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CASE REPORT

Multifocal pyoderma gangrenosum involving breast and oral mucosa: a diagnostic enigma

Pioderma gangrenoso multifocal com envolvimento da mama e da mucosa oral: um enigma diagnóstico

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Abstract

Pyoderma gangrenosum (PG) is a rare neutrophilic dermatosis typically affecting the lower limbs, often associated with systemic diseases such as inflammatory bowel disease (IBD). We report a rare case of a 36-year-old female presenting with painful ulcers over atypical sites, including the left breast and tongue, alongside the buttocks, groin, thighs, and abdomen. Lesions evolved from pustules to necrotic ulcers, accompanied by fever, bloody diarrhea, and arthralgia. Histopathology revealed dense neutrophilic infiltrates; colonoscopy showed terminal ileal ulcerations consistent with IBD. Infectious causes were excluded. The patient responded well to systemic corticosteroids and cyclosporine over 3 months. This case underscores the importance of recognizing PG in uncommon sites such as the oral mucosa and breast, which may mimic other conditions. Early diagnosis and immunosuppressive treatment are vital for optimal outcomes, especially in PG associated with systemic disease.

Keywords: Pyoderma gangrenosum. Mucosal involvement. Inflammatory bowel disease.

Resumo

O pioderma gangrenoso (PG) é uma dermatos e neutrofílica rara que afeta tipicamente os membros inferiores, frequentemente associada a doenças sistémicas como a doença inflamatória intestinal (DII). Relatamos um caso raro de uma mulher de 36 anos que apresentou úlceras dolorosas em locais atípicos, incluindo a mama esquerda e a língua, além das nádegas, virilha, coxas e abdómen. As lesões evoluíram de pústulas para úlceras necróticas, acompanhadas de febre, diarreia com sangue e artralgia. A histopatologia revelou infiltrados neutrofílicos densos; a colonoscopia mostrou ulcerações no íleo terminal compatíveis com DII. As causas infecciosas foram excluídas. A paciente respondeu bem ao tratamento com corticosteroides sistémicos e ciclosporina ao longo de três meses. Este caso destaca a importância de reconhecer o PG em locais incomuns, como a mucosa oral e a mama, que podem imitar outras condições. O diagnóstico precoce e o tratamento imunossupressor são fundamentais para resultados ótimos, especialmente no PG associado a doença sistémica.

Palavras-chave: Pioderma gangrenoso. Envolvimento mucoso. Doença inflamatória intestinal.

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Introduction

Pyoderma gangrenosum (PG) is a rare, ulcerative, inflammatory skin disorder categorized within the spectrum of neutrophilic dermatoses. It is estimated to occur at an annual incidence of 3-10 cases/million individuals, underscoring its clinical infrequency and its diagnostic challenges[1]. Classically, PG manifests as rapidly progressive, extremely painful ulcers bordered by characteristic violaceous, undermined edges and accompanied by dense neutrophilic infiltration on histopathology. The most commonly affected anatomical sites are the lower extremities, particularly the pretibial areas. PG is frequently associated with systemic conditions such as inflammatory bowel disease (IBD), autoimmune disorders, and hematologic malignancies, with up to 50% of patients demonstrating such comorbidities[2].

Despite decades of study, the exact pathogenesis of PG remains incompletely understood. Current evidence points toward an interplay of immune dysregulation, genetic predisposition, and neutrophil dysfunction. Pathergy – the phenomenon in which minor trauma incites exaggerated inflammatory responses is well recognized in PG and often complicates both diagnosis and management. Due to the absence of definitive laboratory or histological markers, PG continues to be a diagnosis of exclusion, requiring careful elimination of infectious, vascular, neoplastic, and other inflammatory etiologies.

Although PG typically affects the lower limbs, atypical localizations occur, such as involvement of the breast, oral mucosa, and other non-acral sites, and can significantly delay diagnosis and often be mistaken for infection, vasculitis, or other ulcerative disorders. Herein, we describe a rare case of multifocal PG affecting atypical cutaneous and mucosal sites – including the breast and tongue – in a young woman with gastrointestinal (GI) symptoms suggestive of underlying IBD.

Case report

A 36-year-old female presented with a 10-day history of multiple painful ulcers distributed across the buttocks, abdomen, breast, and thighs. The initial lesion arose as a painful papule in the groin, which evolved into a pustule within 3 days. The pustule ruptured spontaneously, forming an ulcer that rapidly enlarged. Over the ensuing 5 days, additional lesions appeared sequentially across the buttocks, thighs, and breast, exhibiting similar rapid evolution. All lesions were painful and progressively increasing in size.

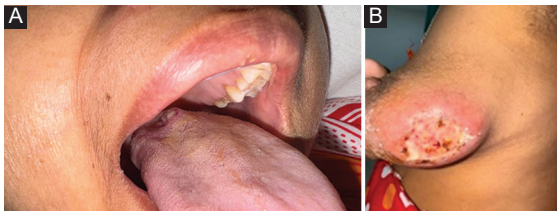

Figure 1. Ulcerations over oral mucosa and breast. **A:** ulcer of 0.5 x 0.5 cm^2 present over the tongue. **B:** 4 x 5 cm^2 ulceration present over the breast with peripheral erythema.

The patient reported intermittent fever, several episodes of bloody diarrhea for 20 days, and diffuse joint pain in the finger digits and lower back for a period of 1 month, associated with morning stiffness, which resolved as the day progressed. She denied any recent drug intake, weight loss, hematemesis, or preceding trauma. There was no history suggestive of sexually transmitted infections or chronic systemic disease.

On cutaneous examination, multiple irregular ulcers with undermined, violaceous borders and necrotic, purulent bases were observed over the abdomen, groin, buttocks, thighs, and the left breast. The breast ulcer measured 4 × 5 cm^2 and was tender with surrounding erythema. Furthermore, a 0.5 × 0.5 cm^2 ulcer was observed on the base of the tongue (Fig. 1A and B).

Given the history of bloody diarrhea and systemic symptoms, GI evaluation was pursued. Upper GI endoscopy revealed erosive esophageal ulcers, while colonoscopy demonstrated ulceration in the terminal ileum, raising suspicion for IBD. Laboratory investigations showed normal hematologic and biochemical parameters, except for markedly elevated C-reactive protein levels, suggesting a systemic inflammatory process.

A 4-mm punch biopsy from an ulcer on the buttock demonstrated mild hyperkeratosis, intraepidermal bullae consisting of polymorphs, epidermal, and dense dermal neutrophilic infiltration. Blood vessels reveal endothelial swelling and congestion with surrounding karyorrhectic debris. Deep dermis shows perivascular neutrophilic infiltration strongly suggestive of PG (Fig. 2A and B).

Pus culture from the ulcer base grew organisms sensitive to linezolid, indicating secondary bacterial infection. The patient was initiated on intravenous linezolid along with intravenous dexamethasone 8 mg (0.133 mg/kg), which was gradually tapered. In addition, oral cyclosporine at 5 mg/kg was started as a steroid-sparing immunosuppressant. Local wound care was provided following standard protocols for inflammatory ulcers.

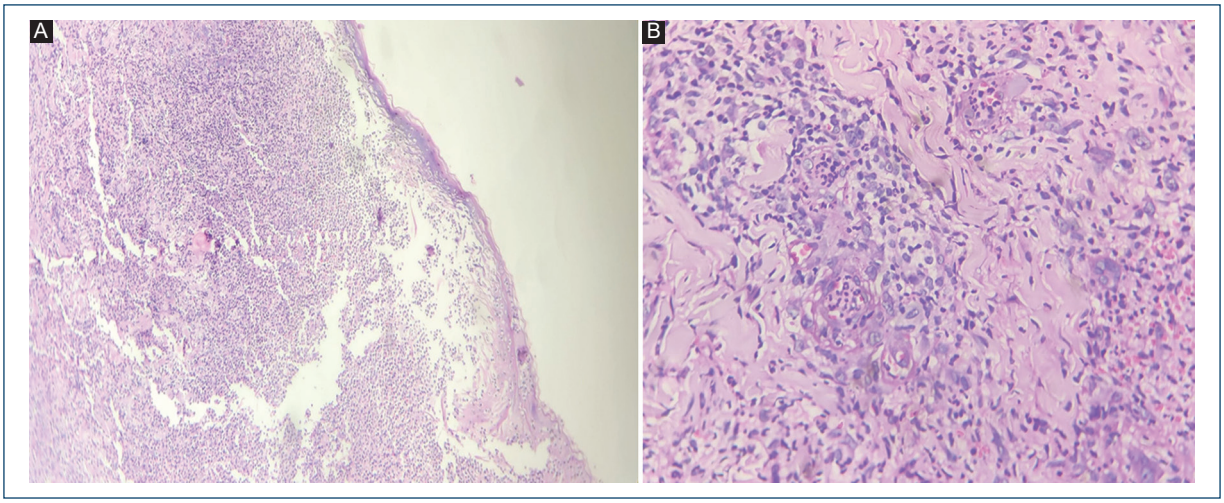

Figure 2. Histopathology of ulcer from buttock. **A:** H&E (10X) mild hyperkeratosis, intraepidermal bullae consisting of polymorphs, epidermal, and dense dermal neutrophilic infiltration. Blood vessels reveal endothelial swelling and congestion with surrounding karyorrhectic debris. **B:** (40X) Deep dermis shows perivascular neutrophilic infiltration strongly suggestive of PG.

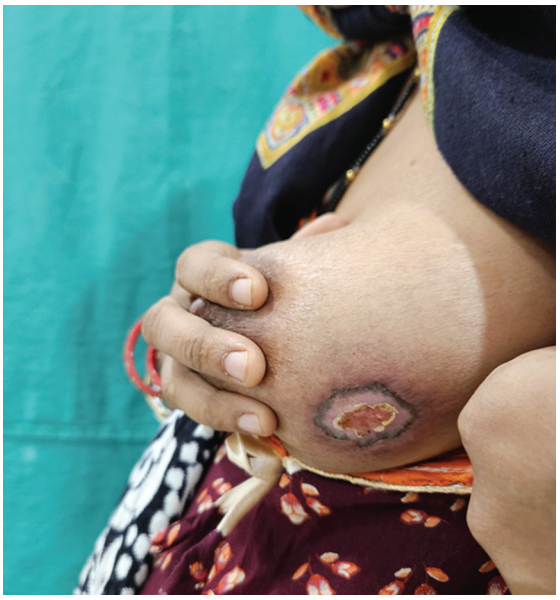

Figure 3. Reduction in the size of ulcer following therapy.

The patient showed significant clinical improvement, with a reduction in ulcer size, pain severity, and surrounding inflammation over the following month. She continues to be monitored on outpatient follow-up (Fig. 3).

Discussion

PG remains a complex and challenging dermatologic condition, particularly when it presents in an atypical manner. The classical ulcerative form is most common and is characterized by painful ulcers with undermined borders[1,2]. Diagnosis is largely clinical and one of exclusion, increasingly aided by consensus guidelines such as those proposed by Maverakis et al., which integrate clinical features with supportive histopathological findings[3]. The presence of dense dermal neutrophilic infiltrates and perivascular infiltrate in our patient corresponds to the major histological criteria proposed in earlier studies, including those of Su et al.[4]

The underlying pathophysiology involves abnormalities in innate immune regulation. Neutrophil dysfunction, upregulation of proinflammatory cytokines such as interleukin-8 (IL-8), IL-17, IL-23, and activation of inflammasome pathways involving IL-1β and IL-1α play central roles in driving neutrophilic infiltration and tissue destruction[5]. Matrix metalloproteinases-9 and 10 are additionally implicated in impaired tissue remodeling and delayed healing in PG lesions[6].

Importantly, the clinical course of PG does not necessarily parallel the severity of underlying systemic diseases. While PG is well associated with IBD, rheumatoid arthritis, and hematologic malignancies, several studies – including those by Weizman et al. – have demonstrated that PG severity often fails to correlate with IBD activity[7]. In our patient, the rapid and widespread evolution of skin lesions occurred independently of the severity of GI symptoms, reinforcing the need for clinicians to maintain vigilance even when systemic disease manifestations appear mild or non-specific.

The oral mucosal ulceration observed in our patient underscores the importance of recognizing non-cutaneous PG manifestations. Mucosal involvement is rare and easily misdiagnosed as aphthous ulcers, infections, Behçet disease, or manifestations of IBD itself. Similarly, the breast ulceration noted in this case is unusual, particularly in young women and in East-Asian populations, where very few cases have been documented. While PG has been reported on the face and head/neck regions, predominantly in elderly individuals and children[8], breast involvement has been sparsely described and can mimic abscesses, necrotizing infections, or malignancy, leading to unnecessary surgical intervention. Given the propensity for pathergy in PG, surgical procedures can precipitate lesion worsening, further complicating management.

In addition to classical PG, a number of PG-like ulcerative conditions have been described in the literature, including those associated with rare pathogens such as *Helicobacter cinaedi*, as reported by Dua et al.[9] These rare presentations further highlight the broad pathology spectrum and the importance of thorough infectious workup before initiating immunosuppressive therapy.

Treatment of PG includes mostly immunosuppression. Corticosteroids and cyclosporine remain first-line agents, with response rates of approximately 40-50% documented in earlier reviews[10]. Tumor necrosis factor-α inhibitors and newer biologics targeting IL-1, IL-17, or IL-23 have shown efficacy in refractory cases[11]. Adjunctive wound care is crucial, with emphasis on preventing secondary infections and avoiding mechanical trauma to minimize pathergy. Our patient responded favorably to combined intravenous corticosteroids and oral cyclosporine, along with judicious antibiotic use and careful wound care following the tissue, infection, moisture, and edge framework.

Conclusion

This case highlights a rare and multifocal presentation of PG, involving atypical anatomical sites such as the breast and oral mucosa. In the context of possible underlying IBD, recognizing such unusual patterns is crucial for timely diagnosis. The atypical distribution in this patient underscores the need for heightened clinical awareness, as PG at uncommon sites can easily mimic infectious, autoimmune, or neoplastic processes. Early diagnosis and prompt initiation of immunosuppressive therapy remain essential to prevent morbidity and avoid complications related to pathergy.

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Conflicts of interest

None.

Ethical considerations

Protection of human subjects and animals. The authors declare that no experiments on humans or animals were performed for this research.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from all patients, and secured approval from the Ethics Committee. SAGER guidelines have been followed as applicable to the nature of the study.

Declaration on the use of artificial intelligence (AI). The authors declare that no generative artificial intelligence was used in the writing or creation of the content of this manuscript.

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