CASE REPORT

Surgical intervention in aggressive ulcerative pyoderma gangrenosum

Maham Mudassir[1], Rabia Ali[2], Sikandar Saeed[3], Ali Rafique Mirza[4]

Abstract

Pyoderma Gangrenosum (PG) is a rare, debilitating, and painful disease of the skin. Its aetiology and pathophysiology are not well understood. However, it is known that PG is not bacterial in origin, as previously believed. A significant number of cases of PG report a phenomenon called pathergy, which is characterized by the appearance of new lesions after the application of trauma to the skin. This represents a unique challenge for surgeons in cases that are refractory to medical therapy. The objective of this study is to review past literature and report a case of PG in a 19-year old woman, who presented with recurrence, after undergoing skin grafting one year back. The patient was referred for split thickness skin grafting (STSG) to reduce the psychological and physical morbidity as a result of this disease.

Keywords: pyoderma gangrenosum, wound, split thickness skin grafting.

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Introduction

Ulcerative Pyoderma Gangrenosum (PG) is a neutrophilic necrotizing dermatitis. It typically presents as recurrent cutaneous ulcers with mucopurulent discharge. The ulcers are painful and have bluish borders surrounded with erythema. In many cases, it is associated with an underlying inflammatory disease such as rheumatic disorders and inflammatory bowel disease.[1] The etiology of PG is poorly understood. However, it has been suggested to be a neutrophilic pathology with overexpression of inflammatory mediators. In 30% of patients with PG, pathergy has been observed. Pathergy is a term used for skin reaction to any mechanical trauma such as a cut or a laceration.[2] Immunosuppressive therapy is the first-line treatment for this disease. However, the following case report is presented to highlight the role of STSG combined with medical treatment.

Case Report

A 19-year old girl with PG was referred to the plastic surgery department of Shaikh Zayed Medical Complex, Lahore on 20th September 2019 for the management of extensive ulcers on both legs, covering them almost circumferentially, both thighs and dorsum of both feet including digits. She also had ulcers on dorsum of both hands around metacarpophalangeal joints and dorsum of proximal interphalangeal joints of all fingers, both elbows, buttocks, groin area and in the oral cavity. She had developed the disease seven years ago when she noticed papules on both feet. Later, they became itchy and turned into pustules. She also developed similar pustules on her hands. These pustules turned into ulcers within days and she was diagnosed with PG on histopathology. A year ago, she had skin grafting done for the coverage of lower limb wounds from another tertiary care hospital and was now admitted to the dermatology ward and was placed on oral steroids, topical steroids, dapsone and intravenous antibiotics.

The ulcers were painful with purulent discharge, so multiple debridements were performed under general anaesthesia and steroid cover until all the wounds developed healthy granulation. Split-thickness skin grafting (STSG) of the wounds was planned and her steroid dose was tapered to a minimum till the procedure day. Her pre-operative blood tests were as follows: Haemoglobin, 9.27 mg/dL (12-16 mh/dL); white cell count, 12.7 x 10^9/L (4-11 x 10^9/L); platelets, 464 x 10^9/L (150-400 x 10^9/L); RBC count, 3.45 x 10^{12}/L (4.2-5.4 x10^{12}/L); Haematocrit, 29.9 % (36.1-44.3%). Serum albumin level was as low as 2.1 g/dL (3.4-5.4 g/dL) and total serum proteins were 4.36 g/dL (6.4-8.3 g/dL). Rest of the liver function tests; renal function tests and serum electrolytes were normal. Serum iron level was

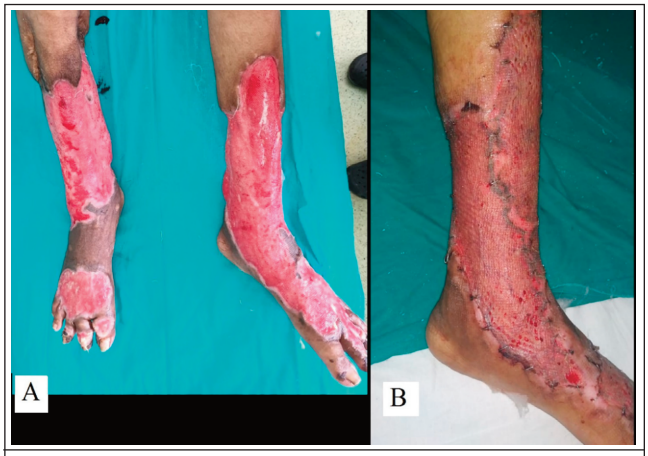

Figure-1: A; granulated ulcers after multiple debridement, B; fourth post-operative day.

[1,4]Department of Plastic Surgery, Shaikh Zayed Hospital, Lahore, Pakistan;
[2,3]5th Year MBBS Student, Shifa College of Medicine, Islamabad, Pakistan
Correspondence: Rabia Ali. e-mail: rabiaali5266@gmail.com

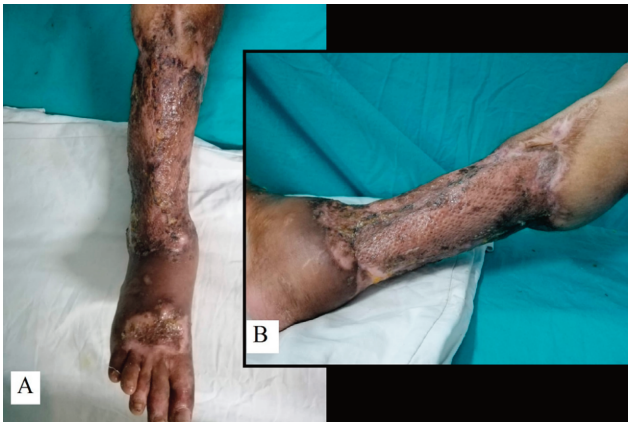

Figure-1: A and B; split thickness skin graft on right leg one month post-operatively.

23 µg/dL (60-170 µg/dL) and total iron binding capacity was 185 µg/dL (240-450 µg/dL). She was transfused one unit of packed red blood cells (RBC) a day before surgery.

STSG of thickness between 0.4 – 0.6 mm were harvested with a dermatome from available areas of virgin or healed skin on both her thighs, sparing the ulcers. Grafts were meshed on a 1.5:1 Mesher plate. Recipient wounds on thighs, legs and feet were debrided and grafts were applied with surgical skin staples. Dressing was applied with a sterile, paraffin-soaked tulle.

Post-operatively, the patient had low-grade fever and persistent sinus tachycardia for which she was shifted to intensive care unit (ICU). Systemic steroids, dapsone and intravenous antibiotics were continued post-operatively as the patient was monitored in the ICU for two days. She was shifted back to the dermatology ward with stable vitals. Two units of packed RBCs were transfused. Her dressing was changed on the fourth post-operative day with satisfactory results. Dressings were then applied on alternate days. On tenth post-operative day, excellent graft take was observed at all recipient sites. Donor site dressing was changed on the twentieth post-operative day and wounds had started to heal from the edges whereas central areas were still raw. These wounds were managed conservatively with sterile paraffin-soaked tulle dressings and later saline dressings. Recipient sites healed completely three weeks post-operatively, while donor sites took two months to completely epithelialize. Systemic steroids and dapsone were administered throughout this period while intravenous antibiotics were discontinued three weeks post-operatively. Wounds on dorsum of hands and fingers, elbows, groin and buttocks were left to heal by secondary intention. Consent was taken from the patient for publishing her case report along with the pictures and maintaining the anonymity.

Discussion

The incidence and distribution of PG have never been properly determined in a population study. The estimated incidence is said to be 3-10 per million population per year.[3] PG has been categorized into four types. They are known as ulcerative, vesicular-bullous, pustular and lastly, superficial granulomatous. The ulcerative type is usually found on the lower extremities as a rapidly progressing, purplish ulcer with unclear borders. The vesicular-bullous PG appears on the face, trunk and upper extremities. It presents as blisters or superficial bullas. The pustular PG is commonly found on the legs and the trunk and presents as multiple painful pustules. The last type, superficial granulomatous PG appears on the trunk as superficial ulcers with a granulomatous appearance.[4] Treatment of PG mainly lies in the domain of a dermatologist. Before the start of the treatment, obtaining culture and sensitivity for the wounds is required. Initially the patient should be started on a systemic corticosteroid with pulsed doses, infliximab and cyclosporine.[5] Immunosuppressive agents such as azathioprine, colchicine and cyclophosphamide have been used alongside corticosteroids. Tacrolimus and intravenous immunoglobulins have also been prescribed. The therapeutic results of corticosteroids with cyclosporine are by far the best.[6]

Surgical intervention in PG is controversial due to pathergy and has to be undertaken with extreme caution. However, if the disease is quiescent with sufficient immunosuppressive therapy, it may be beneficial. Healing of the wounds can be improved with negative-pressure wound therapy, split-thickness or full-thickness skin grafting.[7] Surgical methods include aggressive ulcer excision, recipient-site preparation, grafting of autologous cultured keratinocytes, skin as well as muscle flap coverage. In a study carried out by Saracino et al.,[8] no incidences of pathergy were reported after performing 16 surgical interventions in conjunction with immunosuppressive therapy. In contrast, some studies have observed that PG is aggravated at surgical sites due to pathergy.[8] Long et al proposed a solution to approximate the edges of the wound by subcuticular sutures. This method will avoid any skin trauma and the inflammatory cytokines would not be triggered. However, so far no research has been conducted on this method.[9]

In our case, the patient presented with ulcerative PG which did not resolve completely by previous grafting. We performed STSG under cover of systemic steroids, dapsone and antibiotics due to the extensiveness of ulceration, which could not be left to heal secondarily with medical therapy alone. Although the recipient wounds healed completely in three weeks, the donor sites took more time

than it does in a patient without PG. Nevertheless, with good wound care complete healing was observed after two months.

Conclusion

The rationale for STSG combined with immunosuppressive therapy in this patient was to reduce physical and psychological morbidity, as well as to reduce the overall cost of wound care and length of hospital stay, as compared to healing with secondary intention with medical therapy alone. However, more interventional studies are needed in order to make a direct comparison between the treatment modalities.

Abbreviations: PG: pyoderma gangrenosum; STSG: Split-thickness skin grafts; RBC: red blood cells; ICU: intensive care unit

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