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# Extensive Perianal Pyoderma Gangrenosum, Diagnostic-Therapeutic Approach to A Complicated Patient. A Case Report

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## ABSTRACT

**Background:** Pyoderma gangrenosum is a rare neutrophilic dermatosis with very low incidence and varied clinical presentation associated with different comorbidities, also a diagnostic-therapeutic challenge. This article reports an individualized and multidisciplinary diagnostic-treatment approach of a relevant additionally infrequent extensive perianal pyoderma gangrenosum case.

**Clinical Case:** A 79-year-old female patient with 18 months dermatosis evolution began as a perianal pustule and progressed to a 20 cm in diameter ulcer was presented. Pyoderma gangrenosum diagnosis was established based on international accepted criteria. Due to her comorbidities moreover systemic complication, she received multidisciplinary and individualized topic treatment with satisfactory evolution.

**Conclusions:** Pyoderma gangrenosum is frequently associated with a systemic disease, consequently it must be approached from a systemic perspective with the support of a multidisciplinary team to provide the most appropriate treatment.

**Keywords:** Pyoderma gangrenosum, neutrophilic dermatosis, peptic ulcer hemorrhage, hypothyroidism, case report.

## BACKGROUND

Pyoderma gangrenosum is a chronic inflammatory neutrophilic dermatosis characterized by single or multiple, painful ulcers, with varied clinic presentation depending on the infiltrated regions[1] and its approach continues to be an exclusion diagnosis.[2] The incidence is 3-10 patients per 1 million worldwide.[3]

Etiology is poorly understood, complex, multifactorial and involves genetic interaction, environmental factors, immune dysregulation, altered neutrophil chemotaxis, keratinocyte apoptosis with the release of molecular patterns associated with damage.[4] It is estimated that in up to 77% of cases there is an underlying systemic disease.[5] Inflammatory bowel disease, rheumatoid arthritis, and hematological malignancies, among others, are the more frequent associated diseases.[2] There are five clinical variants: classic (ulcerous), bullous, vegetative, pustular, and peristomal[6] bring about a diagnostic challenge. Delphi criteria with 86% sensitivity and 90% specificity were recently defined, (Table 1)[7] and PARACELSUS score in order to improve the diagnostic approach (Table 2).[2]

**Table 1:** Delphi criteria. Positive by having the major criterion and 4 minors.[7] The patient presents the major criterion and 5 minor criteria.

| **Major criterion** |
|---|
| 1. Biopsy with neutrophilic infiltrate |
| **Minor criteria** |
| 1. Exclusion of infection |
| 2. Pathergy |
| 3. Personal history of Inflammatory Bowel Disease or inflammatory arthritis |
| 4. History of papule, pustule, or vesicle that rapidly ulcerates within 4 days of appearing |
| 5. Peripheral erythema, undermining border, and tenderness at ulceration site |
| 6. Multiple ulcerations (at least one on anterior lower leg) |
| 7. Cribriform or “wrinkled paper” scar (s) at healed ulcer sites |
| 8. Decrease in ulcer size after immunosuppressive treatment |

**Table 2:** PARACELSUS SCORE. Suggestive of Pyoderma=10 or more points. Venous ulcers = < 7 points.[2] The patient has 13 points

| | |
|---|---|
| P | Progressing disease (ulcer developing within 6 weeks). Mayor. 3 points |
| A | Assessment of relevant differential diagnoses. Mayor. 3 points |
| R | Reddish-violaceous wound margin. Mayor. 3 points |
| A | Amelioration (Alleviation) by immunosuppressant drugs. Minor. 2 points |
| C | Characteristically irregular (bizarre) ulcer shape. Minor. 2 points |
| E | Extreme pain >4/10 on visual analogue scale. Minor. 2 points |
| L | Localization of lesion at site of trauma (pathergy). Minor. 2 points |
| S | Suppurative inflammation in histopathology. Additional. 1 point |
| U | Undermined wound border. Additional. 1 point |
| S | Systemic disease associated. Additional. 1 point |

Skin ulcers are mainly located in the pretibial area, chest, hands, head, neck, and peristomal skin, and infrequently in the perianal area.[8] Their therapeutic response can be clinically evaluated by Gulliver's sign, due to epithelial growth in the ulcer edge towards the center leaving cribriform scars.[9]

Histopathology of early lesions classically shows deep suppurative, folliculocentric inflammation, dense neutrophilic infiltrates, and leukocytoclastic vasculitis; then, the mixed inflammatory infiltrate predominates with neutrophils and epidermal ulceration, however, it is an inconsistent finding, not pathognomonic.[10]

Regarding treatment, there is currently no drug approved by the FDA, traditionally therapies include corticosteroids and equally important other immunosuppressants, either local or systemic and recently biological therapies are the trend[2] however there is lacking scientific evidence from large clinical trials that evaluate safety, efficacy, adverse events and outcomes.

Pyoderma gangrenosum is a rare disease,[3,4] which makes it challenging to carry out large clinical trials and adequate statistical significance observational studies. Hence, observational studies such as case reports, case series, and trials in reference centers, among other strategies, become relevant[11] to evaluate the diagnostic-therapeutic approach, reporting their outcomes toward establishing records to provide the basis for future extended review development.

Therefore, a case of extensive perianal pyoderma gangrenosum in the presence of gastrointestinal bleeding secondary to duodenal ulcer and hypothyroidism is presented, highlighting the importance of an individualized diagnostic-therapeutic approach based on comorbidities and clinical history.

## PRESENTATION OF THE CASE

A 79-year-old female patient with a history of primary hypothyroidism, systemic arterial hypertension, peptic acid disease, hemorrhoidal disease and bilateral gonarthrosis with chronic use of non-steroidal anti-inflammatory drugs during 1 year was referred to our unit for ulcerative dermatosis.

On physical examination, she presented a localized dermatosis in the perianal region consisting of a 20 cm approximately ulcer with undermined and erythematous-violaceous edges with fibrinous tissue associated. She began her condition 18 months before her admission, with a pustule that progressed to a painful ulcer. Figure 1. At the beginning of the dermatosis, she was treated with a torpid evolution due to diagnostic-therapeutic approach. Then, she went to a dermatologist, where a skin biopsy was performed which reports leukocytoclastic vasculitis. Figure 2. Upon admission to the hospital, the patient presented pale skin and low cardiac output data, so she was admitted to the internal medicine service, protocolized by the endoscopy service for suspected inflammatory bowel disease, observing digestive bleeding secondary to duodenal ulcer, and an endoscopy is performed. Pyoderma

gangrenosum treatment started based on high-potency topical steroid due to the impossibility to use systemic treatment with improvement.

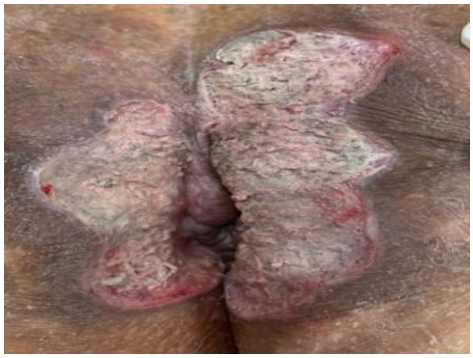

**Figure 1:** Perianal ulcer characterized by irregular, violaceous, raised borders, with a granulomatous background.

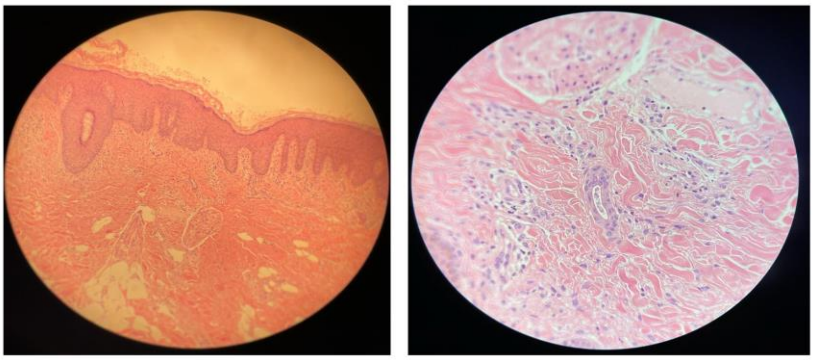

**Figure 2:** Skin biopsy showing epidermis with lamellar horny layer, horny plugs, moderate irregular acanthosis, dermis with vessels with fibrinoid degeneration in its wall, surrounded and invaded by neutrophils, moderate and diffuse infiltrate between collagen fibers by lymphocytes, histiocytes, epithelioid cells and neutrophils.

## DISCUSSION

The diagnostic-therapeutic approach to rare pyoderma gangrenosum location associated with comorbidities was presented, based on the Delphi and PARACELSUS scores the diagnostic was established, which are currently internationally accepted, generating homogeneity at the time of reporting the results, reducing the time of diagnosis. And start of treatment, however, future studies of greater statistical relevance are required to evaluate the above.

Hypothyroidism associated with this disease is new in the medical literature, being added to its list of systemic associations, so attention should be paid in future reviews, since it is relevant to the disease pathophysiology and its relationship with the decrease in collagen and hydroxyproline in wounds during the inflammatory phase and the proliferative phase, interfering with the proliferation and secretion of fibroblasts in the wound healing process.[11] We emphasize maintaining endocrinological control to improve healing conditions, however there is a lack of scientific evidence on its relationship and clinical results.

Regarding treatment, in superficial and localized forms, topical treatment has shown an excellent response,[12] observing in a prospective study that when using topical tacrolimus 0.03% and clobetasol 0.05% 44% had a response at 6 months, 33% required systemic treatment and 15% had recurrent disease,[13] considering that steroids increase the risk of bleeding or perforation in hospitalized patients[14] we opted for clobetasol only, observing improvement of the lesion in a 3-month follow-up.

Finally, as there are no international guidelines that evaluate the results in patients with comorbidities that contraindicate the use of systemic and biological therapies, the use of topical therapy was justified and the clinical result was favorable. The foregoing may form the basis for future reviews and research projects focused on topical treatment in complicated patient that contraindicates the systemic treatment, however, our patient should be followed up for better evidence of the outcome.

This article has well-established limitations as it is a case report, so the results presented here should be taken with caution. However, international guidelines (CARE guidelines) were used to report them, thus increasing their internal validity. On the other hand, the use of this resource acquires epidemiological relevance in rare diseases such as this case.[11]

## CONCLUSIONS

Pyoderma gangrenosum is a neutrophilic dermatosis with a diagnostic-therapeutic challenge, notwithstanding there are few clinical studies due to the low prevalence of the disease, leading to limited information or with little statistical relevance on the clinical outcome and response to treatment. On the other hand, pyoderma gangrenosum in the perianal area in the presence of multiple comorbidities has not been reported yet in the literature. The recent standardization of diagnostic criteria is a tool that can facilitate timely diagnosis and treatment, in order to avoid the risk of developing squamous cell carcinoma.[4]

It is frequently associated with a systemic disease, so it must be approached from a systemic perspective with the support of a multidisciplinary team in order to provide the best treatment.

Patient perspective: During my hospitalization, I was approached by multiple specialties which evaluated each of my diseases in order to give an appropriate and individualized treatment. I went to several doctors for more than a year; however, it was not until this last hospitalization that I was able to improve.

## CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest.

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