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An extensive pyoderma gangrenosum mimicking necrotizing fasciitis: An unusual case report

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ABSTRACT

INTRODUCTION: Pyoderma Gangrenosum (PG) is a rare, benign and inflammatory disease characterized by ulcerative skin lesions. We report the successful management of an unusual case of PG following a caesarean section, with extensive cutaneous skin involvement and mimicking necrotizing fasciitis.
PRESENTATION OF CASE: A 36-year-old woman was admitted with extensive surgical site inflammation after a caesarean section. Despite antibiotic treatment and wound care, the clinical course deteriorated rapidly. Wound debridement following negative pressure closure was performed due to an immediate increase in skin necrosis. A diagnosis of PG was reached based on the absence of a positive wound culture, resistance to wound debridement and the histopathological results. A course of high-dose corticosteroids was started, and a successful clinical course was finally achieved. The patient is now in the 14th month of remission, with no recurrence.
DISCUSSION: PG is often reported after bowel surgery, especially after complicated stoma or diverticulitis, breast surgery and occasionally after C-sections. The diagnosis of pyoderma gangrenosum may be challenging because of a wide variety of macroscopic features and its pronounced similarity to necrotizing fasciitis. Treatment with systemic corticosteroids is the most common management option, while surgical treatment is extremely controversial.
CONCLUSION: An extensive PG following surgery can mimic necrotizing fasciitis. An interdisciplinary treatment approach provides early diagnosis and effective treatment resulting in less morbidity.
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1. Introduction

Pyoderma gangrenosum (PG) is a rare inflammatory, non-neoplastic and ulcerative skin condition [1]. Although the pathogenesis is unclear, at least 17%–74% of PG patients are found to have an underlying systemic disease [1–3]. The most common associations are with inflammatory bowel diseases, rheumatoid arthritis, hematological malignancies and monoclonal immunoglobin A gammopathy, but many other comorbidities may be seen [4]. The annual rate for PG is approximately 3-10/1000000 in a year, and commonly occurs between the ages of 20–50 [4,5].

PG lesions typically develop at sites of injury caused by trauma or surgery, also known as the pathergy phenomena [6]. PG is sometimes encountered after surgery, especially after complicated stoma or diverticulitis [4], breast surgery [6] and occasionally after C-sections [7]. Necrotizing fasciitis and other neutrophilic dermatomes are important alternatives to consider in differential diagnosis [8,9]. Medical treatment methods including systemic corticosteroids are effective for PG treatment and early diagnosis and treatment can have a significant impact on morbidity [9–11].

We report the successful management of a case of PG with extensive cutaneous involvement of the skin, following a caesarean section.

2. Case report

A 36-year-old woman underwent an uneventful C-section at normal term. The pregnancy was uncomplicated, and her medical history presented no co-morbidities such as diabetes mellitus (DM), hypertension (HT) or coagulopathy disorders, and she was a non-smoker. In addition, the patient didn't have drug history, including any psychosocial history and relevant genetic information. The body mass index of the patient was 25 kg/m2. After seven days postpartum, she was admitted to the emergency department with complaints of chills, fever and surgical site infection. Physical examination revealed swelling, erythema and fluid discharge at the pfannenstiel incision. The patient had constipation although there was release of gas, and there were no signs of

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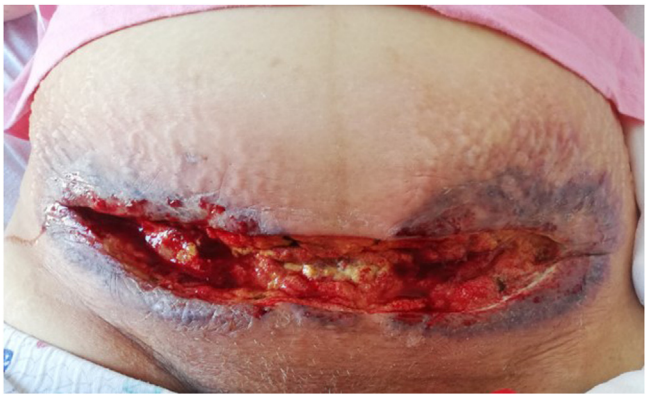

Fig. 1. Pyoderma gangrenosum in the C-section incision (7th day postpartum).

ileus or acute abdomen. The laboratory results indicated a white blood cell level of 8360/mm [3], hemoglobin of 7.8 (normal range: 12.2–18.1), hematocrit at 24.6% (normal range: 37.2%–53.1%), and platelet levels at 166 K/Ul (normal range:142–424). Moreover, her C-reactive protein (CRP) was 261 mg/dl (normal range: 0–5). An abdominal ultrasonography showed no intraperitoneal collection or abscess. A contrast-enhanced computed tomography detected a fluid collection above the C-section scar under the umbilicus, 25 × 15 mm in diameter and including air density. Furthermore, CT showed minimal free fluid in the pelvis, and increased density in the anterior abdominal wall, compatible with widespread edema, in adjacent fat planes close to the operation site. There was no intraperitoneal abscess. The patient was hospitalized to the department of obstetrics and gynecology, and, following wound culture results, prophylactic antibiotic (ceftriaxone 1 g. IV, twice a day, and metronidazole 500 mg. IV, at eight hourly intervals) was administered. The general surgery and dermatology consultation were performed for differential diagnosis. Hematocrit levels reached the normal range with erythroid suspension. The patient underwent wound debridement followed by vacuum assisted closure (Figs. 1 and 2). Unfortunately, after one day, the surgical site infection had expanded into the near umbilicus and bilateral lumber region. The borders of this surgical site infection were marked with a pencil to track its development, and the infection was seen to have increased after a few hours (Fig. 3). There was no growth in the wound culture; however, meropenem and 'tekosit' (Teicoplaninum) were given to counter the infection due to increased CRP, white blood cell count and uncontrolled fever. A skin biopsy was taken to determine the probability of pyoderma gangrenosum or necrotizing fasciitis and because of the lack of a positive culture, its resistance to wound debridement, and histopathological features, a diagnosis of "Pyoderma gangrenosum" was returned (Fig. 4). High dose systemic corticosteroids (prednisolone 1 mg/kg, IV per day) were given, resulting in rapid clinical improvement. This corticosteroid therapy was tapered off slowly and stopped within one month (Fig. 5). The patient is now 14 months postoperative with no recurrence (Fig. 6). The first intervention and follow up were managed by senior surgeons and experience dermatologist with interdisciplinary approach. Moreover, the patient presented a positive attitude toward her treatment in the surgical intervention, medical treatment and at follow-up. The present case report study was edited by SCARE 2020 guidelines [12].

3. Discussion

Pyoderma gangrenosum is a rare inflammatory disease which is painful, progresses rapidly, and is characterized by large ulcers in the neutrophilic dermatoses [1]. PG was first described by Burnsting et al. in 1930 [2] as being in the spectrum of inflammatory

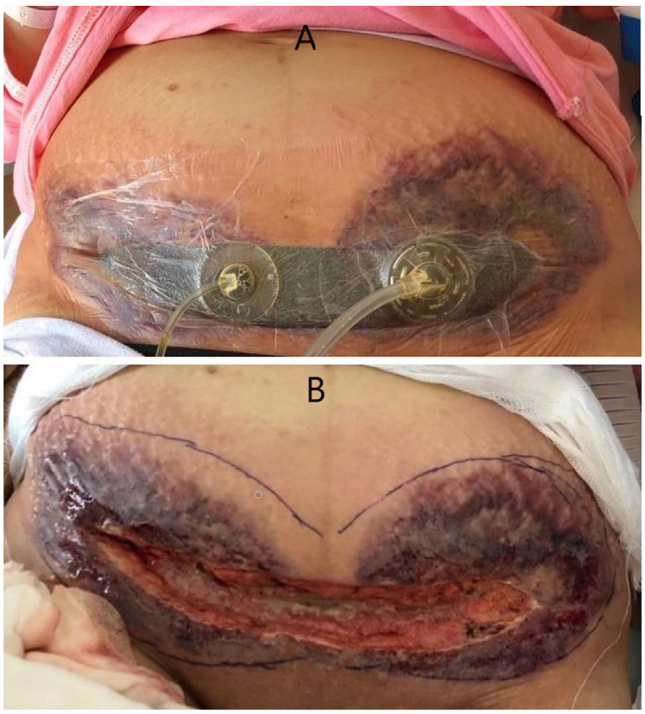

Fig. 2. Violaceous wound borders have spread, progressively expanding the ulcerative area (8th day postpartum) (A), and tretmanet of vacuum assisted closure (B).

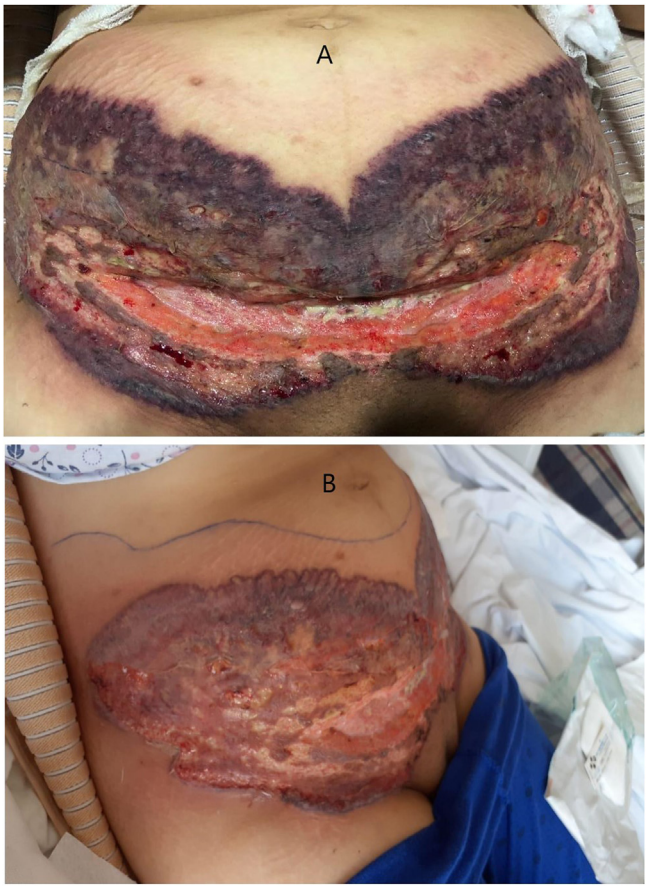

Fig. 3. Extensive painful ulcerative open wound with erythematous plaques and violaceous border involving the whole lower abdomen (9th day postpartum) (A) and late of second weeks of postoperative (B).

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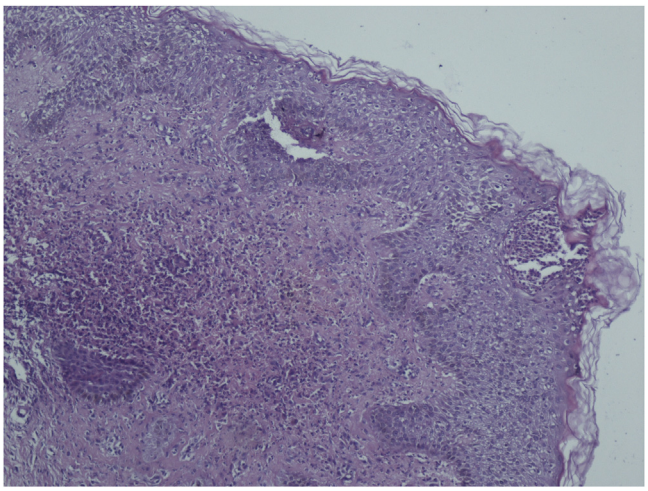

Fig. 4. The abscess formation: a neutrophilic inflammation within subcuticular and deep dermis layers (Hematoxylin&Eosin X100).

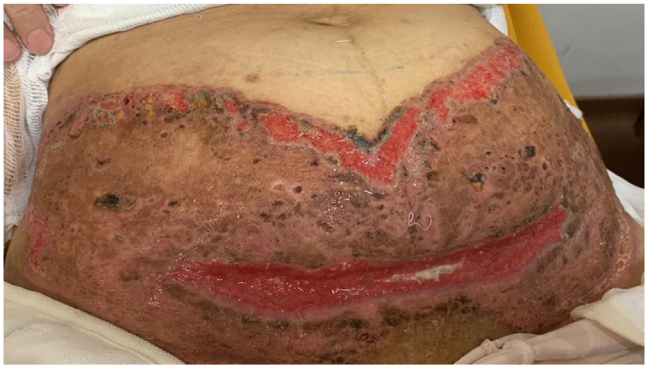

Fig. 5. Recovery of the wound after corticosteroid treatment (1st month).

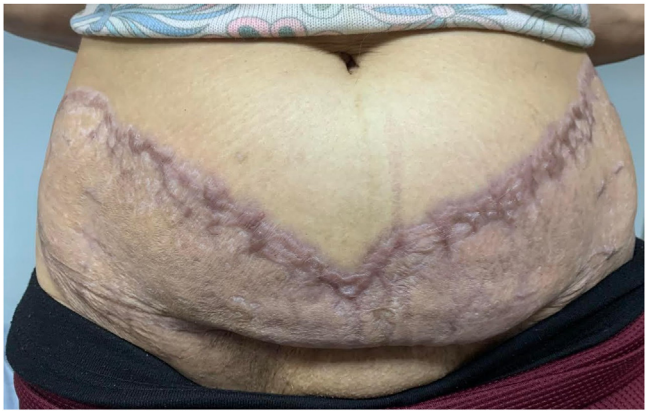

Fig. 6. The complete healing of the wound (A, 10th month and B, 1st year postpartum).

neutrophilic diseases. As in other reactive neutrophilic dermatoses, females are more frequently affected by PG [3,4]. The present case is a 36-year-old woman without major co-morbidity.

The etiology is thought to be a manifestation of immunological response to injury leading to neutrophil dysfunction [4,5]. PG is most commonly seen as a single lesion in the lower extremities, on the anterior aspect of the tibia. Atypical forms are more superficially localized, usually on the face, head-neck, breast, arm, hand, and especially on the peristomal skin [1,5]. Von den Driesch et al. [6] reported that 73% of PG cases were accompanied by vasculitis of which 68% was leukocytoclastic and 32% lymphocytic. In over half of PG patients, there is an association with systemic diseases, particularly inflammatory bowel disease, rheumatologic and hematological diseases, monoclonal gammopathies, suppurative hidradenitis and immunosuppression or malignancies [5,6]. In present case report study, our patient had not the systemic disease.

PG can be difficult to diagnose in a variety of clinical situations. It is usually established by excluding other clinical features, and from the histopathological findings and similar cutaneous pathologies of the patient. Laboratory findings may also reflect symptoms of the underlying disease. Necrotic-based ulcers, dark red or purple fresh hemorrhagic pustules are clinical ulcer features. Pathergy tests are positive in 30% of patients. There are several different forms of PG such as ulcerative, bullous, vegetative and pustulous and the clinical course and localization of the disease may vary with each form of PG. In the classical ulcerative form, lesions tend to occur after minor trauma and generally on the lower extremities and trunk whereas in its pustular form, common in patients with inflammatory bowel disease, multiple eruptive, inflammatory cutaneous pustules develop. Although in its ulcerative form PG may begin with a pustule, it can rapidly turn into a large ulceration [3,7]. The clinical and histopathological findings of our case were consistent with ulcerative PG.

The main treatment for PG is medical with systemic corticosteroid being the primary choice. In patients with less severe PG, topical and intralesional corticosteroids can be used effectively. Clungston et al. [8] reported using intradermal triamcinolone successfully for the treatment of PG, without the need for systemic corticosteroids. On the other hand, a patient whose disease rapidly progresses or is resistant to local treatment may be prescribed immunosuppressive drugs, intravenous immunoglobin or biological agents [3,6,9–11]. High-dose prednisolone (40–120 mg or 1 mg/kg per day) is suggested as a primary treatment option to achieve remission of disease while combination treatment, adding cyclosporine (2–6 mg/kg per day), may be preferred [3,9]. Reichrath et al. reported the combined use of prednisolone (0.3–1 mg/kg per day) and cyclosporin (5 mg/kg per day) as first line treatment for PG.

After surgery problems such as surgical site infection, wound dehiscence, cellulitis and necrotizing fasciitis may occur [9,10]. While cellulitis generally has mild symptoms such as erythema, and pain in the extremities or flank, localized necrotizing fasciitis frequently involves severe symptoms including a sudden pain in the scrotum or surgical site, together with pallor and fever [10]. These clinical entities can be diagnosed easily in surgical departments. However, the diagnosis of pyoderma gangrenosum, which is seen less often in surgery clinics, may be harder because of its indistinguishable macroscopic features. PG is often reported after bowel surgery, especially after complicated stoma or diverticulitis [4], breast surgery [6] and infrequently after C-sections [7], but can mimic or be misdiagnosed as a surgical site infection, wound dehiscence, or necrotizing fasciitis. Unfortunately, the patient may unwittingly undergo surgery because of misdiagnosis or confusion with an infectious disease, in which case correct diagnosis and treatment may be unavoidably delayed.

Moreover, using surgery to treat PG is considered an extremely controversial approach. As most authors consider PG to be a “pathergy” reaction and expect trauma to exacerbate the lesions [6,11–14], an aggressive surgical treatment approach is not recommended. Wong et al. [15] misdiagnosed 29 of their 35 PG patients as having an infection and subsequently treated them with antibiotics; 13 of these patients also underwent debridement of their lesions. Cabalag et al. [16] suggested using a combination of surgery, hyperbaric and immunosuppressive therapy to resolve both the disease and disease-associated co-morbidities, as well as alleviating the consequent adverse effects of long-

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term immunosuppressant therapy. Ultimately, an interdisciplinary approach including surgery, dermatology and pathology is recommended for effective treatment. In the present case report study, the patient was originally assessed as having localized necrotizing fasciitis, however, both general surgery and dermatology consultations were requested for the differential diagnosis of PG and necrotizing fasciitis due to the enlargement of the erythema area at the lesion borders in the early postoperative period after debridement. The diagnosis was revised to PG when clinical response was observed within 1 day after steroid therapy initiated after punch biopsy was taken. As a result, the patient was discharged successfully after treatment with systemic corticosteroids.

4. Conclusion

Pyoderma gangrenosum is a rare inflammatory disease that is difficult to diagnose, and for which the use of surgical treatment is not recommended in clinical practice. The clinical features, laboratory findings and progressive course of PG can mimic necrotizing fasciitis. Severe pyoderma gangrenosum is an interdisciplinary issue that concerns both internal and surgical medical sciences; therefore, early diagnosis and successful treatment depend on input from both these fields.

4.1. Learning points

- Pyoderma gangrenosum is a rare, benign ulcerative disease of the skin.
- PG can mimic necrotizing fasciitis
- The correct diagnosis of Pyoderma gangrenosum is essential for treatment
- In case of suspected necrotizing fasciitis, it can be a reasonable approach to take a dermo-epidermal biopsy to exclude PG during debridement and to stimulate the pathology for early diagnosis.
- The primary treatment for PG is systemic corticosteroids

Declaration of Competing Interest

There is not conflict of interest.

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Ethical approval

This is case report.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Mehmet Aziret → Study design and writer
Şeyma Kara → Data collection
Mahizer Yaldız → Data Analysis
Nur Köse → Data collection
Feyza Aşkınuzunoğlu → Interpretation
Arif Serhan Cevrioğlu → Study concept

Registration of research studies

Not applicable.

Guarantor

Mehmet Aziret.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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