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Pyoderma Gangrenosum of the breast: A case report study



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ABSTRACT

INTRODUCTION: Pyoderma gangrenosum (PG) of the breast is a rare and rapidly spreading disease, which usually co-exists with severe underlying systemic conditions. PG often presents secondary to breast surgery with skin lesions and signs of infection, even though it is a non-infectious, necrotizing dermatological entity.

PRESENTATION OF CASE: We present a case of de novo unilateral breast PG in 37-year-old woman, with a clear medical history whatsoever. The patient was treated with corticosteroids and, in a two-month follow up, presents with nearly no signs of PG.

DISCUSSION: PG of the breast presents with atypical clinical signs and is characterized by an exclusion-based diagnosis. It often mimics inflammation but is resistant to antibiotics.

CONCLUSION: The optimal treatment for PG is systemic use of corticosteroids and surgical debridement of the necrotic tissue, while the timely onset of the therapeutic approach is of outmost importance.

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1. Introduction

Pyoderma gangrenosum (PG) is a rare, non-infectious, rapidly spreading cutaneous necrotizing dermatological disease. It was first described in 1908 by the French dermatologist Brocq with the term “geometric phagedenism”, considering it as a bacterial infectious disease [1]. Although the etiology and pathophysiology of PG are still not enlightened, nowadays it is categorized into the spectrum of neutrophilic dermatoses caused by genetic predisposition and modified immune/inflammation response [2]. Annually, 3–10 in a million are reported as newly diagnosed cases, with 50–70% of these patients suffering from severe underlying systemic diseases; autoimmune (most frequently inflammatory bowel disease and rheumatoid arthritis), as well as hematologic disorders (leukemia and lymphoma) [3].

PG manifests as an afflictive inflammatory papule, nodule or pustule, expeditiously evolving into an ulcerative lesion with undermined borders. The ulcers are usually located at the lower extremities and the pretibia area, whilst infrequently it presents in other sites (trunk, head, neck hands, peristomal skin) or extra-cutaneous tissues (lungs, liver, bones) [4]. Breast PG is uncommon, with only 43 reported cases in the literature, 70% of which emerged after breast surgical intervention. It is important to include PG in the differential diagnosis of breast ulceration, as early diagnosis can

reduce morbidity and needless therapies. We present a case report [5] of de novo unilateral breast PG in a 37-year-old woman, who had no history of associated systemic disorder or surgical intervention.

2. Case presentation

A 37 year old Caucasian woman G1P1 presented in our outpatients' department with painful, left breast ulceration and fever of 38 °C. The patient was non-smoker and had a free medical history. Three days earlier, the patient had noticed an edematous pustule which progressed into a small ulcer of 1 cm diameter at the lower outer quadrant of the left breast. Oral antibiotics were prescribed (cefuroxim) and another visit after three days was recommended. The patient reappeared after 7 days with persistent fever in the afternoons (38,5 °C), excruciating pain and a rapidly developing cutaneous ulcer of dimensions 10 × 5 cm, occupying the inferior half of the left breast but relatively sparing part of the areola and nipple (Fig. 1). She was immediately admitted to our hospital for further investigation, diagnosis and treatment. Her vital signs were normal (temp: 36,6 °C, BP: 112/69 mm Hg, HR: 90 bpm), but the blood tests revealed signs of infection with leukocytosis – neutrophilia (23.000) and elevated CRP levels (144 µg/ml).

During her hospitalization, despite the blind double antibiotic intravenous treatment (ampicillin-sulbactam and metronidazole), the patient continued with afternoon fevers while the breast ulcer deteriorated by 1–2 cm per day. The microbiological cultures and the blood cultures for aerobic and anaerobic bacteria came back negative, so we proceeded to perform surgical debridement and

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Fig. 1. Elcotic, inflammatory lesions upon presentation.



Fig. 2. One month after treatment during follow up.

obtain tissue biopsies. The fascia was not affected macroscopically, which was established in the surgical specimen. As the lesion did not respond to our treatment, at the same time we initiated corticosteroid administration, since PG was part of our differential diagnosis. The pathology report confirmed our hypothesis, revealing skin ulceration covered with purulent exudate, diffuse and perivascular lymphoplasmacytic infiltration at the ulcer borders with necrotizing vasculitis, as in PG (Fig. 3). Following treatment with intravenous pulse corticosteroids (1 g methylprednisolone per day for five days) because of the very aggressive and painful manifestation of the disease, we recorded a rapid response (Fig. 2). Within a two-month follow up period, we have recorded an optimum healing process and the patient has, at the moment, nearly no signs of PG (Fig. 4).

3. Discussion

Pyoderma gangrenosum (PG) is an uncommon neutrophilic dermatosis that presents as an inflammatory and ulcerative dis-



Fig. 3. Two months after treatment. Significant improvement.

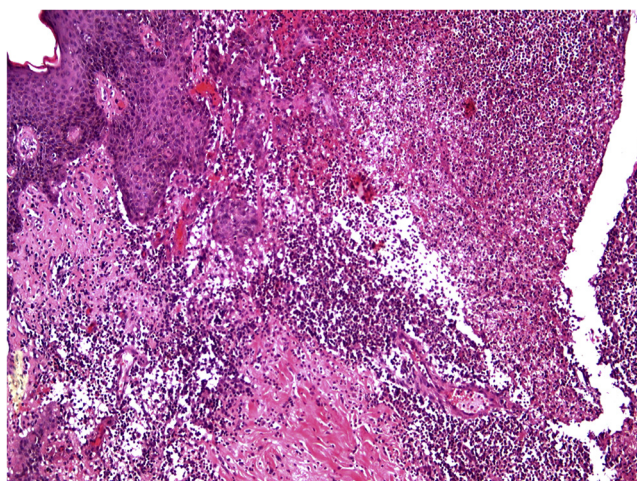


Fig. 4. No specific ulceration with abscess formation in deep dermis and subcutis.

order of the skin, characterized by atypical clinical signs, similar to infectious diseases. Since there are no diagnostic laboratory or histological findings, PG is a diagnosis of exclusion, based on ulcerative characteristics, resistance to antibiotic and surgical therapy and improvement after steroid treatment [6]. Even though histopathological examination is useful in order to distinct PG from other causes of ulceration, ulcerative disease combined with dermal neutrophilic infiltration, makes the diagnosis often difficult and sometimes, impossible [7].

In the case presented, our patient had a minor ulcer that was originally treated with a course of antibiotics. One week later she was reevaluated, only to discover that the ulcer had evolved in size had affected almost the whole of the breast. It is notable that our patient had a free medical history and no surgery to either breast whatsoever.

The later is of great importance to the diagnosis, since the onset of lesions in cases with a previous surgery is rapid, thus PG could be mistaken for wound infection [8]. In our case, the edges of the ulcer were sharp and undermined, while there was no indication of inflammation. There was no bullae present, no thrombosis of small blood vessels in histology, no compartment syndrome and no myonecrosis requiring fasciotomy. We also recorded absence of gram-positive cocci. This helped us in the differential diagnosis from necrotizing fasciitis, which is an infection of the deeper tissues that results in progressive destruction of the muscle fas-

cia and overlying subcutaneous fat; muscle tissue is frequently spared because of its generous blood supply. Post-surgical PG affects the surgical incision site in the immediate post-operative period. Patients usually present with fever, malaise and areas of wound dehiscence, progressing to painful ulcers with violaceous, undermined borders [9]. Our patient presented with the same aforementioned symptoms, but with no history of prior surgery.

Interestingly, appearance of PG has a predisposition during pregnancy and post partum. A possible explanation could be based upon the alterations to the immune system of pregnant women, since pregnancy is an immunosuppressive condition [10]. However, our patient was neither pregnant nor lactating upon the presentation of PG.

The association of PG to systemic diseases such as rheumatoid arthritis, hepatologic malignancy, IgA monoclonal gammopathy and inflammatory bowel disease is well established. However, it seems that there could be a connection between new-onset of PG and a previously diagnosed solid organ malignancy, while recurrent PG could also be of the same origin [11]. Nevertheless, our patient did not have a known history of cancer, nor was diagnosed during her investigational and follow up period.

Regarding the therapeutic approach, the severity of PG influences the choice of initial therapy. Although local interventions successfully induce healing in cases with mild disease, patients with more extensive PG require systemic therapy. Glucocorticoids are the most common systemic drugs used, since a rapid response is often observed, while systemic cyclosporine works as an alternative treatment to patients who cannot tolerate systemic glucocorticoid therapy [12,13]. In our case, the initial treatment with antibiotics, was based on signs of inflammation and probable infection. Since the tissue and exudation cultures proved negative, a therapeutic approach with corticosteroids was initiated which, in conjunction to surgical debridement, provided with optimum results.

4. Conclusions

Pyoderma gangrenosum of the breast is a rare entity and is often correlated to previous surgical treatment. It usually coexists with systemic inflammatory diseases and hematologic disorders. A careful clinical examination, macroscopic evaluation and laboratory investigation is of utmost importance, while treatment with oral or systemic glucocorticosteroids seems to be the optimum first-line therapeutic approach. Although seldom encountered, its clinical existence should be recalled when rapidly progressing ulcerative lesions are observed. Severe morbidity can be substantially reduced with timely onset of appropriate treatment, as discussed.

Conflicts of interest

The authors declare no conflict of interest.

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Ethical approval

This is not a research study.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Authors contribution

Spyridon Marinopoulos: Editor
Charalampos Theofanakis: Writer
Theoni Zacharouli: Writer
Maria Sotiropoulou: Histopathology report
Constantine Dimitrakakis: Reviewer

Registration of research studies

This is not a research study.

Guarantor

Spyridon Marinopoulos.

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