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Simultaneous Perianal Manifestations And Pyoderma Gangrenosum Revealing Crohn's Disease

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Abstract

Up to 40% of patients with inflammatory bowel disease (IBD) develop an extraintestinal manifestation of the disease with the skin being the most commonly involved organ. Pyoderma gangrenosum (PG), an autoinflammatory non-infectious neutrophilic dermatosis, occurs in 1–2% of patients with IBD. PG can follow a course independent to that of the bowel disease, however, most reported cases describe PG occurring in patients with an established diagnosis of IBD. We present a case of a young patient who presented with axillary skin ulceration, which was subsequently diagnosed as PG. On further investigation for a possible underlying cause, she was found to have Crohn's disease. She had not developed any preceding change in her bowels and did not have abdominal pain; the IBD was diagnosed on endoscopic findings. This case is also unusual for the distribution of the PG lesions that typically occur in the lower limbs.

Keywords: Crohn's disease, Pyoderma gangrenosum, extraintestinal manifestations, perianal manifestations, inflammatory bowel disease.

Introduction

Extraintestinal manifestations (EIMs) occur in up to 50% of patients with inflammatory bowel disease (IBD). They can affect practically any body system, from the eyes to the skeletal system, and have a significant impact on the number of symptoms experienced by patients. The majority of EIMs have a direct connection to an ongoing intestinal flare. This comprises episcleritis, erythema nodosum (EN), pauciarticular arthritis, and aphthous ulcers. Other EIMs, such as uveitis and ankylosing spondylitis (AS), are unrelated to intestinal disease activity.[4]

Our case reports a pyoderma gangrenosum that coincided with Ano perineal manifestations revealing Crohn's disease in a misdiagnosed patient.

Case presentation

During a follow-up consultation, a 24-year-old patient, who had been treated in our department for a year for UC discovered following severe acute colitis during pregnancy and initially put on 5-ASA with very good clinical improvement, presented with painful skin lesions that had been evolving for 1 week, as well as proctalgia and purulent anal emissions.

A proctological examination revealed a large anal fissure, with edematous detached edges and a major inflammatory component. We also noted the presence of an anal fistula at 10 o'clock, surmounted by an

intraductal bud with intermittent pus emission.
Meticulous mucocutaneous examination revealed several lesions in the form of necrotic ulcers, with well-defined, purplish hypertrophic margins. The surrounding skin tissue is erythematous, infiltrated and indurated.

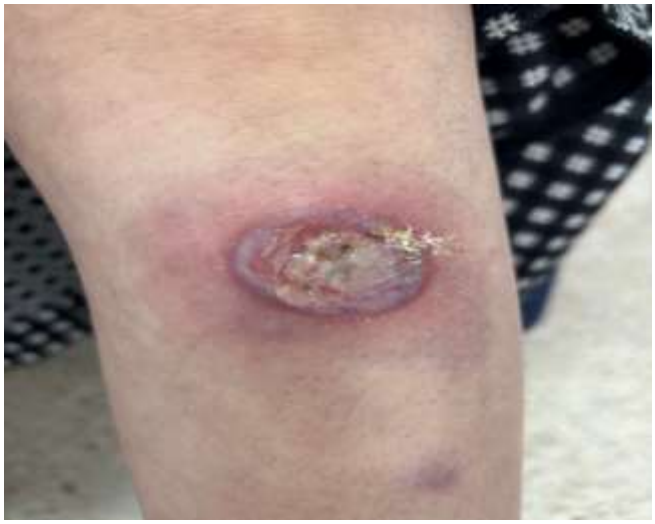


Figure 1 Necrotic ulcer on the leg

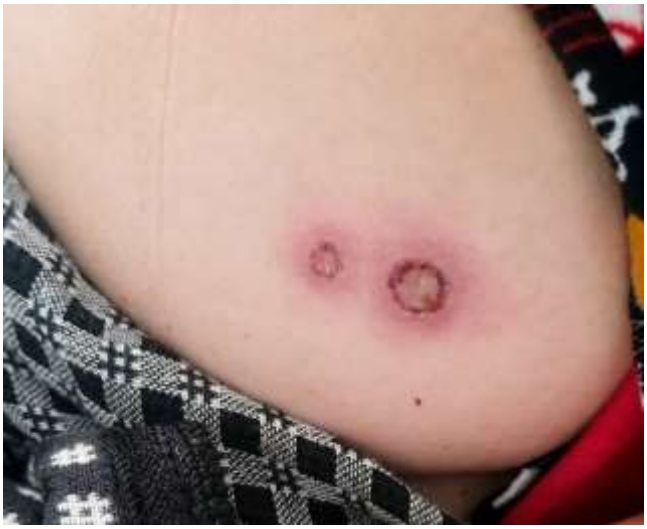
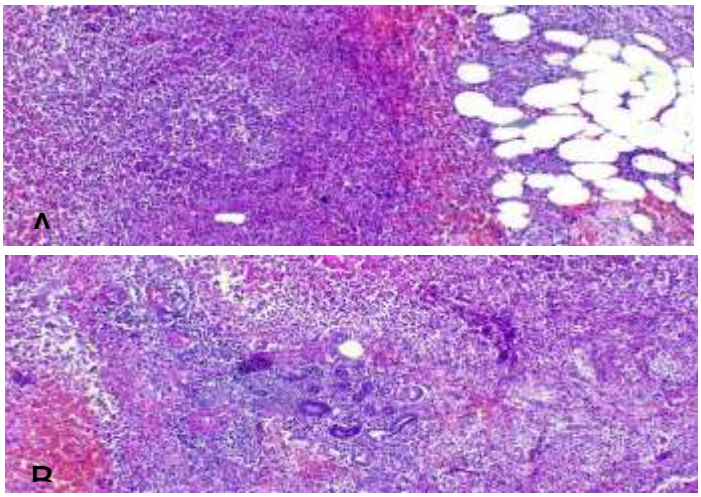


Figure 2 Necrotic ulcers on the upper-external quandrant of the right buttock

The largest is located on the anterior aspect of the left leg, measuring 6 cm in diameter.(Figure 1) The rest of the lesions are located on the upper-external quadrant of the right buttock (Figure 2).

Iniatial blood tests revealed a microcytic anaemia (haemoglobin 9.5 g/dL), hypoalbuminemia (21 g/L) and a high C reactive protein (166 mg/L). Her renal and liver function tests were normal.
A skin biopsy was performed on the leg lesion. The diagnosis of UC was changed to Crohn's disease, and the patient was put on ciprofloxacin and metronidazole to dry out the ano-perineal lesions.



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Anatomopathological results revealed a deep suppurative folliculitis with a dense neutrophilic infiltrate confirming the diagnosis of pyoderma gangrenosum. (Figure 3)

The indication for systemic and topical corticosteroid therapy has been established. However, with the patient ongoing active fistula, it was put on hold. The evolution was marked by the drying of the anal fistula after 5 days and complete healing of the anal fissure.

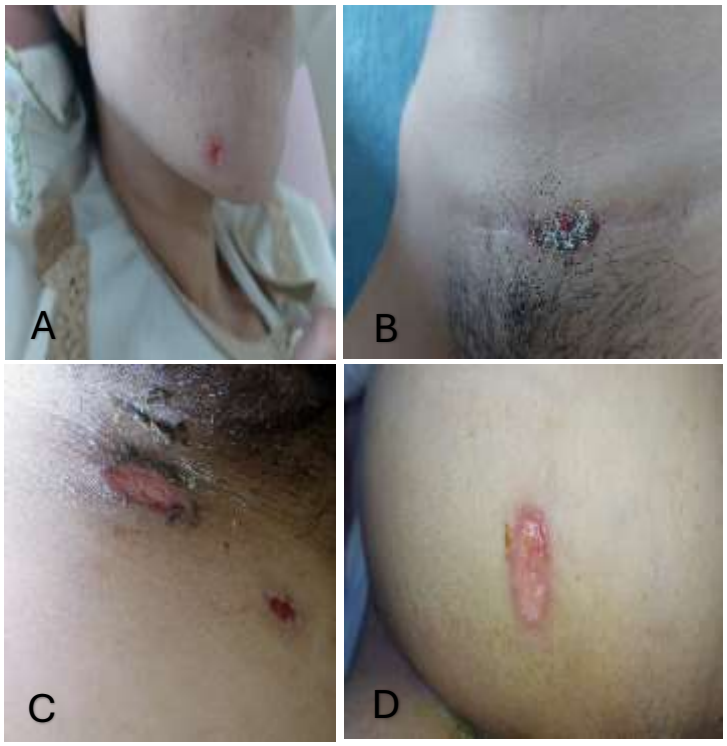

Figure 4 : A. Ulcerative lesion of the chin B. ulcerative lesion on caesarean scar C. ulcerative lesions on the groin D. ulcerative lesion on the buttock

During this period, the patient developed further ulcerative lesions on her chin, groin fold, genitals and buttocks,(figure 4) as well as pustules on both ankles and the elbow (Figure6) and worsening of existing lesions.(Figure 5)

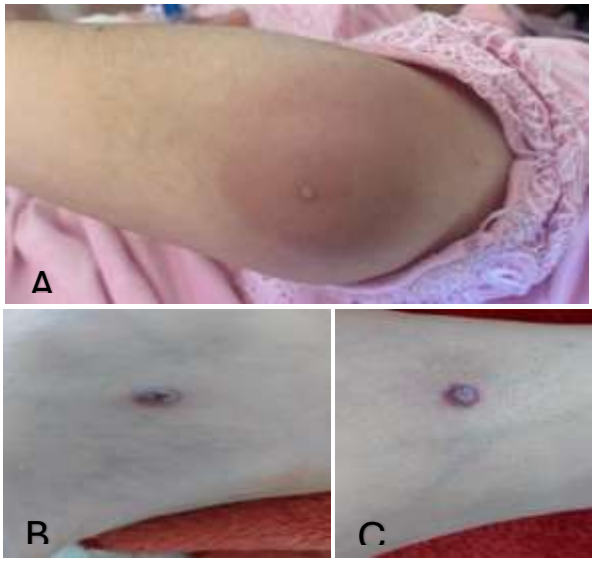

Figure 5 : A. Pustula on the elbow

B. Pustula on the left ankle C. Pustula on the right ankle

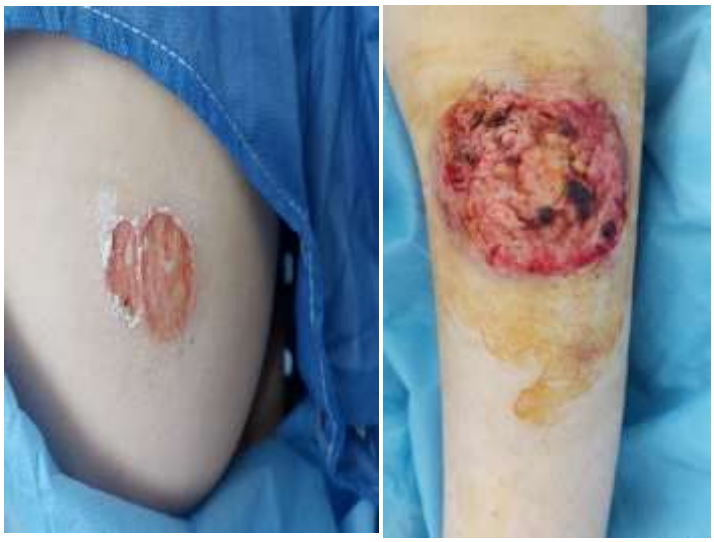

Figure 6 : worsening of lesions on the front of the leg and buttocks

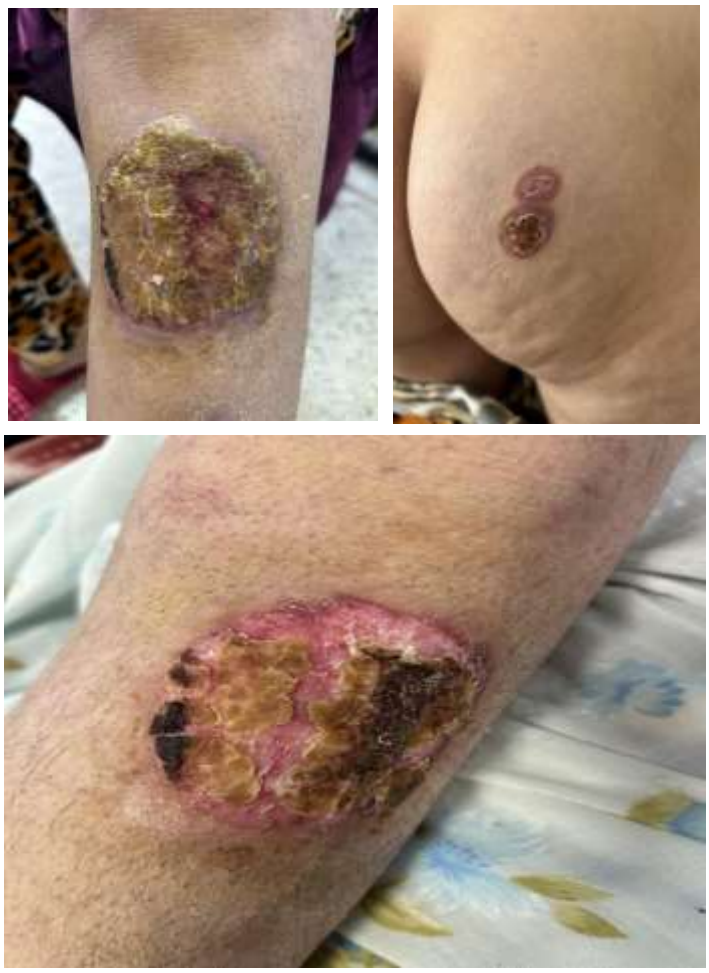

Figure 7 : A. Evolution after one month

B. Evolution after two months

We were able to start prednisolone-based corticosteroid therapy at 1 mg/Kg/day with a good improvement in skin lesions. (Figure 6) The patient was later discharged on corticosteroids and was put on infliximab. The evolution was marked by healing of the lesion. (Figure 7)

Discussion

Pyoderma gangrenosum (PG) is an ulcerative form of neutrophilic dermatosis of unknown etiology.
The pathophysiological mechanisms of PG remain very hypothetical at present. Nearly half of the cases involve patients with systemic inflammatory diseases. In 20 to 60% of cases, patients with PG have a digestive inflammatory disease such as Crohn's disease or ulcerative colitis.[2]In contrast to erythema nodosum, pyoderma gangrenosum was more common in UC than Crohn's disease with a female predominance. [3, 10] The colonic location of the disease has been linked to this complication.[6]
The prevalence of PG in chronic inflammatory bowel disease is estimated to be between 0.4 and 2%.[8] In the majority of cases, pyoderma presented in known IBD patients.[9]PG often presents as an erythematous lesion centered by a pustule that rapidly evolves into an ulcerated lesion with irregular, purplish borders. The bottom of the ulcer contains purulent material, sterile in culture.[10]

The areas most affected are the extensible surfaces of the legs (shins) and areas adjacent to a post-surgical stoma, but other locations have been reported including the genitalia. [6]

The diagnosis of PG is based primarily on clinical signs, and is supported by a biopsy for histopathological purposes. The patient's history must be taken in order to detect any underlying disease, and specific examinations must be carried out on the basis of this history. PG is a diagnosis of exclusion.There are no laboratory parameters . The histopathology of PG is non-specific and evolves according to the stage of the lesion. Initial lesions present a deep suppurative folliculitis with a dense neutrophilic infiltrate.[11] There is no gold standard for the treatment of PG. PG has both a systemic inflammatory component and a wound-related component, so an effective treatment strategy must address both processes.

PG lesions are painful. Pain management should include judicious use of NSAIDs, opioids, pain specialists and psychiatric consultants if necessary for patients with chronic pain or depression.[5]

A mix of wound care, topical therapy, and systemic therapy may be used to treat PG. Every lesion requires specific wound care based on the kind of lesion and level of exudation. It is frequently advised to keep an area moist to encourage wound healing. Additionally, it's critical to look out for symptoms of overlaid illnesses.

For localized, moderate disease, topical therapy can be used alone; however, for more severe lesions, topical therapy can be combined with systemic therapy. Topical treatments such as corticosteroids, tacrolimus, ciclosporin, 5-aminosalicylic acid, and dapsone have been used with some success.

The literature shows that systemic corticosteroids (prednisone 0.5-1 mg/kg/day, methylprednisolone up to 0.8 mg/kg/day) are effective in a large number of cases and are the first-line systemic treatment.[7]

Biological agents are useful therapeutic options for PG and are frequently used to treat specific associated pathologies, notably Crohn's disease. Infliximab, an anti-TNFα monoclonal antibody that binds to both soluble and membrane-bound TNFα, is the only biological agent that has shown efficacy in classic PG.[1]

Conclusion

In summary, extraintestinal symptoms are a common symptom of inflammatory bowel disease (IBD). Large, painful ulcers are the primary presenting feature of pyoderma gangrenosum; histology is supportive of the clinical diagnosis. Although the course of pyoderma gangrenosum can differ from that of IBD, treatment for both diseases entails wound care and immunosuppressive drugs.

References

1. 1Brooklyn TN, Dunnill MGS, Shetty A, Bowden JJ, Williams JDL, Griffiths CEM, et al. *Infliximab for the treatment of pyoderma gangrenosum: a randomised, double blind, placebo controlled trial*. 2006;55(4):505-509.
2. Carron DP-N. *P.-N. Carron S.Yerly R. Ksontini P. Meylan T. Calandra*. 2008;
3. Freeman HJ. *Erythema nodosum and pyoderma gangrenosum in 50 patients with Crohn's disease*. Can J Gastroenterol. 2005;19(10):603-606.
4. Malik TF, Aurelio DM. *Extraintestinal Manifestations of Inflammatory Bowel Disease*. In: StatPearls. Treasure Island (FL): StatPearls Publishing; 2023. Disponible sur: http://www.ncbi.nlm.nih.gov/books/NBK568797/.
5. Miller J, Yentzer BA, Clark A, Jorizzo JL, Feldman SR. *Pyoderma gangrenosum: A review and update on new therapies*. Journal of the American Academy of Dermatology. 2010;62(4):646-654.

6. Orchard T. *Extraintestinal complications of inflammatory bowel disease*. Curr Gastroenterol Rep. 2003;5(6):512-517.
7. Reichrath J, Bens G, Bonowitz A, Tilgen W. *Treatment recommendations for pyoderma gangrenosum: An evidence-based review of the literature based on more than 350 patients*. Journal of the American Academy of Dermatology. 2005;53(2):273-283.
8. Vavricka SR, Brun L, Ballabeni P, Pittet V, Prinz Vavricka BM, Zeitz J, et al. *Frequency and risk factors for extraintestinal manifestations in the Swiss inflammatory bowel disease cohort*. Am J Gastroenterol. 2011;106(1):110-119.
9. Vavricka SR, Rogler G, Gantenbein C, Spoerri M, Prinz Vavricka M, Navarini AA, et al. *Chronological Order of Appearance of Extraintestinal Manifestations Relative to the Time of IBD Diagnosis in the Swiss Inflammatory Bowel Disease Cohort:* Inflammatory Bowel Diseases. 2015;21(8):1794-1800.
10. Vavricka SR, Schoepfer A, Scharl M, Lakatos PL, Navarini A, Rogler G. *Extraintestinal Manifestations of Inflammatory Bowel Disease*. 2015;21(8):1982-1992.
11. Wollina U. *Pyoderma gangrenosum – a review*. Orphanet J Rare Dis. 2007;2:19.