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A Rare Case of Recurrent Pyoderma Gangrenosum

Willy Oematan[1], Syeben Hezer Epatah Hietingwati[2], Robertus Arian Datusanantyo[3]

[1]Department of Surgery, Prof. Dr. W. Z. Johannes General Hospital, Kupang, East Nusa Tenggara, Indonesia
[2]Pathologist, Department of Anatomical Pathology Prof. Dr. W. Z. Johannes General Hospital, Kupang, East Nusa Tenggara, Indonesia
[3]Plastic Surgeon, Department of Surgery Prof. Dr. W. Z. Johannes General Hospital, Kupang, East Nusa Tenggara, Indonesia

Corresponding Author: Willy Oematan

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ABSTRACT

Introduction: Pyoderma gangrenosum (PG) is a rare non-infectious disorder, and presents as solitary or multiple, fragile papules that progresses to ulcer and necrotic tissue. Autoimmune is the favored pathophysiology associated with this chronic inflammatory skin disease. Systemic disease such as inflammatory bowel disease, rheumatoid arthritis, and haematological disorders often accompany PG. We report a case of recurred PG in 60 year old male with no evidence of associated systemic condition.

Case Presentation: A 60 year-old male complained of 1 month-long painful ulceration on his left chest. The lesion started as a pustule which gradually increased in size and broke into painful ulcer. The patient had similar skin lesion, five and three years prior to this admission on the lower abdomen and lower limbs respectively. Both episodes healed more than a year. The diagnosis of ulcerative PG was established based on the physical and histopathological examination. The patient was treated with prednisone and moist wound care. The ulceration responded rapidly.

Discussion: Pyoderma gangrenosum is associated with underlying disease in up to two-thirds of case. It occurs typically in adults between 40 and 60 years of age. Although PG predilection is typically lower extremities, any body site can be affected, especially following a trauma. Our patient presented with recurred PG but showed no other systemic condition. Autoinflammatory disease are clinically characterized by recurrent episode and the treatment is quite challenging for its tendency to become chronic, relapsing, or reversible ulcer.
Conclusion. We report a rare case of recurred ulcerative PG with no symptom of associated systemic disease who responded rapidly with oral steroid treatment and moist wound care.

Keyword: Pyoderma gangrenosum, ulcerative, autoimmune, case report, steroid.

INTRODUCTION

Pyoderma gangrenosum (PG) is a rare non-infectious disorder, and presents as solitary or multiple, fragile papules that can progress to ulcers and necrotic tissue (1,2). It was described by Brunstring in 1930 as a rapidly spreading ulceration of soft tissue and the causing of this skin ulceration was believed to be a streptococcal infection. Currently, the pathophysiology of PG remains mostly uncertain, it has been defined that PG is not caused by bacteria. The favored PG pathophysiology is autoimmune with defect in cell-mediated immunity, neutrophil and monocyte function, and humoral immunity (3,4).

Pyodema gangrenosum occurs typically in adults between 40 and 60 years of age. It rarely affects children, in which case it is commonly associated with other systemic diseases. In large meta-analysis study including 2,611 patients the prevalence of immune-related systemic diseases in PG was 56.8% (2,4). Systemic diseases that

associated with PG such as inflammatory bowel disease (IBD) (17,6%), rheumatoid arthritis (12,8%), haematological disorders (8,9%), and solid tumors (7,4%) (5).

The incidence of PG is estimated at 10 cases per million people per year, and affect women more often than men. Based on the clinical manifestation, PG is classified into four subtypes: ulcerative PG, a painful ulcers evolve from tender pustules or papules; pustular PG, pustules with symmetric erythematous borders associated with IBD; bullous PG, rapidly evolving, painful bullae that can progresses to ulcer; and vegetative PG, a non-painful superficial ulcer generally without the classic purple edge. The best documented factor that can induce PG is trauma, the disease follows acute trauma or injury to the area (including surgery), in a process known as pathergy (2,4,5).

Making a PG diagnosis can be challenging. The diagnosis is clinical and requires the exclusion of other disorders in the differential diagnosis. International consensus diagnostic criteria for ulcerative PG assign four equal points categories: histology to exclude infection; history of pathergy, systemic disease, and pustule or vesicle that rapidly ulcerates; clinical or photographic evidence such as peripheral erythema, multiple ulcerations, and 'Wrinkled-paper" scar; and respond to treatment characterized by reduction in ulcer size within 1 month of immunosuppressive therapy (5,6).

Pyoderma gangrenosum management consist of local wound care with moist dressing to relieve pain, promote re-epithelialization and prevent trauma, topical therapy with potent topical glucocorticoids is useful in early ulcers or there are small isolated ulcers, and systemic therapy with high dose corticosteroids (3).

CASE PRESENTATION

A 60 year-old male complained of one month-long painful ulceration on his left chest. The lesion started as a pustule which gradually increased in size and broke into painful ulcer. The patient had similar skin lesion five and three years prior to this admission on the lower abdomen and lower limbs respectively. Both episodes healed more than a year.

The patient had no history of or complaints related to inflammatory bowel disease, arthritis, autoimmune disease and tumor.

Figure 1. A well-defined ulcer with irregular shape. Skin was necrotic whereas nipple aerola complex was normal.

On admission to our facility, his vital signs were within normal limit as well as afebrile. His laboratory work-up was unremarkable. On physical examination, we found a large painful ulcerated wound on the anterolateral aspect of his left chest. The wound was in irregular shape, partially covered with crust and necrotic tissue, wound margin induration, edema, granulation centrally, and hyperemia (figure 1). Nipple areola complex showed neither sign of inflammation nor necrosis. The final biopsy report supported clinical diagnosis of pyoderma gangrenosum (figure 2).

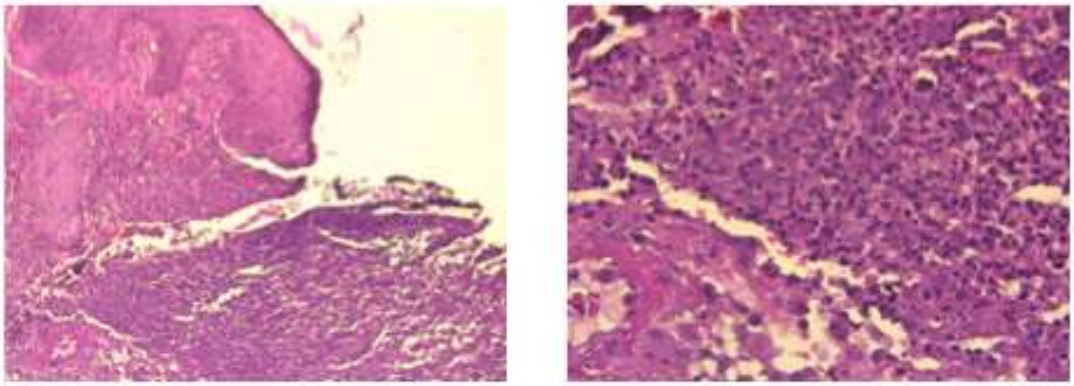

Figure 2. Histopathological findings showed eroded skin to ulceration, containing massive distribution of lymphocytes, histiocytes, neutrophils, and eosinophils; blood vessels that contain erythrocytes and multiple necrotic areas. No visible signs of malignancy.

The diagnosis of ulcerative PG was established based on the physical and histopathological examination. The patient was treated with oral high-dose prednisone and moist wound care. The ulceration responded rapidly (figure 3). Contralateral, there was similar lesion which healed faster despite conservative wound treatment. The healed lesion formed "paper-wrinkled" scar which is characteristic of PG (figure 4).

Figure 3. After 9 weeks, the lesions have resolved, granulation tissue is clearly visible and there are early signs of islands of re-epithelialisation.

DISCUSSION

Pyoderma gangrenosum present as solitary or multiple fragile papules that can rapidly progress to ulcer and necrosis. In up to two-third case, PG is associated with underlying disease. In this report, the clinical manifestations of the ulcers were characteristic of PG lesions. The patient had initial lesion in the form of pustules that enlarged and ruptured into tender ulcers with uneven edges and showed no sign and symptom of systemic condition. Although PG is commonly associated with underlying diseases, PG itself is a rare disease. It may be found as an isolated autoimmune disorders (5).

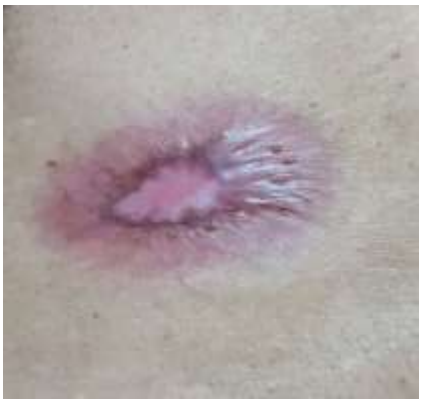

Figure 4. Paper wrinkled scar on patient right chest.

Trauma is the best documented factor that can induce PG ulceration. It is known to induce the cytokines that trigger innate immune responses in a process known as pathergy (2,5,6). While anybody site can be

affected, the lesions are often take place on the lower extremities (7). Autoinflammatory diseases such as PG are clinically characterized by recurrent episodes (2,5), similar with the case of our patient presented with a history of similar skin lesion five and three years prior to this admission on the lower abdomen and lower limbs.

There are no specific laboratory finding or pathognomonic to diagnose PG. The international consensus diagnostic criteria for ulcerative PG assign four equals point category that depends exclusively on the observation of clinical characteristics and disease progression, history, histopathological examination to exclude other etiologies of skin ulcers, and response to therapy (4,5). In this case the clinical manifestations of the ulcers were characteristic of PG lesions. The pathergy sign was negative, there was no history of underlying disease, but he had history of similar skin lesion that healed more than a year, the biopsy confirmed pyoderma gangrenosum, and ulceration responded rapidly to therapy.

Administration of systemic corticosteroids, wound treatment, and aseptic dressing is the most effective treatment of PG. The objective treatment for PG is to limit tissue destruction, promote wound healing and obtain a good esthetic result. Autoinflammatory diseases are clinically characterized by recurrent episodes. Therefore the treatment of PG is quite challenging for its tendency to become chronic, relapsing, or reversible ulcer (4,5). In this case, our patient was treated with systemic corticosteroids and moist wound treatment. The ulceration responded rapidly.

CONCLUSION

Pyoderma gangrenosum is a rare non-infection, autoimmune skin ulceration that can be challenging in diagnosis. We report a rare case of recurred ulcerative PG with no associated systemic disease who responded rapidly with oral steroid treatment and moist wound care.

Declaration by Authors

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