Pyoderma Gangrenosum Over Hypertrophic Scar: Report of a Rare Presentation

Abstract

Pyoderma gangrenosum (PG) is a chronic neutrophilic dermatoses characterised by large ulcer healing with cribriform scar. PG can rarely occur over the healed scar. Its diagnosis depends on clinical morphology and exclusion of other diseases. We report a case of large pyoderma gangrenosum ulcer on a hypertrophic scar since 1 year. Biopsy from ulcer edge was suggestive of PG and hypertrophic scar. The hypertrophic scar had been formed by healing of a large ulcer 2 years back. The ulcer healed to two-third size over next three months with monthly dexamethasone pulse, daily oral steroid and dapsone. This case was unique since the PG ulcer had occurred over a hypertrophic scar. Such a presentation in PG is rare and physicians should be aware of such occurrence of PG.

Keywords: *Dexamethasone pulse, hypertrophic scar, keloid, pathergy, pyoderma gangrenosum*

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Pyoderma gangrenosum (PG) is a rare, chronic neutrophilic dermatoses of unknown etiology.[1] The ulcer is characterized by large size, bluish raised margin, undermining edge, floor covered with granulation tissue, and necrotic slough. The PG ulcers heal with atrophic cribriform scar.[2] PG ulcer can occur over surgical scar sites rarely.[3] Occurrence of PG on hypertrophic scar has not been described. We report a case of PG occurring over the hypertrophic scar of another healed ulcer.

Case Report

A 17-year-old female presented with painful nonhealing ulcer over a large scar on the left leg since 1 year. The ulcer did not improve on treatment with various topical and oral medications. The lesions started with formation of red blister, which doubled in size within 2 days and ruptured to form an ulcer within 1 week period. It took about a year to heal forming a large hypertrophic scar. After around 1 year, she developed another ulcer over the scar, which progressed to the present size over 1 month. There was no history of any prior trauma, abnormal bowel habit, joint pain, or frequent bleeding from the ulcer. On examination, there was a well-defined ulcer of size 11.5 cm × 6.7 cm over a hypertrophic scar (20 cm × 8 cm) over the left lower leg [Figure 1]. The base of ulcer had red granulation tissue. The margin was pigmented with undermining at some portions and punched out edge. The healed scar had resulted in flexion deformity at the ankle joint [Figure 1]. The examination of other parts of skin was normal. The site of Bacille Calmette Guerin injection had healed without hypertrophic scar. Provisional diagnosis of PG, Marjolin ulcer and posttraumatic eczema was made. Biopsy from edge of ulcer was done for histopathological examination. All routine investigations, such as complete blood count, peripheral smear comments, were within normal limits. Pus on Gram stain and culture sensitivity did not show any organism. Biopsy showed subcorneal neutrophilic pustules with dermis showing neutrophilic infiltration [Figures 2-4]. The upper and mid dermis showed fibrillar collagen in orderly arrangement [Figures 2 and 3]. Based on history, clinical morphology and histopathology a final diagnosis of PG over a healed hypertrophic scar was made. The patient was treated with dexamethasone pulse (100 mg dexamethasone in 300 mL 5% dextrose infused over 2 hours) on 3 consecutive days with 30 mg oral prednisolone and 100 mg dapsone daily.

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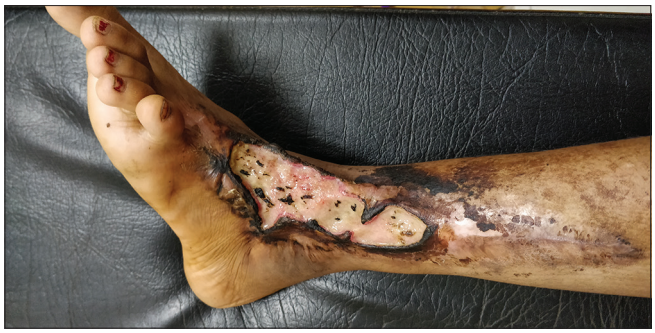

Figure 1: Healed scar with central part showing hypertrophy over which well-defined ulcer with raised margin, undermined edges, and floor covered with necrotic slough

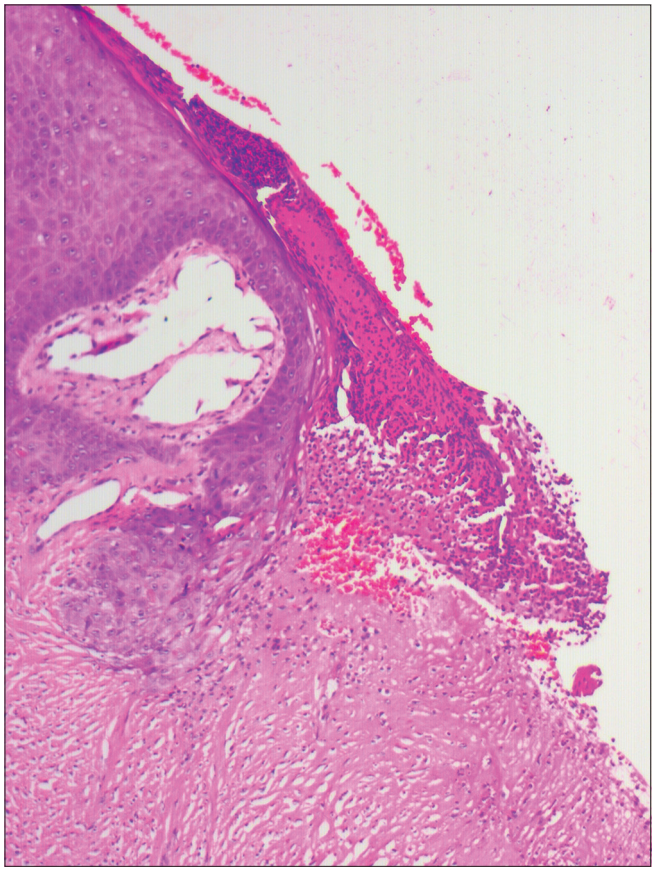

Figure 3: Low power view showing shallow ulcer with infiltrates in the floor. [H&E, 100x]

There were signs of improvement in the form of decrease size and re-epithelization of the ulcer characterized by decreased depth of ulcer over 2 weeks with slopping of the edge. After seven doses of monthly dexamethasone pulse and oral prednisolone in tapering doses and dapsone 100 mg, the ulcer healed completely [Figure 5a and b]. The patient is on regular follow-up.

Discussion

PG is a rare, noninfectious, and neutrophilic dermatosis. The lesions typically begin as pustules or papules that

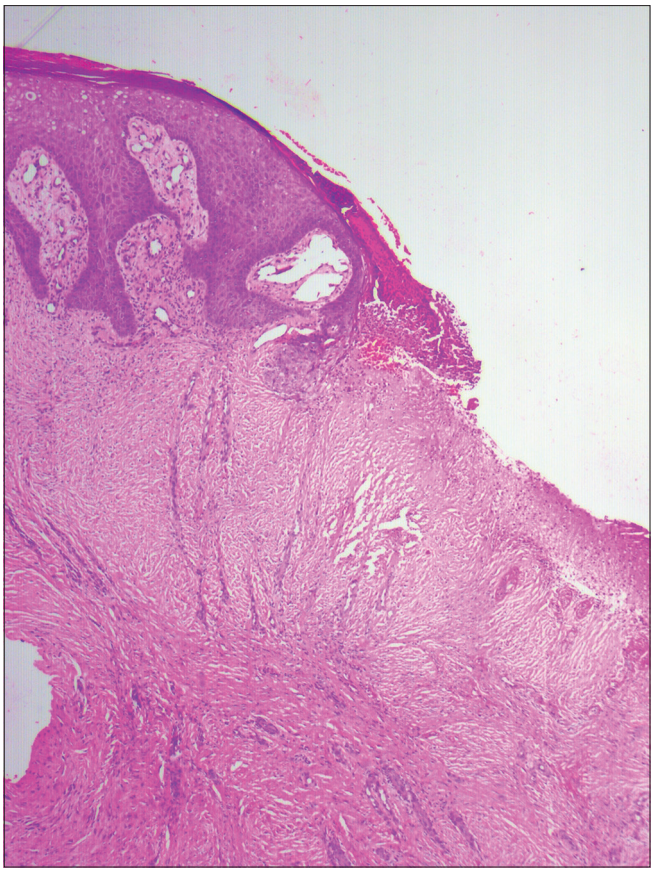

Figure 2: Scanner view of histopathology showing ulcer edge with base showing fibrillar collagen. [H&E, 40x]

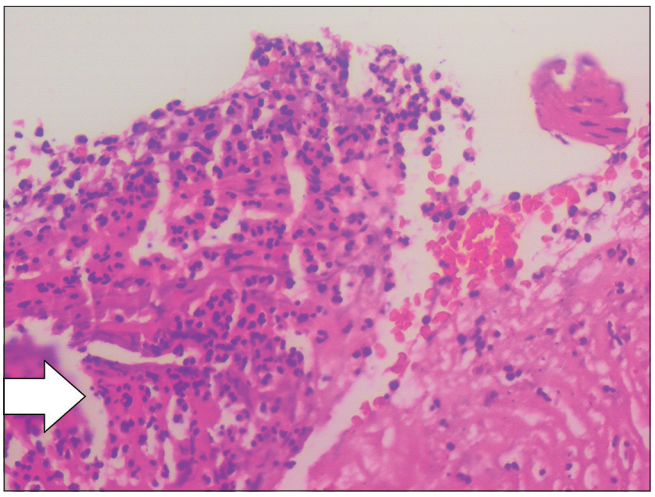

Figure 4: High power view showing neutrophilic infiltrates with segmented nuclei. [H&E, 400x]

undergo central necrosis and extend peripherally to produce ulcers.[4]

Abnormalities in the function of inflammatory cytokines, the immune system, and neutrophils combined with specific genetic mutations are implicated in the pathogenesis of PG.[5]

Pathergy phenomenon can give rise to enlargement of ulcer and new lesion formation at the site of trauma-like

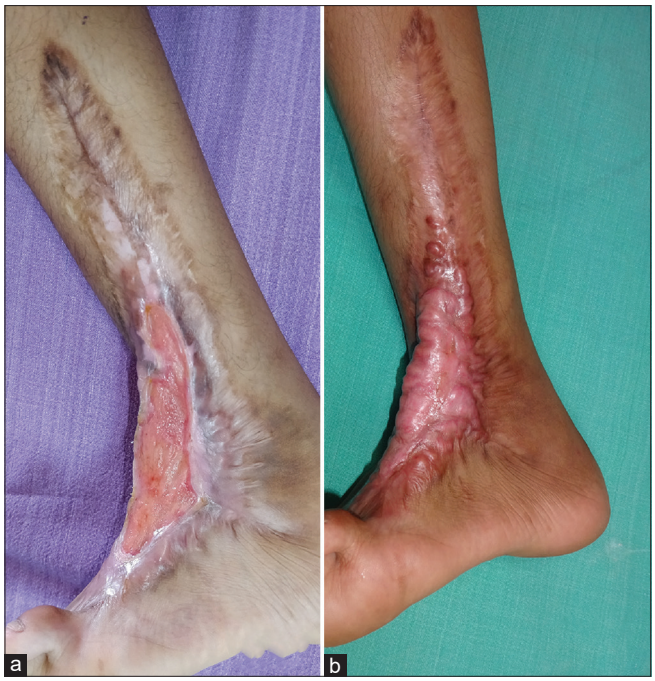

Figure 5: (a) Response after 1 month of treatment with sloping edges and decrease in depth of floor of the ulcer. (b) Response after 7 months of treatment with complete healing of the ulcer

injection site and debridement/dressing of PG ulcer.[6] These ulcers heal with atrophic cribriform scars. Literature search did not reveal any report of PG ulcer occurring over hypertrophic scars. Postsurgical scar PG occuring within 7 days to as long as 7 years of surgical intervention is known.[3,7] Morphology of PG ulcer occurring over postsurgical scar is similar to that of classical ulcer with undermined edges, bluish margins, and floor covered with granulation tissue.

Diagnosis of PG is by clinical suspicion and exclusion of other causes. The clinical suspicion is mostly from large size ulcer, rapid evolution of ulcer to reach the large size in short period (days to weeks), and pathergy phenomenon. The diagnostic criteria include 1 major criterion-biopsy of ulcer edge demonstrating neutrophilic infiltrate-and 8 minor criteria: (1) exclusion of infection; (2) pathergy; (3) history of inflammatory bowel disease or inflammatory arthritis; (4) history of papule, pustule, or vesicle ulcerating within 4 days of appearing; (5) peripheral erythema, undermining border, and tenderness at ulceration site; (6) multiple ulcerations, at least 1 on an anterior lower leg; (7) cribriform or "wrinkled paper" scar(s) at healed ulcer sites; and (8) decreased ulcer size within 1 month of initiating immunosuppressive medication(s). Fulfilling 4 of 8 minor criteria yields sensitivity of 86% and specificity of 90%, respectively in the diagnosis of the PG ulcers.[2]

Histopathologic features of PG are nonspecific, but can aid the diagnosis by excluding other potential diagnoses, such as a primary vasculitis, infections, and malignancies. Characteristically, PG lesions have a dense neutrophilic infiltrate, but leukocytoclastic vasculitis, necrosis, and even granulomatous inflammation also have been described.[8]

Treatment of PG is variable. PG ulcer can heal spontaneously.[9] Drugs, such as glucocorticoids, cyclosporin-A, dapsone, clofazimine, colchicine, azathioprine, mycophenolate mofetil, and cyclophosphamide have been used.[8] Dexamethasone pulse therapy has been used in treatment of PG.[10]

In the present case, the PG ulcer over a hypertrophic scar fulfilled 1 major and 5 minor criteria. There was history of spontaneous healing of the previous ulcer with disfiguring hypertrophic scar. However, formation of hypertrophic scar could not be explained in our patient since she did not have any history of keloidal tendency. In the present case, the ulcer had rapid course of evolution within 1 month to reach the large size, possibly because of site of occurrence (i.e., scar tissue) and classical morphology, thus clinically suggestive of PG. The ulcer improved significantly with dexamethasone pulse and daily doses of 30 mg prednisolone and 100 mg dapsone.

We report this case because of its unique presentation and to emphasize upon the fact that the morphology of PG ulcer appearing over a scar is similar to the classical PG.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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