Pyoderma gangrenosum associated with Burkitt lymphoma

G Shopana[1], K L P D Seneviwickrama[2], S K P Lekamge[3], H M P Gunathilake[3]

Sri Lanka Journal of Dermatology, 2022-2023, **23**: 47-49

Abstract

Pyoderma gangrenosum (PG) is a cutaneous condition characterised by the presence of a painful cutaneous ulcer showing rapid progression. PG is associated with a concomitant systemic disease in 50 to 70% of cases[1].

Herein, we report a 7-year-old patient, with a history of Burkitt lymphoma in remission, who presented with PG, and was treated successfully with corticosteroid and azathioprine.

Introduction

Pyoderma gangrenosum is an uncommon neutrophilic dermatosis characterized by rapid development of painful ulceration with undermined edge and gun metal gray borders.

The aetiology of PG is unclear but is thought to be a reactive inflammatory dermatosis. Although half of the cases are idiopathic, PG may be associated with inflammatory bowel disease, connective tissue disease, and hematological disorders[1]. PG may be associated with myeloid malignancies. Less information is available about the association of PG with lymphoid malignancies.

Burkitt lymphoma (BL) is an aggressive non-hodgkin B-cell lymphoma but with intense chemotherapy treatment disease prognosis is excellent in children but poor in adults.

Case presentation

5 year old girl with childhood bronchial asthma presented with gradual onset of colicky abdominal pain which worsened over 3 days with loss of appetite. There was no alteration in bowel habits or vomiting. Further investigation showed presence of intussusception requiring surgical intervention. During the surgery, child was found to have gastrointestinal tumor which predisposed the intussusception and it was histologically confirmed as Burkitt's lymphoma. Child was treated with cycles of chemotherapy.

Nearly 1 year after the diagnosis of Burkitt's lymphoma child was transfused with blood products due to investigation abnormality but child developed painful papules which rapidly progressed to ulceration over few weeks, followed by multiple similar ulcerations developed over various sites. She was investigated with wound swab cultures, biopsy for histology and managed according to the positivity of culture for *staphylococcus aureus* and then for pseudomonas and lymphocytic cellular infiltrate in histology. Over 6 months all her lesions slowly healed.

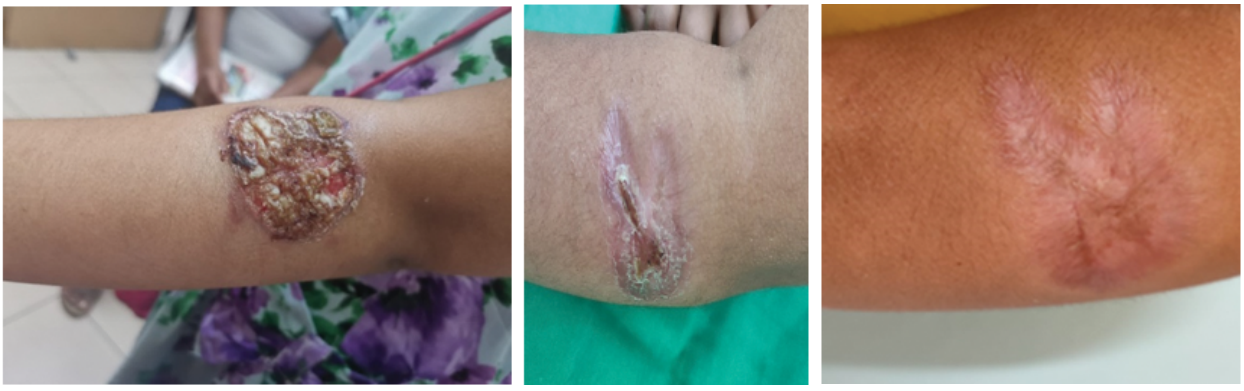

Figure 1. *Evolution of lesion with cigaretle paper scarring.*

[1]*Senior Registrar in Dermatology,* [2]*Consultant Dermatologist,* [3]*Medical Officer in Dermatology.*

After a year again presented with similar lesions and on examination the child had multiple ulceration with various stages of healing. Individual ulcer had undermined border with necrotic base contain yellowish slough and cigarette paper scarring at the site of healed lesion. The child was investigated again with multiple skin biopsy for histology and cultures were obtained. The histology came as suppurative necrosis and the cultures for fungal and mycobacterium were negative.

With the clinical findings and exclusion of other causes of recurrent painful ulcers, the diagnosis of PG was made and started on systemic corticosteroid. Azathioprine started as steroid sparing drug. She improved dramatically with no recurrences.

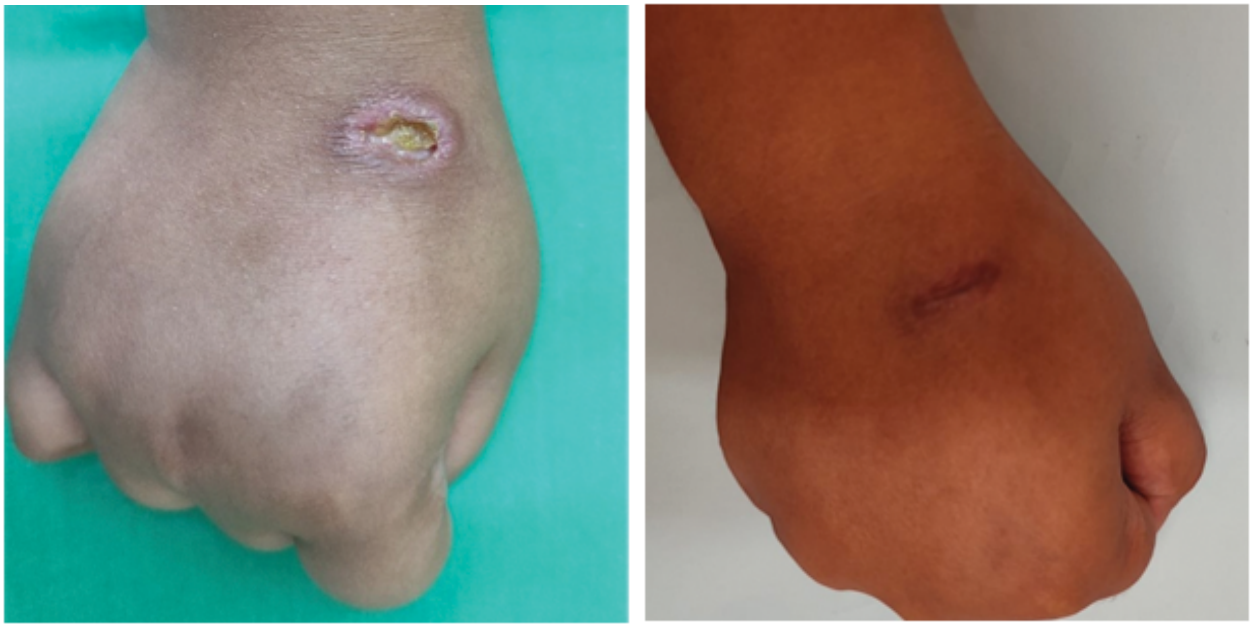

Figure 2. *Pathergy phenomenon following cannulation.*

Discussion

Diagnosis of PG is often made after failure of initial treatment and absence of gram stain on culture. Furthermore, the presence of secondary infections can obscure the diagnosis of this condition.

Diagnosis of PG requires both major criteria and at least two minor criteria. The major criteria include the characteristic appearance of a painful, irregular ulcer with a violaceous border, and exclusion of other causes of ulceration such as malignancy, vasculitis, primary infection, and drugs. The minor criteria include history of pathergy, associated systemic illness, histology findings of dermal neutrophilia, and response to steroid treatment.

Currently, there are no standard guidelines for the treatment of PG. In general, the treatment of PG involves both topical and systemic approaches. Topical therapy involves the use of dressings to control exudation, improve auto-debridement, protect the surrounding skin, and provide analgesia[3]. Topical tacrolimus has been shown to be effective in the treatment of less severe PG[4].

Systemic therapy is the mainstay of treatment for rapidly progressive PG. Corticosteroids such as prednisolone are initially used to prevent progression and arrest the inflammatory process. Combinations of steroid and cytotoxic drugs such as azathioprine, cyclophosphamide, cyclosporine, or mycophenolate mofetil are used in patients with steroid-resistant disease or as a steroid-sparing measure. In the presence of secondary infections, corticosteroids should be combined with antibiotics. Recently, tumor necrosis factor (TNF)-α blockers and other biologics have been used with some success.

Surgical debridement is contraindicated due to pathergy. Surgical treatments such as skin grafts may be worthwhile but can only be performed with

concomitant immunosuppression and in patients with stable disease or partial remission[5].

Despite the advances in medical therapy, the outlook of PG is unpredictable. Delayed diagnosis or misdiagnosis of PG will certainly lead to exacerbation and progression of the disease.

References

1. Bolognia j. n.d. *Dermatology* 4th ed. ; **1**: 459
2. Braun-Falco M, Kovnerystyy O, Lohse P, Ruzicka T. Pyoderma gangrenosum, acne, and suppuratives hidradenitis (PASH) - a new autoinflammatory syndrome distint from PAPA syndrome. *J Am Acad Dermatol*. 2012; **66**: 408-15.
3. Wollina U. Clinical management of pyoderma gangrenosum. *Am J Clin Dermatol*. 2002; **3**(3): 149-58.
4. Marzano AV, Trevisan V, Lazzari R, Crosti C. Topical tacrolimus for the treatment of localized, idiopathic, newly diagnosed pyoderma gangrenosum. *J Dermatolog Treat*. 2010; **21**(3): 140-3.
5. Fulbright RK, Wolf JE, Tschen JA. Pyoderma gangrenosum at surgery sites. *J Dermatol Surg Oncol*. 1985; **11**: 883-6.