

Case report of ulcerative pyoderma gangrenosum of the forearm: A challenging diagnosis

Gautam Kumar Vasnik¹, Raghvendra Singh Chittoria², Gitanjali Jain³, Sangram Bhattacharya⁴

From ¹Graded Specialist, Department of Pathology and Lab Medicine, ²Classified Specialist, Department of General Surgery, ³Graded Specialist, Department of Pediatrics, ⁴Classified Specialist, Department of Anesthesiology, Military Hospital, Nasirabad, Rajasthan, India

Correspondence to: Gautam Kumar Vasnik, Department of Pathology and Lab Medicine, Military Hospital, Nasirabad - 305 601, Rajasthan, India. E-mail: gautam_vasnik@yahoo.co.in

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ABSTRACT

Pyoderma gangrenosum (PG) is an uncommon disease associated with significant morbidity, characterized by the rapid development of a painful, necrolytic ulcer with undermined, irregular margins. Various clinical or histological subtypes have been illustrated in the literature. The diagnoses of various subtypes of PG are difficult due to its uneven presentation, clinical overlap with other conditions and not have specific diagnostic laboratory tests or classical histopathological findings. PG is frequently misdiagnosed as infection, results in delay or inappropriate treatment of the same, which leads to devastating results such as amputation of limbs and even demise. We report here the rare case of a 64-year-old woman who had initially presented with tender papules over the right forearm, later developed expanding ulcers and underwent serial debridements, which resulted in extensive ulcerative PG at the same site. This case highlights the challenges being faced while diagnosing ulcerative PG, emphasizes the key clinical and histopathological findings to aid in the final diagnosis, and also the clinical consequences of delayed diagnosis of this condition.

Key words: *Pyoderma gangrenosum, Skin ulcer, Surgical wound*

P yoderma gangrenosum (PG) is an uncommon disease associated with significant morbidity. The incidence is 3–10 per million per year, commonly affects women between 20 and 50 years [1]. PG was initially considered to be of infectious origin, now a neutrophilic inflammatory disease that is frequently related to inflammatory conditions such as inflammatory bowel disease (IBD), inflammatory arthritis (IA), and hematolymphoid malignancy [2-5]. At present, many aspects of PG are not well understood, and etiology still remains uncertain.

PG is usually associated with the rapid expansion of a painful, necrolytic ulcer with undermined, irregular margins and involves the lower appendages and trunk [2]. Many clinical subtypes have been illustrated in literature: Vegetative, bullous, pustular, and ulcerative [2]. PG lesions do not exhibit any specific histopathology. PG is further more a diagnosis of exclusion and to be considered only after frequent causes of painful necrolytic ulcers like infection and malignancy ruled out. PG ulcers typical exacerbate post-debridement through a mechanism known as pathergy (ulcer at trauma site) [5].

We report here a case of extensive ulcerative PG of the forearm as a consequence of serial debridement for suspected infected painful ulcers and further stressing on diagnosis and surgical management of the same. Neglect to reevaluate the patient after repetitive surgical attempts for non-healing ulcers promulgates the disease process and further the risk of infection and later with devastating patient morbidity.

CASE REPORT

A 64-year-old woman presented with painful erythematous lesion over the dorsal aspect of the right forearm for the past 7 days. Later after 3 days, she developed a superficial painful ulcer at the same site.

General examination revealed mild pallor and other vital signs were normal. Systemic examinations were unremarkable. Local examination revealed a superficial tender ulcer with irregular margins, measuring 2 cm×2 cm with surrounding erythema and serous discharge. Serous fluid examination revealed no viral inclusions and no growth on culture. She was admitted at this time and started on injectable antibiotics.

In the next few days, her wound had worsened with enhanced in ulcer size, measuring 5 cm×3 cm with serous discharge. Keeping in mind, surgical debridement of the necroulcerative area was done. Subsequently, when the patient underwent a dressing change, it was found that the ulcer had even worsened and the once healthy wound margins had become necrotic. Hence, further debridement was done and excised tissues were sent for histopathology. Higher antibiotics were added for suspected postoperative wound infection.

The patient's wounds continued to worsen in spite of all measures in subsequent days. After the third debridement, the ulcer had further increased in size, measuring 25 cm×7 cm and also involved deeper tissues, as shown in Fig. 1a and b. Biopsy revealed ulcerated epidermis and suppurative dense inflammation

comprising of polymorphs and eosinophils, reaching up to dermis and subcutis, as shown in Fig. 2a-c. No organisms, acid-fast bacilli, and fungal elements were seen. No vasculitis, granuloma, and malignancy noted. Antinuclear antibodies, rheumatoid factor, and anti-neutrophil cytoplasmic antibodies were negative. Considering history, clinical examination, pathology, negative culture, non-responsiveness to antibiotics, and histopathology, a diagnosis of ulcerative PG was considered.

The patient was given 50 mg prednisolone with appropriate wound care. After 1 week, significant improvement was noted, with a resolution of necrotic margins and the formation of granulation tissue. A month after admission, the patient underwent successful split skin grafts, as shown in Fig. 1c. At monthly follow-up, there was no recurrence of the disease.

DISCUSSION

Most of the cases of PG are commonly mismanaged as ulcers with an infective etiology resulting in repetitive surgical debridement. Conditions with similar presentation as PG are skin infections



Figure 1: (a) Right forearm pyoderma gangrenosum (PG) after multiple debridements; (b) PG involving subcutaneous fat; (c) after treatment with immunosuppressive medications and split skin graft

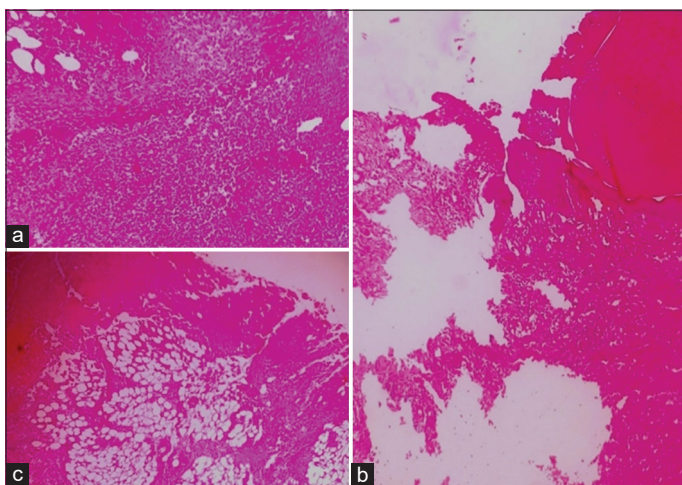


Figure 2: Histological findings from punch biopsy of the right forearm ulcer. (a) Upper left image (×400): Dense suppurative inflammation comprising predominantly of neutrophils and debris; (b) right image (×400): Extensive skin ulceration; (c) lower left image (×400): Inflammation involving subcutaneous tissue

(most common) followed by other causes such as venous insufficiency, peripheral arterial disease, exogenous skin damage (e.g., burns and insect bites), cutaneous autoimmune disorders (e.g., bullous pemphigoid), cutaneous malignancy, cutaneous vasculitis (e.g., cryoglobulinemia and polyarteritis nodosa), and necrobiosis lipoidica. This necessitates that a detailed history for initial lesion such as a papule/vesicle, its progression, associated pain, any other underlying systemic disorder, and symptoms associated with pathology to be considered followed by relevant investigations [6]. Unfortunately, repeated debridement worsens PG and most of the time diagnosis being made after failure of initial treatment, as was in our case [7].

Maverakis *et al.* [5] assembled a group of diagnostic criteria for PG following the Delphi consensus exercise using the RAND/ University of California, Los Angeles appropriateness method [8] and validated the same. Delphi exercise integrated one major and eight minor criteria. The major criterion is the presence of neutrophilic infiltrates in histopathology. The eight minor criteria are (a) exclude infection; (b) pathology; (c) IBD or IA; (d) papule, pustule, or vesicle ulcerating within 4 days since the time of appearance; (e) peripheral erythema, undermining margins, and pain at ulcer site; (f) multiple ulcerations, at least one on an anterior lower leg; (g) cribriform or wrinkled paper scars at healed ulcer sites; and (h) decreased ulcer size within 1 month of initiating immunosuppressant. As per the Delphi exercise, any 4/8 minor criteria had a sensitivity of 86% and specificity of 90%. In our case, major and six minor criteria, namely, (a), (b), (d), (e), (g), and (h) were encountered.

At present, there are no specific management protocols for PG. No risk factors or perioperative management protocol have been identified as statistically significant predictors of disease reappearance [7]. There are no gold standard treatment guidelines for PG, and current therapy mainly based on the severity and progression of the disease course [9]. Topical therapy by means of control exudation, wound dressings, and systemic therapy in the form of immunosuppressant's like corticosteroids is at first used to avert progression further seize the inflammation. In steroid-resistant cases, cytotoxic drugs like cyclosporine should be used. Split skin grafts to be considered in cases of aggressive disease and only after partial remission with immunosuppression [10].

CONCLUSION

Ulcerative PG is rare in occurrence and often demonstrates to be a diagnostic challenge. It is imperative to reconsider a diagnosis when the disease course deteriorates or does not react to management. This case report emphasizes the key clinical and histopathological findings to aid in the final diagnosis and also shows the clinical consequences of delayed diagnosis of this condition.

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