ECRONICON OPEN ACCESS

EC GASTROENTEROLOGY AND DIGESTIVE SYSTEM

Case Report

# Peristomal Pyoderma Gangrenosum in Crohn's Disease: Case Report

**W Hliwa*, M Ettachfini, S Banani, F El Rhaoussi, M Tahiri, F Haddad, A Bellabah and W Badre**

*Department of Gastroenterology, University Hospital Ibn Rochd, Casablanca, Morocco*

***Corresponding Author:** W Hliwa, Department of Gastroenterology, University Hospital Ibn Rochd, Casablanca, Morocco.

**Received:** May 08, 2023; **published:** May 31, 2023

## Abstract

**Introduction:** Peristomal PG (PPG) is a dreaded and (fortunately) rare subtype of PG that develops on the skin adjacent to a stoma. There are few case reports describing this association. It is a challenging condition to diagnose and treat and no evidence-based guidelines exist. We report a case report of a patient with crohn's disease who had presented a peristomal pyoderma gangrenosum.

**Observation:** A 22-year-old woman, with a history of ileocolonic and perianal crohn's disease classified A2L3B3P according to the montreal classification, associated with ocular and articular involvement since 2014. She is currently on a combination therapy (Infliximab+ azathioprine). Due to the severity of anoperineal lesions, and non-response to combination therapy the patient underwent surgery for sigmoidal colostomy. Two years later the patient presented a peristomal PPG confirmed on histology. The treatment consisted on local corticotherapy associated with local care and parenteral nutrition, albumin human perfusions and infliximab 5 mg/kg with good clinical evolution.

**Conclusion:** Peristomal PG during IBD is an exceptional localization. Early diagnosis and treatment is important to avoid disease progression and subsequent patient discomfort. Its management is poorly codified and empirical.

***Keywords:*** *Peristomal Pyoderma Gangrenosum; Crohn's Disease; Local Corticotherapy; Infliximab*

## Abbreviations

PG: Pyoderma Gangrenosum; PPG: Peristomal Pyoderma Gangrenosum; CDEIS: Crohn's Disease Endoscopic Index Score

## Introduction

Pyoderma gangrenosum (PG) is a neutrophilic dermatosis considered as a common extraintestinal lesion associated with IBD. Peristomal PG (PPG) is a rare localization of PG that makes stoma management extremely difficult. There are few case reports describing this situation, due to the presence of other differential diagnosis related to stomial complications. There are no evidence based guidelines in the literature for the diagnosis and the therapeutic management. We report a medical case of a young patient with crohn's disease who presented a peristomal pyoderma gangrenosum.

## Observation

A 22-year-old woman, with a history of ileocolonic and perianal crohn's disease, classified as A2L3B3P according to the montreal classification, associated with ocular and articular manifestations since 2014.

She is currently on a combination therapy (Infliximab and azathioprine), but due to the severity of anoperineal lesions, and non-response to medical treatment, the patient underwent surgery for sigmoidal colostomy.

Two years later, the patient was admitted to our department for painful peristomal ulcerations prohibiting the placement of the colostomy pouch, the clinical examination of the peristomal skin revealed two large ulcerations with an oozing and fibrinous base with raised purplish margins in the peristomal level, the largest one was linear and measured approximately 4 cm in diameter with retraction of the stomial orifice and vegetated skin opposite associated with echymotic plaque with central necrotic (Figure 1), a PPG was suspected.

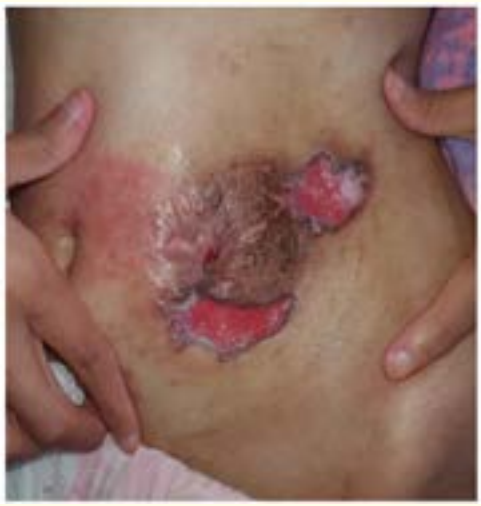

***Figure 1:*** *Parastomal pyoderma gangrenosum.*

Biopsies of the skin lesion revealed: neutrophilic and granulomatous dermatitis, compatible with a pyoderma gangrenosum in its granulomatous subtype.

Laboratory tests shows an inflammatory syndrome associated with malabsorption.

A colonoscopy was performed showing an endoscopic aspect compatible with a severe ileocolic crohn's disease (CDEIS score 34).

The treatment consisted on local corticotherapy: 1 application per day associated with local care by vaseline and parenteral nutrition, albumin human perfusion and infliximab 5 mg/kg with good clinical evolution marked by healing of the lesions from the second week (Figure 2 and 3).

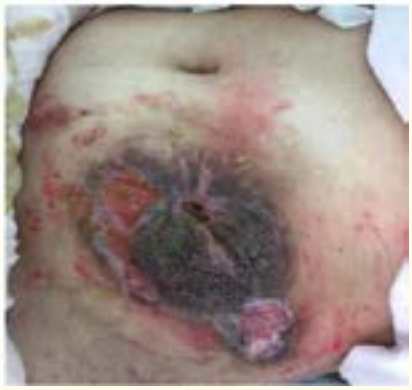

***Figure 2:*** *Parastomal pyoderma gangrenosum after one week of treatment.*

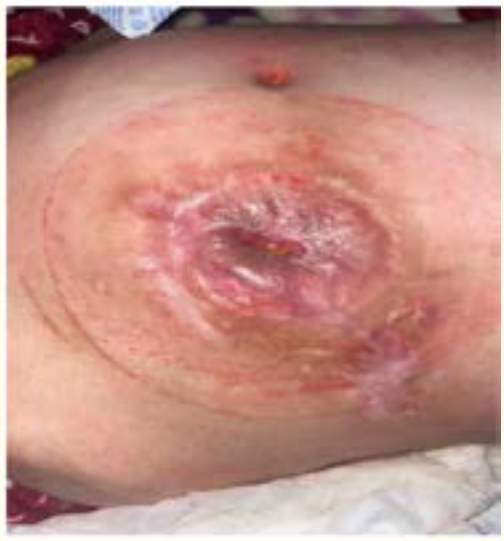

***Figure 3:*** *Peristomal pyoderma gangrenosum at the end of the treatment.*

## Discussion

Pyoderma gangrenosum is a severe extra-intestinal manifestation of IBD with painful lesions and scarring, requiring in most of the case the hospitalization of the patient [1].

It occurs in 0.75 - 1.5% of IBD patients [2,3], more frequently in crohn's disease, which is more often pancolic, but is not correlated with the severity of the intestinal disease. Conversely, IBD is the primary cause of PG (20 - 30% of cases) [4]. 2 to 4% of patients with IBD and a stomy, can develop peristomal pyoderma gangrenosum (PPG), this type of PG (PPG) is favored by the various aggressions to which the peristomal skin is subjected [5], and it's most frequently located in the lower extremities (77,7%) [6,7].

In terms of clinical presentation, PPG manifests initially as painful pustules that increase in size forming ulcers with purple edges and a necrotic base. PPG may be multiple and keep scars after healing [8].

The diagnosis of PPG is made clinically and requires the exclusion of other causes to institute the appropriate management [9].

Differential diagnoses of PPG can be either:

- Early postoperative excoriation that could be: a stitch abscess, a poorly fashioned stomy allowing irritation from the ileal effluent, allergic contact dermatitis, a leaking appliance, an inappropriately placed stomy, or recurrent Crohn's disease and fistulization should be ruled out with the appropriate endoscopy or radiography studies [9].
- Chronic ulcerating, include infectious diseases, malignancy, vasculitis [10,11].

Skin biopsy and culture are valuable in the diagnosis of PPG, it is highly recommended to rule out other conditions since 10% of PPG ulcers are misdiagnosed. It also may be useful in refractory cases or when infectious (fungal) or malignant causes are suspected.

Histologic findings are non-specific and tend to reveal a polymorphonuclear infiltration and edema in the epidermis associated with interstitial and perivascular infiltration with lymphocytes, mononuclear cells, and plasma cells in the dermis. The chronic dermal inflammatory process results in thrombosis of blood vessels and secondary necrosis and ulceration of the epidermis and dermis [12-15]. In our case the biopsy was done which confirmed our diagnosis.

Treatment of PPG is empirical and there is no gold standard treatment. Generally based on successful studies of the management of patients with PPG, which does not always respond to IBD treatment [6,9,16,17].

Most patients need systemic treatment to induce remission. Systemic corticosteroids are usually given until clinical improvement occurs. Subsequent doses are gradually reduced to prevent recurrence. Corticosteroids, like prednisolone, 1 to 2 mg per kg per day, are widely used for initial therapy. Rapid improvement has been reported in patients with severe disease given Intravenous corticosteroid therapy (hydrocortisone 100 mg four times daily or methylprednisolone 1 g/day [pulse therapy]) for up to 5 days. Since this treatment may cause fatal side effects in patients with cardiovascular disease on diuretics, patients have to be carefully selected [17-19].

Topical treatment as monotherapy can be applied for localized and mild disease, but it can also be used in combination with systemic treatment for more severe lesions. Topical treatments that have been tried with some success include corticosteroids, tacrolimus, cyclosporine, 5- aminosalicylic acid and dapsone [20].

Immunosuppressive drugs, especially anti-TNFs, represent possible therapeutic alternatives, where their use is well established for intestinal disease and are currently indicated as second- line therapy after failure of corticosteroids [6].

Simultaneous lesions and stoma management is an important adjuvant treatment for successful treatment of PPG, this management includes keeping the stoma clean and maintaining a well- fitting, irritation-free stoma [9]. Zinc oxide paste and silver nitrate baths have been used to improve symptoms [9].

It should be noted that surgery is not indicated for PG. Ulcer excision or stoma revision leads to exacerbation of PG, locally and distantly [9].

In most cases, the prognosis of PPG is good as the lesions disappear after treatment, but may reoccur in the long term. In other less frequent cases, patients may present chronic lesions, requiring long term combination therapy, whose toxicity and efficacy must be monitored [21]. In our patient's case, the lesions were localized, so the treatment used was local care with good cleaning of the peristomial skin, followed by application of vaseline dressings, local corticosteroid therapy, and biotherapy (infliximab) as a maintenance treatment for her disease, which were effective on this lesion, with healing after 2 weeks of treatment leaving an atrophic scar.

## Conclusion

Peristomal PG in IBD is an exceptional localization, as it is probably underdiagnosed. Early diagnosis and treatment is important to avoid disease progression and subsequent patient discomfort. Its management is poorly codified and empirical.

In our case the evolution under treatment was favorable with local corticosteroid therapy and anti-TNF as a maintenance treatment of her disease.

## Bibliography

1. AV Weizman., *et al*. "Pyoderma gangrenosum among patients with inflammatory bowel disease: a descriptive cohort study". *Journal of Cutaneous Medicine and Surgery* 18.5 (2014): 361.

2. Veloso FT., *et al*. "Immune-related systemic manifestations of inflammatory bowel disease. A prospective study of 792 patients". *Journal of Clinical Gastroenterology* 23 (1996): 29-34.

3. Christodoulou DK., *et al*. "Frequency of extraintestinal manifestations in patients with inflammatory bowel disease in Northwest Greece and review of the literature". *Digestive and Liver Disease* 34 (2002): 781-786.

4. Barrie A and Regueiro M. "Biological therapy in the management of extraintestinal manifestations of inflammatory bowel disease". *Inflammatory Bowel Diseases* 13 (2007): 1424-1429.

5. Saigal R., *et al*. "Pyoderma gangrenosum". *Journal of the Association of Physicians of India* 58 (2010): 378-383.

6. Ladan Afifi MS., *et al*. "Diagnosis and management of peristomal pyoderma gangrenosum: A systematic review". *Journal of the American Academy of Dermatology* 78.6 (2018): 1195-1204.

7. Manifestations dermatologiques au cours des maladies inflammatoires chroniques de l'intestin; POST'U (2016).

8. D Agrawal., *et al*. "Pathogenesis and clinical approach to extraintestinal manifestations of inflammatory bowel disease". *Minerva Gastroenterologica e Dietologica* 53 (2007): 233-248.

9. Bruce A Cairns., *et al*. "Peristomal Pyoderma Gangrenosum and Inflammatory Bowel Disease". *The Archives of Surgery* 129 (1994): 769.

10. Ruocco E., *et al*. "Pyoderma gangrenosum: an updated review". *Journal of the European Academy of Dermatology Venereology* 23 (2009): 1008-1017.

11. Ruhl AP., *et al*. "Neutrophilic folliculitis and the spectrum of pyoderma gangrenosum in inflammatory bowel disease". *Digestive Diseases and Sciences* 52 (2007): 18-24.

12. Holt PJ. "The current status of pyoderma gangrenosum". *Clinical and Experimental Dermatology* 4 (1979): 509-516.

13. Powell FC., *et al*. "Pyoderma gangrenosum: a review of 86 patients". *QJM: An International Journal of Medicine* 217 (1985): 173-178.

14. Weenig RH., *et al*. "Skin ulcers misdiagnosed as pyoderma gangrenosum". *The New England Journal of Medicine* 347.18 (2002): 1412-1418.

15. TjandraJJ and Hughes LE. "Parastomal pyoderma gangrenosum in inflammatory bowel disease". *Diseases of the Colon and Rectum* 37 (1994): 938-942.

16. Jourabchi N and Lazarus G. "Pyoderma gangrenosum". In: Kang S, Amagai M, Bruckner AL., *et al*. editors. Fitzpatrick's Dermatology. 9th edition. New York: McGraw- Hill 1 (2019): 605-616.

17. Wollina U. "Pyoderma gangrenosum-a review". *Orphanet Journal of Rare Diseases* 2 (2007): 9.

18. Uwe Wollina. "Review Pyoderma gangrenosum - a review". *Orphanet Journal of Rare Diseases* (2007): 19.

19. Xian-rui Wu and Bo Shen. "Diagnosis and management of parastomal pyoderma gangrenosum". *Gastroenterology Report* 1.1 (2013): 1-8.

20. Ahronowitz I., *et al*. "Etiology and management of pyoderma gangrenosum". *American Journal of Clinical Dermatology* 13 (2012): 191-211.

21. L Antonio Carlos., *et al*. "Case report and brief review, pyoderma gangrenosum: An uncommon Case report and review of the literature". *Index Wounds* 29.9 (2017): E61-E69.

**Volume 10 Issue 4 April 2023**