of Ulcerative Pyoderma Gangrenosum: A Delphi Consensus of International Experts. JAMA dermatology, 154(4), 461–466. https://doi.org/10.1001/jamadermatol.2017.5980
8. Jockenhöfer, F., Wollina, U., Salva, K. A., Benson, S., & Dissemond, J. (2019). The PARACELSUS score: a novel diagnostic tool for pyoderma gangrenosum. The British journal of dermatology, 180(3), 615–620. https://doi.org/10.1111/bjd.16401
9. Su, W. P., Davis, M. D., Weenig, R. H., Powell, F. C., & Perry, H. O. (2004). Pyoderma gangrenosum: clinicopathologic correlation and proposed diagnostic criteria. International journal of dermatology, 43(11), 790–800. https://doi.org/10.1111/j.1365-4632.2004.02128.x
10. Nishimura, M., Mizutani, K., Yokota, N., Goto, H., Akeda, T., Kitagawa, H., Habe, K., Hayashi, A., & Yamanaka, K. (2022). Treatment Strategy for Pyoderma Gangrenosum: Skin Grafting with Immunosuppressive Drugs. Journal of clinical medicine, 11(23), 6924. https://doi.org/10.3390/jcm11236924