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Single Case

An Unusual Presentation of Pyoderma Gangrenosum Leading to Systemic Inflammatory Response Syndrome

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Keywords
Pyoderma gangrenosum · Systemic inflammatory response syndrome · Circulatory shock

Abstract
This is a report of an atypical presentation of pyoderma gangrenosum (PG) in a 26-year-old male who had a negative septic screen. The patient had a life-threatening presentation requiring an intensive care unit (ICU) admission for vasopressor support. It was thought that the likely cause of circulatory collapse was an overwhelming cytokine reaction or systemic inflammatory response syndrome (SIRS) secondary to extensive PG lesions rather than septic shock. The patient presented with multiple large ulcers, the largest being 4 cm in diameter on the central chest. He developed fevers and circulatory shock preceding his ICU admission. Microbiological specimens, including blood cultures and wound swabs, were negative for any growth (bacterial, fungal, and tuberculosis). No infective foci could be identified as a cause of hemodynamic instability. During admission, the patient's condition was complicated by multi-organ dysfunction. Wound debridement extending to the deep fascia on the anterior chest, back, bilateral shoulders, and right upper thigh was deemed necessary and performed by the plastic surgery team. Histopathology showed abundant neutrophils but could not confirm an infective process. Overall, the patient made an impressive recovery with almost complete healing of all lesions following oral prednisolone alone. Based on the history and clinical and laboratory findings, a diagnosis of PG complicated by a SIRS was favored. Very few cases of neutrophilic dermatoses have been described in this way. A similar presentation

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has been described in a 76-year-old female with lower-leg ulcers who developed circulatory shock and required an amputation. Lesions continued to appear despite antibiotics and surgical treatment. Septic screen was negative. She was subsequently diagnosed with PG and recovered rapidly after steroid therapy.

Introduction

Pyoderma gangrenosum (PG) is an uncommon neutrophilic dermatosis that can affect individuals of any age, but it most commonly occurs in young and middle-aged adults [1, 2]. PG typically presents as an inflammatory papule or pustule which later develops into a painful ulcer with a violaceous rim and purulent base. Patients may have an underlying autoimmune disease, such as inflammatory bowel disease or rheumatoid arthritis. Neutrophilic dermatoses are a diverse group of disorders characterized histologically by the presence of abundant neutrophils in the absence of infection or true vasculitis. Neutrophilic dermatoses include PG, Sweet syndrome, SAPHO syndrome, generalized pustular psoriasis, Bechet syndrome, and several others. Typically, PG or other neutrophilic dermatoses do not present with life-threatening sepsis or circulatory collapse.

Case Report

A 26-year-old male with stable inflammatory bowel disease presented with folliculitis and multiple carbuncles and was admitted to the acute surgical unit for debridement and wound management. He had a background of ulcerative colitis and had had a similar presentation in 2012 requiring an intensive care unit (ICU) admission. Multiple lesions then developed into large ulcers with a violaceous border on the chest, back, and thighs. The patient then became tachycardic and hypotensive, requiring vasopressor support in the ICU. Wounds were debrided by the plastic surgery team, and broad-spectrum antibiotics were commenced. The patient remained unstable until steroid therapy was initiated following dermatology input. His 4-day ICU stay was complicated by multi-organ dysfunction with a creatinine of 125 μmol/L, mild ALT rise, INR of 1.8, pancytopenia, and increasing oxygen requirements. There was negative growth on blood cultures and wound swabs. In particular, he was negative for QuantiFERON assay, treponemal serology, HIV, hepatitis serology, acid-fast bacilli, and fungal elements. Central chest histology showed severe active inflammation with large numbers of neutrophils, leucocytes, necrosis, and microabscess formation. No organisms were identified. The lesions continued to heal well with steroid therapy with almost complete resolution at ~2 months of follow-up (Fig. 1, 2, 3, 4).

Discussion

This case showed an atypical and unusual presentation of PG. Considering the clinical features, the negative septic screen, and the rapid recovery after commencing steroid therapy, PG complicated by a systemic inflammatory response syndrome (SIRS) was considered as the most likely differential diagnosis.

Very few cases of neutrophilic dermatoses have been described in this way. A similar presentation has been described in a 76-year-old female with lower-leg ulcers who devel-

oped circulatory shock and required amputation. Lesions continued to appear despite antibiotics and surgical treatment [3]. The septic screen was negative. She was subsequently diagnosed with PG and recovered rapidly after steroid therapy. In 2 other cases, Sweet syndrome with SIRS was reported, where both patients required ICU admission and had a negative septic screen [4, 5]. There have also been reports of Sweet syndrome and cardiogenic shock following the initiation of azathioprine [6]. These cases demonstrated atypical but life-threatening presentations of some neutrophilic dermatoses in the absence of infection.

Conclusion

This case highlights a severe and life-threatening presentation of PG. SIRS may be a less recognized association with some neutrophilic dermatoses, as there have only been very few reports. SIRS requires early commencement of steroid therapy. This may prevent the need for ICU or surgical treatment.

Statement of Ethics

We would like to acknowledge the patient for providing informed consent of his case report and images to be used for the purpose of medical education. In addition, all authors meet the International Committee of Medical Journal Editors guidelines, and no authors or contributors have been omitted. Ethical standards have been complied with.

Disclosure Statement

All authors have no conflicts of interest or funding sources to declare.

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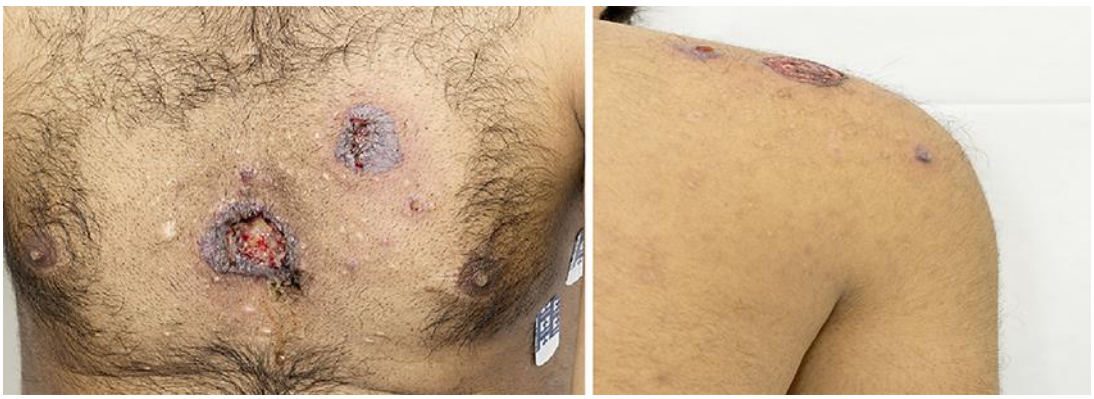

Fig. 1. Progress on day 2 after presentation: large violaceous ulcers on the anterior chest (left) and right shoulder (right).

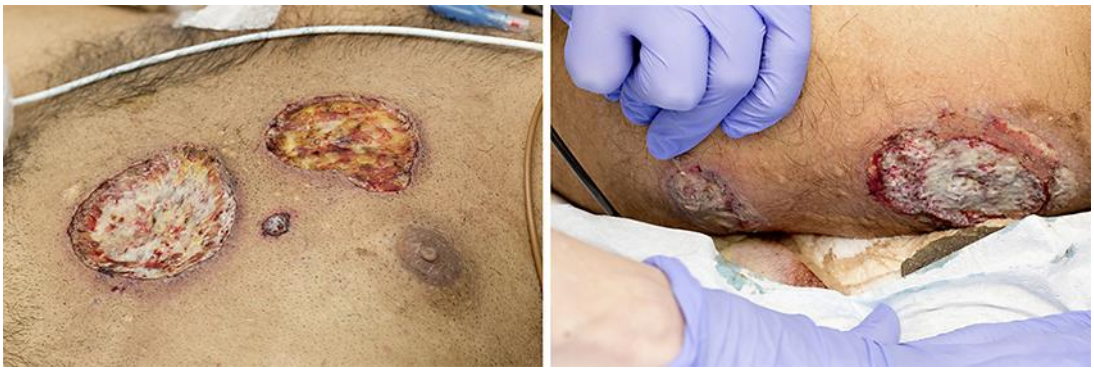

Fig. 2. Progress on day 4 after debridement: anterior chest (left) and back (right).

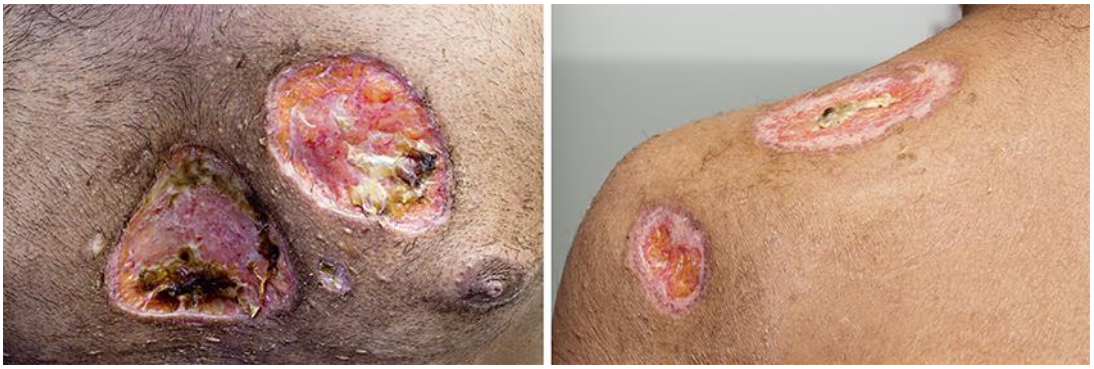

Fig. 3. Progress on day 23 after presentation: healing ulcers on the anterior chest (left) and left shoulder (right).

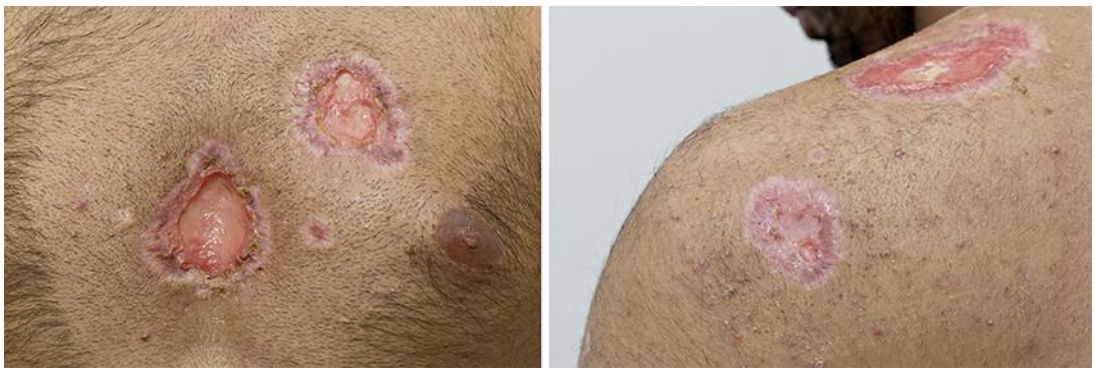

Fig. 4. Progress at 2 months of follow-up: anterior chest (left) and left shoulder (right) showing almost complete resolution.