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Case Report

Multiple Disseminated Pyoderma Gangrenosum: Case Report

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Abstract: Pyoderma gangrenosum (PG), first described by Brocq and named by Brunsting *et al*., in 1930, is a rare, ulcerating, neutrophilic dermatosis affecting patients any age, with a peak between 40-60 years; diagnosis is based on its clinical morphological features. Approximately 50% of cases are associated with systemic disease. PG is frequently misdiagnosed, leading to a delay in the appropriate medication with corticosteroids.

Keywords: Pyoderma, gangrenosum, skin, ulceration, dermatology, dermatosis.

INTRODUCTION

Pyoderma gangrenosum (PG) is a rare neutrophilic dermatosis characterized by painful necrotic ulceration [1, 2] with an incidence of 3-10 cases per million per year [3] it can affect any age group, with a peak between 40 and 60 years, it has a slight female predominance [4]. The diagnosis is based on its clinical morphological features: ulcers have a purulent base and an overhanging gunmetal gray border [5]. If pyoderma gangrenosum is suspected, a punch biopsy specimen should be collected from the ulcer's edge and sent for pathological examination, another fragment should be sent for culture of bacteria, atypical mycobacteria and deep fungi [5]. Literature reports that 50% of cases are associated with systemic disease, most commonly inflammatory bowel disease, arthritis and hematologic disease, underlying systemic disease should always be sought [5, 6]. We present an 84-year-old without systemic disease, who presented with clinical features of multiple pyoderma gangrenosum.

CASE PRESENTATION

A 84-year old woman with chronic degenerative history of hypertension; she presents with a sixth- month ulcerated painful lesions on abdomen, anterior region of right shinbone and right foot. The first lesion appeared six months ago as pustules that grew forming ulcers with a purulent discharge; she was prescribed with silver sulfadiazine, however lesions continued to enlarge. She was referred to a dermatologist, the bacterial swab of the ulcer was positive to *Pseudomona aeruginosa* sensitive to ceftazidime and imipenem, she was prescribed a course of systemic antibiotics without improval. She presented to our service and received dermatological evaluation; On examination she presented with a disseminated dermatosis, asymmetrical, located in abdomen, shinbone and foot, characterized for ulcerative lesions, the largest of them with a diameter 22 x 15 cm, the smallest of them with a diameter of 3x2 cm, some oval, others round, non-confluent, with well-defined edges, a dirty bottom made up of granulation, fibrin and necrotic tissue. A biopsy was performed of the lesion in the abdomen that showed deep neutrophilic dermatitis, compatible with; pyoderma gangrenosum, classic variant. She received prednisone at an initial dose of 50 mg per day which was tapered every week and alibour; hematological disease was ruled out; endoscopy was normal. Antinuclear antibodies, extractable nuclear antigen antibodies, antineutrophil cytoplasmic antibodies, a QuantiFERON test, and serology for hepatitis, HIV, and syphilis were all negative. Lesions improved significantly with treatment and the patient was discharged.

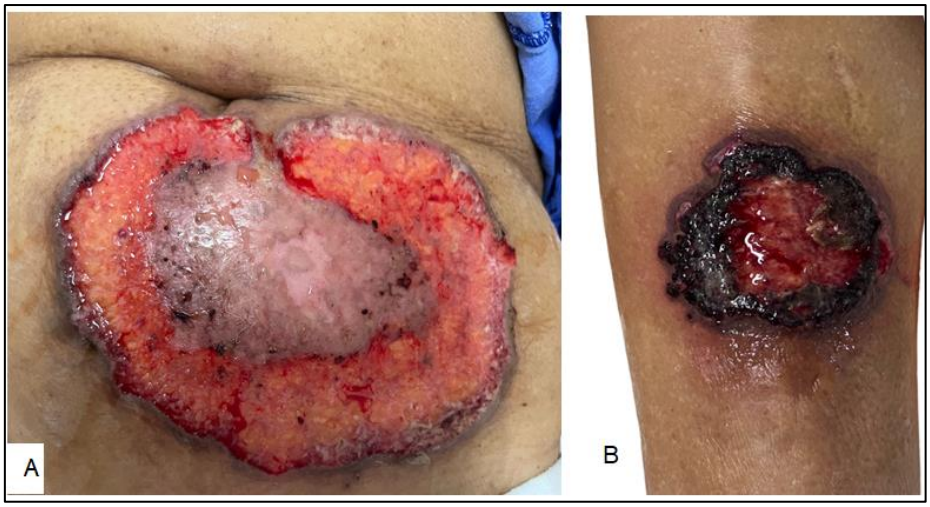

Figure 1: Disseminated initial lesions A) A large ulcerated lesion in abdomen with granulation tissue B) Ulcerated lesion in right anterior leg with necrotic border and granulation tissue

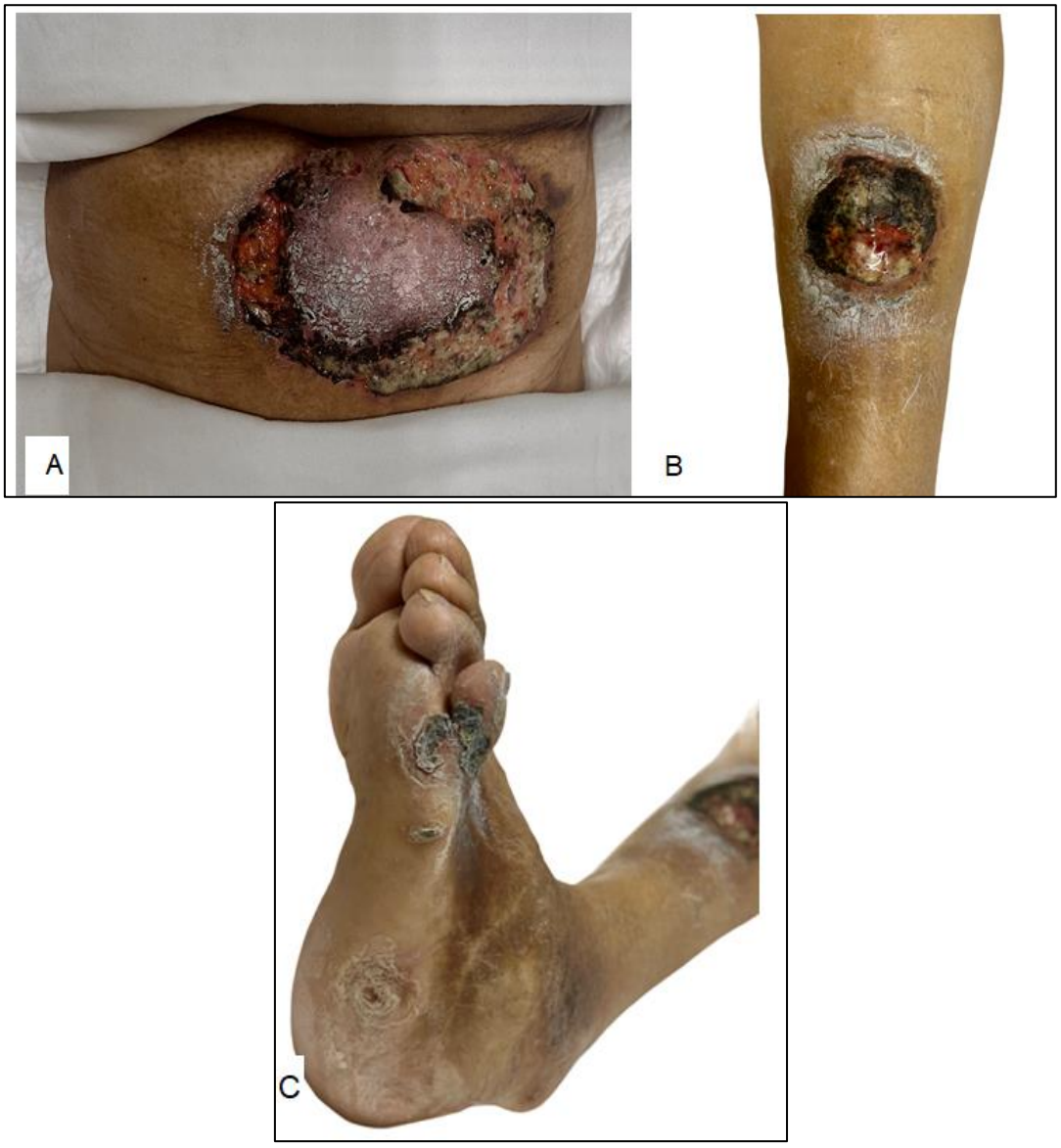

Figure 2: Lesions after one month of treatment A) Ulcerated lesion in abdomen with granulation and ample fibrin tissue B) Ulcerated lesion in right anterior leg C) Necrotic lesion in right foot

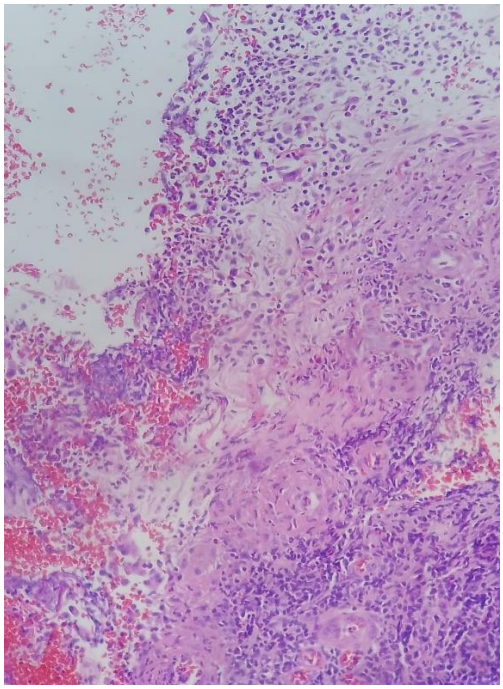

Figure 3: Skin biopsy of ulcerated abdominal lesion: Reticular dermis showing hemorrhage, ischemia and diffuse neutrophils consisting in neutrophilic dermatitis. HyE (20x)

DISCUSSION

PG is a reactive, noninfectious neutrophilic dermatosis [7] characterized by neutrophil-predominant infiltrates; the exact reason for development of inflammation remains unknown. Abnormalities in neutrophil function, genetic variations and dysregulation of the innate immune system are considered to be involved in the pathogenesis [2]. PG is characterized by a spectrum of clinical presentations with variable courses, usually starts as an inflammatory tender papule, pustule, vesicle or nodule that quickly enlarges and erodes to a painful ulcer with sharply marginated, undetermined, violaceous borders surrounded by erythema and a purulent necrotic base [2,7], patients usually refers important pain; lesions are often multiple and recurrent, they can occur or worsen at areas of constant trauma [2]. Various PG variants are known: ulcerative, bullous, pustular, vegetative, peristomal, post-surgical, the most common variant is ulcerative (classic) [2]. Each subtype has a common location and associated disease, the most common location are the lower extremities [8]. More than 50% of patients with PG have an associated systemic disease; the most common is inflammatory bowel disease, inflammatory arthritis, hematologic disorders and malignancies and other neoplasm, they alway should be considered and ruled out [2]. Maverakis *et al*., proposed new diagnostic criteria for ulcerative PG that required only the exclusion of infection and shows sensitivity and specificity of 86% and 90%, respectively [9]; one major criteria requires biopsy of ulcer edge demonstrating a neutrophilic infiltrate, and four of eight minor criteria including (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 one 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 [9,10]. PG is frequently misdiagnosed, leading to a delay in the appropriate medication with corticosteroids. Some cases do indeed need antibiotic therapy as there is an infection compromise. The use of systemic corticosteroids is considered to be the first-line treatment for PG, as oral prednisone (0.5–1 mg/kg/day) or intravenous corticosteroid (1000 mg/day) and signs of improvement can be seen within two-three days [11]. Additional measures depend on whether the disease is localized or extensive and can include wound care, topical treatments, immunosuppressants, and immuno-modulators [12]. PG lesions demonstrate pathergy, a phenomenon where skin trauma triggers the development of new lesions at the site of injury. Therefore, surgical debridement is contraindicated in the management of PG [13].

CONCLUSION

The diagnosis of PG remains a challenge, in the absence of a disease-specific laboratory or histologic finding; however, recently proposed clinical criteria are useful. Understanding the histopathological findings, the role of neutrophils in tissue destruction and the potential for pathergy aids in the comprehensive management of this challenging condition. Approximately half of all cases are associated with a systemic disease, justifying a thorough search for the most common associated conditions in any patient with a new presentation of PG. There is no gold standard for treatment. The central goals of management are to control inflammation and optimize wound healing.

Conflict of Interest: The authors declare that there are no conflicts of interest at the time of publication of this article.

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