

Extensive Refractory Perineal Pyoderma Gangrenosum Treated with Infliximab, Fecal Diversion, and Negative-Pressure Wound Therapy

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Keywords

Pyoderma gangrenosum · Infliximab · Fecal diversion ·
Negative-pressure wound therapy

Abstract

Background: Pyoderma gangrenosum (PG) is a rare and difficult-to-diagnose disease that often associates with inflammatory bowel disease. **Case:** We present a case of a 57-year-old female with ulcerative colitis receiving 5-ASA who presented with rapidly progressive ulcers in the right foot and on the inside of the thigh, extending from the left large vaginal lip to the perianal area, compatible with PG. She was initially treated with corticosteroids with no response. After multidisciplinary consultation, it was decided to initiate infliximab 5 mg/kg, and to perform ileostomy for fecal diversion and negative-pressure wound therapy. The patient presented with marked improvement of the lesions, being discharged after 2 months and demonstrating almost complete resolution of the lesions within 4 months. **Conclusion:** Due to the rarity of PG, there is no evidence of the optimal man-

agement. The role of surgery is controversial as PG lesions can demonstrate pathergy and theoretically could worsen with surgical intervention. In this case it was decided based on the extent of the lesions and the experience in other septic/ulcerative perianal conditions.

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Extenso pioderma gangrenoso perineal

Palavras Chave

Pioderma gangrenoso · Infliximab · Ileostomia · Terapia de vácuo com pressão negativa

Resumo

Introdução: O pioderma gangrenoso (PG) é uma doença rara e de difícil diagnóstico, que frequentemente se associa à doença inflamatória intestinal. **Caso:** Apresentamos

um caso de uma mulher de 57 anos de idade com colite ulcerosa sob 5-ASA que recorreu ao serviço de urgência pelo aparecimento de úlceras rapidamente progressivas no pé direito e no interior da coxa, estendendo-se desde o grande lábio vaginal esquerdo até à área perianal, compatível com PG. A doente foi inicialmente tratada com corticosteroides, sem resposta. Após consulta multidisciplinar, optou-se por iniciar o infliximab 5 mg/kg, realizar ileostomia para desvio fecal e iniciar terapia de vácuo com pressão negativa. A doente apresentou melhoria marcada das lesões tendo tido alta após 2 meses e demonstrando resolução quase completa das lesões aos 4 meses. **Conclusão:** Devido à raridade do PG, não há evidências de qual a abordagem ideal. O papel da cirurgia é controverso, pois as lesões do PG podem demonstrar patergia e teoricamente poderiam piorar com a intervenção cirúrgica. Neste caso, a opção cirúrgica foi tomada com base na extensão das lesões e na experiência em outras condições perianais sépticas/ulcerativas.

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Introduction

Pyoderma gangrenosum (PG) is a rare ulcerating skin disease often associated with inflammatory bowel disease (IBD), with an estimated incidence of 3–10 cases per million population per year [1, 2]. The etiology of PG is poorly understood but probably involves an interplay of genetic and environmental factors with loss of innate immune regulation and altered neutrophil chemotaxis [2, 3]. Diagnosis is challenging since this condition can exhibit different clinical presentations (namely in morphology and affected sites), may overlap with other conditions, and is associated with several systemic diseases [1, 4].

Recently a group of international experts defined criteria for its diagnosis – the Delphi criteria – yielding a sensitivity and specificity of 86 and 90%, respectively. They include 1 major criterion (biopsy with neutrophilic infiltrate) and 8 minor criteria (exclusion of infection on histology; pathergy; personal history of IBD or inflammatory arthritis; papule, pustule or vesicle that rapidly ulcerates; peripheral erythema, undetermined borders, and tenderness at the site of ulceration; multiple ulcerations [at least one occurring on an anterior lower leg]; cribriform or wrinkled paper scars at healed ulcer sites; and a decrease in ulcer size after immunosuppressive treatment) [4].

Case Report

We report a case of a 57-year-old female patient receiving 5-ASA who was diagnosed with ulcerative colitis (UC) 8 years ago. She was admitted to the emergency department due to the appearance of a profound ulcer on the inside of the thigh, extending from the left large vaginal lip to the perianal area (Fig. 1a) and a rapidly progressive painful ulcer in the right foot with peripheral erythema and undetermined borders (Fig. 2a). She had two bowel movements/day with blood in the stool, and sigmoidoscopy showed lesions compatible with Mayo 3 UC.

Biopsy of the lesion from the foot was obtained, showing neutrophilic infiltration compatible with PG. Infections were excluded by serology and skin biopsy. The patient performed a pelvic computed tomography scan that showed a perianal collection lateralized to the left with extension to the vulva with fistulization of the large left vulvar lip and the possibility of rectal fistulization.

Based on the diagnosis of PG, the patient was started on corticosteroids (oral prednisolone 1 mg/kg/day followed by endovenous hydrocortisone 400 mg/day) with no significant clinical improvement. Taking into account the extent of the perineal lesions and after multidisciplinary consultation (gastroenterology, dermatology, and surgery) it was decided to initiate infliximab 5 mg/kg and to perform derivation ileostomy and negative-pressure wound therapy (NPWT). There was marked improvement of the lesions, and the patient was discharged after 2 months. At 4 months, there was almost complete resolution of the lesions (Fig. 1b, 2b).

Considering the excellent evolution of the lesion with complete healing and also the healing of the colonic mucosa on follow-up colonoscopy, the patient was proposed for reconstruction of the intestinal transit, which was uneventful.

Discussion

PG algorithm treatment is controversial and remains poorly characterized due to limited knowledge of its pathophysiology combined with its rarity [1, 2, 5].

It is generally based on wound care, analgesia, topical or intralesional therapy with corticosteroids, and systemic therapy with corticosteroids, cyclosporine, or anti-TNF agents. A randomized, placebo-controlled study found that infliximab had a beneficial clinical response in 69% of the patients and a remission rate of 21% at week 6 [6]. A systematic review with 60 cases found that 92% of the patients had response to anti-TNF [5].

The role of surgical treatment in PG is controversial as 25–50% of the PG lesions demonstrate pathergy and theoretically could worsen with surgical intervention [3].

Nevertheless, NPWT has demonstrated efficacy in the treatment of chronic wounds in a variety of circumstances. The rationale of the use of subatmospheric pressure therapy includes increased tissue perfusion, control of the advancing ischemic process, production of a healthy

Fig. 1. a Ulcerated lesion on the inner thigh from the great lip to the perianal area, showing deep muscles and tendons. **b** Completely closed and healed lesion on the inside of the left thigh after 4 months of treatment.



Fig. 2. a Ulcerative lesion on the right foot with peripheral erythema and undetermined borders. **b** Almost completely healed lesion on the right foot after 4 months of treatment.

granular wound bed, reduced bacterial load, and pain relief. Some reports show its safety and efficacy in PG, but only if combined with proper immunosuppression [7–10].

Ileostomy for fecal diversion as in other perianal septic and ulcerative conditions (such as perianal Crohn's disease or Fournier's gangrene) is also controversial but was made to decrease wound contamination and to promote faster wound healing, especially taking into account the possibility of fistulization to the rectum [11].

In this study, we show an extensive perineal PG in a patient with UC that was successfully treated with infliximab, derivation ileostomy, and NPWT.

Statement of Ethics

Informed patient consent was obtained for publication of the case details.

Disclosure Statement

The authors have no conflicts of interest.

Author Contributions

M.S. wrote the manuscript, M.A.C., A.C.S., J.L., A.R., and J.C. reviewed the manuscript and followed the patient.

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