Case Report

Pyoderma gangrenosum: a challenge for the plastic surgeon

Pioderma gangrenoso: um desafio para o cirurgião plástico

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■ **ABSTRACT**

Introduction: Pyoderma gangrenosum (PG) is a chronic and rare autoimmune dermatosis. Its etiology remains poorly understood, being idiopathic in 25 to 50% of cases; in others, it is associated with systemic diseases with autoimmune background and has an incidence of 2 to 3 cases per 1 million per year. In Brazil, the rate is 0.38 cases per 10,000 clinical visits, and women between the second and fifth decades of life are the most affected. The clinical presentation is variable, and the ulcerous form, which appears on a previous scar, is the most prevalent. **Case Report:** A 39-year-old, previously healthy female underwent reduction mammoplasty, and later developed a necrotic ulcer on a vertical left breast scar. Debridement of devitalized tissue was performed, with significant worsening despite antibiotic therapy. The appearance suggested PG. Treatment with oral and topical corticosteroids was then initiated with remission. **Conclusions:** PG represents a diagnostic challenge, and can be confused with surgical site infection.

Keywords: Pyoderma gangrenosum; Mammoplasty; Corticosteroids; Reconstructive surgical procedures; Immunotherapy.

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■ **RESUMO**

Introdução: O pioderma gangrenoso (PG) corresponde a uma dermatose autoimune crônica e rara. Sua base etiológica ainda permanece pouco conhecida, sendo idiopático em 25 a 50% dos casos, nos demais está associado com doenças sistêmicas de fundo autoimune, tem uma incidência de 2 a 3 casos em 1 milhão de habitantes por ano. No Brasil, este índice é de 0,38 casos por 10.000 atendimentos, as mais acometidas são as mulheres entre a segunda e quinta década de vida. O quadro clínico é variável, sendo que a forma ulcerosa, que surge sobre uma cicatriz prévia, é a mais prevalente. **Relato de Caso:** Paciente do sexo feminino, 39 anos de idade, previamente hígida, foi submetida à mamoplastia redutora, evoluiu com úlcera necrótica em cicatriz vertical de mama esquerda. Realizado desbridamento de tecidos desvitalizados, prescrita antibioticoterapia, apresentando piora importante da lesão, sendo considerada a hipótese de PG. Iniciado tratamento com corticoterapia oral e tópica com remissão do quadro. **Conclusões:** O PG representa um desafio no diagnóstico e, geralmente, demonstra a dificuldade diagnóstica, podendo ser confundido com infecção do sítio cirúrgico.

Descritores: Pioderma gangrenoso; Mamoplastia; Corticosteroides; Procedimentos cirúrgicos reconstrutivos; Imunoterapia.

INTRODUCTION

Pyoderma gangrenosum (PG) is a chronic and rare autoimmune dermatosis, first described by Brunsting and O Leary in 1930, highlighting the absence of an infectious nature. Histopathologically, it is characterized by a nonspecific, noninfectious, non-neoplastic dermal neutrophilic infiltrate, without evidence of primary vasculitis[1].

The etiological basis remains little understood, being idiopathic in 25 to 50% of cases. Other cases are associated with systemic autoimmune diseases, especially inflammatory bowel disease and mainly ulcerative colitis, but also arthritis, IgA gammopathy, and others[2,3]. It may also appear as a paraneoplastic manifestation or after the use of certain medications (propylthiouracil and isotretinoin in particular) and illicit substances such as cocaine[4].

PG has an incidence of 2 to 3 cases per 1 million per year[4,5]. National data from a retrospective analysis indicated that in Brazil, this rate is 0.38 cases per 10,000 clinical visits[5], with women between the second and fifth decades of life being most affected[4].

The clinical presentation is variable, and a single or multiple ulcerous form, which appears on a prior scar, is the most prevalent[6]. The ulcers are well circumscribed, and have a violaceous halo and necrotic-hemorrhagic center, with characteristic purulent and accelerated centrifugal growth, ending with accelerated formation of granulation tissue[7]. In addition to the ulcerated form, PG has pustular and vegetative forms, which are less prevalent and have fewer postoperative complications[8].

Histopathology and immunohistochemistry in PG are nonspecific, and no serological markers are available for laboratory diagnosis; thus, diagnosis is clinical by default[9,10].

Knowledge of the pattern of the cutaneous lesion is important, because diagnosis of post-surgical PG can be delayed. More commonly suspected diagnoses such as wound dehiscence and infection result in unnecessary debridement that tends to worsen the clinical presentation, since the pattern of the PG lesion is related to the phenomenon of pathergy in up to 50% of cases, with minor trauma triggering new lesions[6,11].

The plastic surgeon should include the diagnosis of PG in the differential diagnosis, since the knowledge of cutaneous lesions, predisposing factors, and surgical risk factors enables avoidance of exacerbation.

CASE REPORT

A 39-year-old, previously healthy Caucasian patient, with no surgical or obstetrical history, underwent reduction mammoplasty (Figure 1), and was discharged after 24 hours without complaints and in good general condition.

The patient received antibiotic prophylaxis with cefazolin 1 g every 6 h during hospitalization, followed by cephalexin 500 mg every 6 h, until the postoperative

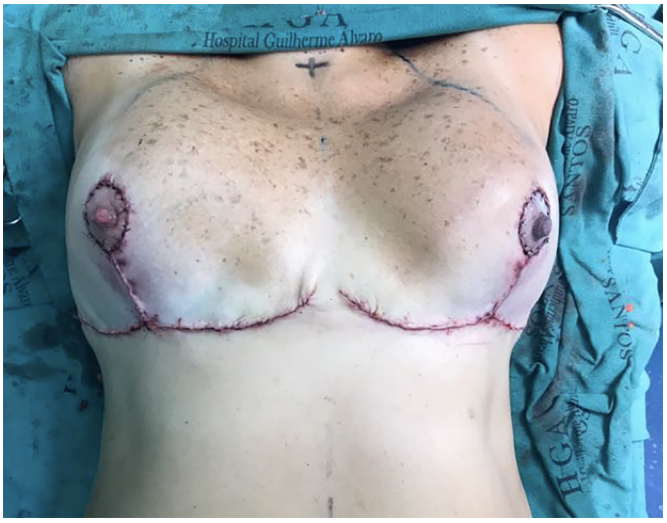

Figure 1. Immediate postoperative appearance.

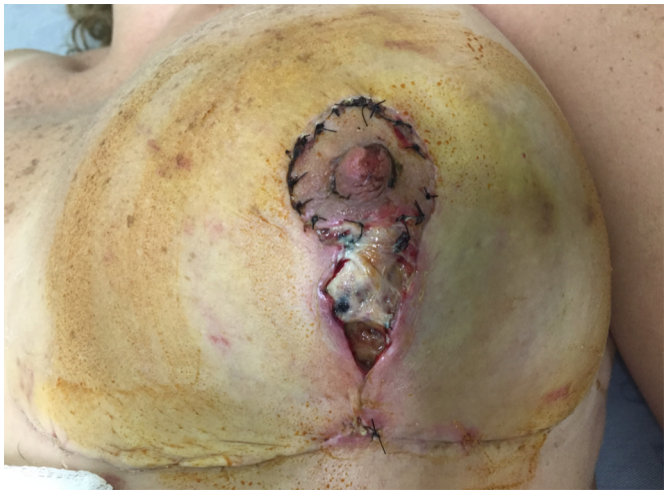

Figure 2. Postoperative day 10 – evolving ulcerated lesion.

day (POD) 7. She was reassessed 72 h after the surgical procedure, still without complaints. The surgical wound had a good appearance, and a micropore dressing was in place.

On 15 POD 15, during routine follow-up, the surgical wound presented a necrotic ulcer in a vertical scar on the left breast, with drainage of moderate seropurulent secretions (Figure 2). Debridement of devitalized tissue was performed, together with antibiotic therapy with ciprofloxacin 500 mg every 12 h and local dressings with collagenase ointment, in addition to micropore dressings. The wound initially appeared improved. On POD 17, the patient was reevaluated for significant worsening, with centripetal progression, affecting segments of the scar in the inframammary groove. The patient denied systemic symptoms.

Due to the surprising change, PG was considered. Consequently, a systematic review was performed in the literature regarding the therapeutic management of PG (Table 1).

First-line treatment was initiated with oral and topical corticosteroids. Prednisone was administered at 60 mg/day for 7 days, 40 mg/day for 7 days, 20 mg/day for 7 days, and finally 10 mg/day for 4 days, ending with 10 mg/day on alternate days, for a total of 28 days (Figure 3).

The topical treatment comprised daily fludroxycortide 0.125 mg/g (Drenison®), maintained from the beginning of oral therapy until the consolidation of the scar, which occurred at the end of the 2nd postoperative month, approximately 20 days after the end of oral corticosteroid therapy (Figure 4). The patient had a favorable course, with complete closure of the lesions after treatment for 1 month. Figure 5 shows the appearance in the 3rd postoperative month.

DISCUSSION

PG is a devastating complication both for the patient and the plastic surgeon, leading to questions about the technical quality of the surgical procedure[12]. The absence of a supplemental exam that confirms the diagnosis of PG, combined with nonspecific findings on histopathology, requires clinical knowledge of this disease to enable diagnosis[1,2,4].

The objective of treatment is to limit tissue destruction, promote healing, and obtain a good esthetic result. Debridement and skin grafts are contraindicated[3-5].

First-line treatment with systemic corticosteroids is the most effective option for PG. Immunosuppressive doses are necessary in the majority of cases, with approximately 100-200 mg/day of prednisolone or 60-80 mg/day of prednisone [4,6,7].

Table 1. Clinical and therapeutic reports on PG, searched in databases of the last 10 years.

Database	Search strategy	Results	Clinical Cases
PubMed	(pyoderma gangrenosum) AND (breast)	81	66
Lilacs	(pyoderma gangrenosum) AND (breast)	3	3
SciELO	(pyoderma gangrenosum) AND (breast)	2	2
Total		86	71

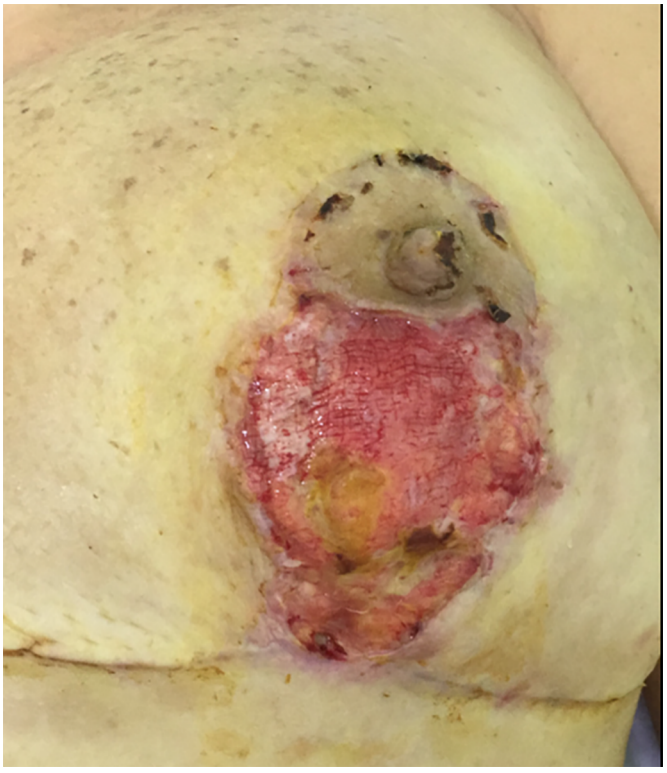

Figure 3. Postoperative day 21 - 4 days after initiation of oral and topical corticosteroids.

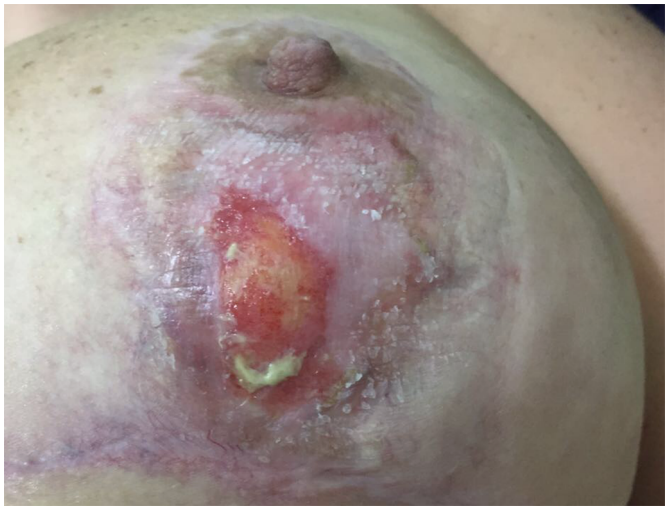

Figure 4. Postoperative day 30 - 13 days after initiation of oral and topical corticosteroids.

Alternatively, cyclosporine, at doses of 6-10 mg/kg/day, can produce significant improvement, with healing in 1 to 3 months, and is indicated for a minority of patients who do not respond to steroid therapy[7,8].

TNF-alpha inhibitors, such as infliximab, show good results in some patients[10,12].

Hyperbaric oxygen therapy may be indicated for patients who cannot tolerate or do not respond to high doses of systemic corticosteroids; however, the therapeutic result is less effective, as demonstrated in some case series[11]. Topical therapy is indicated to complement systemic therapy; corticosteroids alone are the drugs of choice in selected cases of lesser severity[6,7,11].

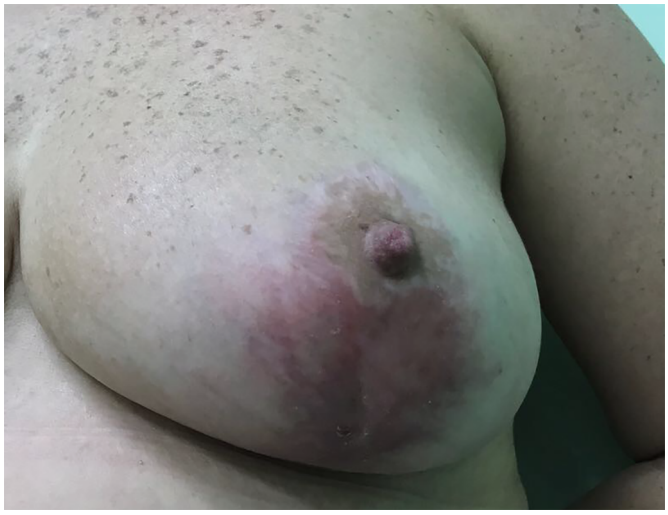

Figure 5. 3 months postoperatively - healed lesion.

Antibiotic therapy is not supported for PG cases, as demonstrated in all series studied; therefore, there are no clinical benefits from the use of antimicrobial agents in these patients[7,9].

With regard to future plastic surgery, the patient should be advised about the likelihood of recurrence of PG, and this point should be explained in an Informed Consent Form to be signed by the patient. Long-term follow-up with a rheumatologist is recommended, as other autoimmune disorders may be found[7,9,12].

COLLABORATIONS

FFGO	Conception and design of the study; completion of surgeries and/or experiments; writing the manuscript or critical review of its contents.
MF	Completion of surgeries and/or experiments.
AMNG	Completion of surgeries and/or experiments.
OSF	Completion of surgeries and/or experiments.
MRM	Completion of surgeries and/or experiments.
EGC	Final approval of the manuscript; conception and design of the study; completion of surgeries and/or experiments.
OS	Final approval of the manuscript.

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