Case Report

Pyoderma gangrenosum: A clinician's nightmare

Bindhu Bhaskaran[1], Mittu John Mathew[2], K. N. Vijayan[2], Asha Zacharia[3]

Departments of [1]Family Medicine, [2]General Surgery and [3]Dermatology and Cosmetology, Kerala Institute of Medical Sciences, Trivandrum, Kerala, India

Abstract

Pyoderma gangrenosum (PG) is a rare disease and that affecting specifically the sole of the foot, is even rarer. Here, we report the case of a 54-year-old female admitted with a painful ulcer on the sole of the right foot which was initially treated with empirical antibiotics and debridement. The disease was found to spread rapidly after each debridement. The culture reports were negative; rheumatology workup and Doppler study were within normal limits. A clinical suspicion of PG was made and was confirmed with tissue biopsy. She was started on oral steroids following which she dramatically improved. Thus, when a patient presents with a rapidly expanding painful ulcer in a vascular limb that is refractory to antibiotic treatment and exacerbating on debridement, it is imperative to consider the possibility of PG.

Keywords: Pyoderma gangrenosum, spreading, ulcer

Introduction

Pyoderma gangrenosum (PG) is a rare dermatological condition characterized by the rapid progression of a painful, necrolytic ulcer with an irregular, undermined border and commonly affects the lower extremities, mainly in the pretibial area.[1] The peak of incidence occurs between the ages of 20 and 50 years with women being more often affected than men.[2,3] Although PG is most common in the pretibial area, it can occur at any site including breast, hand, trunk, head and neck, and peristomal skin. PG has never been reported on the sole of the foot. We report the case of a patient who developed PG on the sole.

Case Report

A 54-year-old female who is known to have diabetes and hypertension presented with a painful ulcer on the sole of the right foot. It developed following an injury sustained by stepping on an earring 1 month ago. She was initially treated in a primary care center with antibiotics and debridement of the ulcer. When she presented to us, she had a painful necrotic patch of 3 cm × 4 cm on the sole of right foot. Blood investigations showed a white blood cell count of 31,700 cells/cumm, C-reactive protein level of 218 mg/l, and erythrocyte sedimentation rate of 105 mm/h. Wound swab was taken, the necrotic patch was debrided totally, and tissue and blood culture samples were sent. Empirical antibiotic therapy with cefoperazone + sulbactam and linezolid were started as per hospital protocol. The next day on review multiple ecchymotic lesions were found surrounding the ulcer which progressed over 2 days to form new necrotic patches [Figure 1]. Vasculitic workup was done after consulting with rheumatologist. Infectious disease consultation was sought and the antibiotics were stepped up to meropenem and teicoplanin. Arterial Doppler study of both lower limbs was done which showed adequate blood flow. Debridement was done again, and tissue biopsy was sent. The next day on review the lesions were found to be spreading in a similar fashion with violaceous purple border and multiple pustules, and it was extremely painful. All the culture reports were sterile. Antinuclear antibody profile, anticardiolipin antibody IgM and IgG, cryoglobulin, rheumatoid factor, anti-lupus antibody were negative. A dermatology opinion was sought, pathergy test was done, and a clinical suspicion of PG was made which was confirmed with the tissue biopsy. Giemsa, Ziehl–Neelson and Grams staining of the tissue specimen were also done which revealed no organisms. The patient was started

Address for correspondence: Dr. Bindhu Bhaskaran, Kerala Institute of Medical Sciences, Trivandrum, Kerala, India.
E-mail: drbindhubhaskar@gmail.com

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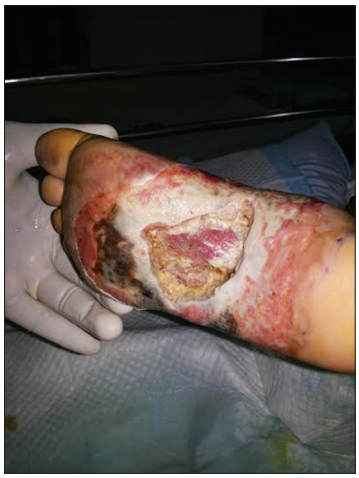

Figure 1: Spreading ulcer on the sole with pustules at the floor

on oral steroids (prednisolone 60 mg) under antibiotic coverage, and the daily dressing was done. The ulcer stopped spreading further, and she improved significantly. She is now kept on regular follow-up.

Discussion

PG is an uncommon, ulcerative cutaneous condition of uncertain etiology.

Classically, lesions begin as tender papules, papulopustules or vesicles, evolving into painful and rapidly enlarging ulcers. Healing frequently leaves a cribriform scar, which may lead to considerable disfiguring.[1,4] On histology, PG lesions do not display specific features, being typically characterized by a prominent dermal neutrophilic infiltrate with abscess formation, with no signs of infection and usually without significant vasculopathy.

PG has a reported incidence of 3-10million cases per year, with nearly half of the cases associated with an underlying systemic disease, such as inflammatory bowel disease, hematologic conditions, or autoimmune disorders.[5]

PG is a diagnosis of exclusion and can only be made after common causes of ulcers such as infection and malignancy have been ruled out. Diagnosis relies on clinical signs first and is supported by histopathology. No laboratory parameters are available. Clinically, it can be mistaken as necrotizing fasciitis, hidradenitis suppurativa, or herpes infection. Pathologically, both PG and necrotizing fasciitis can be characterized by sterile infiltration of the skin by neutrophils with additional nonspecific features of vasculitis, necrosis, edema, and abscess formation.[6] While the treatment for necrotizing fasciitis and hidradenitis includes antibiotics and aggressive surgical debridement, PG is unresponsive to antibiotic therapy and debridement exacerbates PG through a process called pathergy.[7] Its ability to mimic superficial wound necrosis of infectious etiology could lead to a delay in diagnosis and correct management.

Differential diagnosis for PG includes: Infections-bacterial including tuberculosis, viral (herpetic ulcers), parasitic, topical mycoses (sporotrichosis), vascular arterial or venous ulcers, vasculitis (antiphospholipid syndrome, vasculitic rheumatoid arthritis, systemic lupus erythematosus, Wegener's granulomatosis, Behcet's disease), malignancies (squamous cell carcinoma, cutaneous lymphoma), insect bites, self-inflicted ulcerations, and Sweets syndrome.[8]

The mainstay of treatment is long-term immunosuppression often with high dose of corticosteroids (prednisolone 0.5–2 mg/kg/day) or low dose of cyclosporin (3–6 mg/kg/day).[9] Other immunosuppressors and cytostatics can be used as steroid-sparing agents, namely azathioprine, sulfasalazine, dapsone, thalidomide, minocycline, clofazimine, methotrexate, mycophenolate mofetil, tacrolimus, intravenous immunoglobulin (IVIG), and cyclophosphamide.[10] Multidrug therapy should be considered for refractory disease. The combination of systemic steroids and cyclosporine is commonly used. When this combination fails, less conventional treatments are chosen. The common combinations are (1) methotrexate and infliximab, and (2) cyclosporine, mycophenolate mofetil, and prednisone.[9] IVIG is a useful therapeutic option in refractory PG and can be considered in cases of resistance to or intolerance of standard immunomodulatory therapy.[10] In addition, topical corticosteroids or tacrolimus can also be used. The daily dressing of the ulcer with nonadherent foams is recommended. The underlying systemic condition must also be treated.

Conclusion

When a patient is presenting with a rapidly expanding painful ulcer in a vascular limb which is refractory to antibiotic treatment and is exacerbating on debridement, the possibility of PG must be considered. Early diagnosis and treatment with corticosteroids and immunosuppressants can heal the lesion with minimal scarring and prevent devastating consequences such as limb amputation and death.

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Conflicts of interest

There are no conflicts of interest.

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