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THE DIAGNOSTIC ENIGMA OF PYODERMA GANGRENOSUM-A CASE SERIES

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Abstract

Introduction

Pyoderma gangrenosum (PG) is a rare chronic inflammatory disease with an unknown etiology and characterized by neutrophilic infiltration of the skin.[1,2] The term of PG was first introduced by Brunsting et al in 1930.[3]

Based on the Mayo criteria, diagnosis of PG is established if two major and two minor criteria were fulfilled.[4] The major criteria include 1) rapid progression of painful necrotic ulcer, with irregular, violaceous, and undermined edges, 2) elimination of other etiologies of skin ulcers. The minor criteria include 1) history of pathergy or clinical finding of cribriform scarring, 2) PG-related systemic disease, 3) appropriate histopathology finding, and 4) rapid response to systemic corticosteroid administration.[4,9,11] The incidence is estimated at around 3–10 cases per million population per year.[5,6] It tends to affect women more commonly than men,with a median age of 59 years.[7]

Based on the clinical manifestation, PG is classified into five subtypes: peristomal, pustular, bullous, vegetative, or classic (ulcerative).[4,5]

Based on one of the largest case series of PG, 77.7% of the lesions appeared on the leg, followed by the trunk (11.7%), peristomal site, and the upper extremities (8.7% each), the head and neck (7.8%)[8]Hence lower extremity is the most common predilection site for PG.[1,6] and other sites are trunk, upper extremities, head and neck.[7]

Approximately 50% of the PG cases are accompanied by systemic diseases such as inflammatory bowel disease, arthritis, solid malignancies, hematological malignancies, HIV, and rheumatological disorders.[5,13,14]

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Historically, IBD was reported to be the most common associated disease[5] (32%).[6]

Recently, focus has been shifted on detecting underlying endocrine disorders as several studies have found a potential link with diabetes mellitus, thyroid disorders and metabolic syndrome. A multi-centric analysis conducted in 259 patients in Germany revealed that patients of pyodermagangrenosum had concomitant anemia in 45.6% of the patient population, 44.8% had endocrine disease, 12.4% had internal malignancies, and 9.3% had chronic inflammatory bowel disease. 25.5% of the patients had diabetes mellitus with some aspects of metabolic syndrome.[7] Another study found endocrine disease in 38.8% of the patients, diabetes in 28.6% of the patients and obesity in 32.6%.[8] An association of anti-thyoid drug propylthiouracil has been described in literature.[9]

PG can demonstrate pathergy (lesions develop at site of trauma).

PG is an inflammatory neutrophilic dermatosis that comes under the larger umbrella of an autoinflammatory disease. Autoinflammatory conditions present with recurrent attacks of non-infectious inflammation at target sites without associated high levels of circulating autoantibodies or autoreactive T cells. PG immunohistochemistry studies have shown neutrophil recruitment at the lesion bed and T-cell-mediated and macrophage-mediated ulcer formation at wound edges.[3] [6]

Initial PG lesions can be in the form painful pustules, nodules, or plaques.[1,2] Within a few days, the skin lesions may enlarge and rupture to form well-defined ulcers, with irregular violaceous edges.[9,10] The skin and subcutaneous tissue subsequently become necrotic, with a brittle wound bed covered in hemorrhagic or purulent exudative fluid. The ulcers are accompanied by increased severity of pain.[9] Re-epithelialization occurs from the ulcer edge, leaving the cribriform atrophic scars.[4,11]

It is not uncommon for the diagnosis of PG to be delayed or missed, which can lead to inappropriate therapy such as surgical debridement.[5]

It is also important to monitor for signs of superimposed infection.

Treatment for PG can involve a combination of wound care, topical therapy and systemic therapy.

Wound care is essential for all lesions and is tailored to the type of lesion and extent of associated exudate. A moist environment is typically recommended to promote wound healing.

Topical treatment that have been tried with some success include corticosteroids, tacrolimus, ciclosporin, 5-aminosalicylic acid and dapsone.[5]

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More extensive disease is usually initiated with systemic steroids. Cytotoxic agents, most commonly ciclosporin, and others such as azathioprine can also be used. Cyclophosphamide, mycophenolate and tacrolimus have been used as well. Systemic dapsone has also been used in PG with good results. Recently biological therapy, specifically infliximab, for PG in the context of IBD, has shown very good results.[4] [5]

Case report 1

A 18-year-old female presented with a 3 month history of painful ulcerations on her right thigh. The lesions initially appeared as small pustules that rapidly progressed to deep, necrotic ulcers with purulent discharge. The patient was diagnosed as necrotizing fascities for which repeated debridments were done but still showed no signs of healing.

Clinical Examination:On examination, solitary ulcers measuring 12*15cm with irregular borders, undermined edges, and a violaceous border with purulent fowl smelling discharge due to secondary bacterial infection was noted on right tight extending from groin to mid thigh. The patient experienced severe pain, and the lesions were surrounded by an area of erythema. Associated with fever .

Investigations:Laboratory investigations revealed an elevated white blood cell count and inflammatory markers. Wound cultures were positive for bacterial, and negative for viral, and fungal infections. Histopathological examination of a biopsy from the ulcer edge demonstrated a dense neutrophilic infiltrate without evidence of infection or vasculitis, supporting the diagnosis of pyoderma gangrenosum.

Management: Topical wound care, with regular dressing changes and vaccum therapy, was employed to promote ulcer healing. A multidisciplinary team involving dermatologists, nutitionist and wound care specialists collaborated to manage the patient .

Outcome:due to delay in diagnosis as pyoderma gangreosum,patient succumbed to death due to MODS which set in as a result of secondary bacterial infection.

Case report 2

A 48-year-old female presented with a 15 days history of painful ulcerations on her left thigh. The lesions initially appeared as small wound with crustaceans that rapidly progressed to large necrotic ulcers with purulent discharge. She ia a known diabetic since 5 years but not compliant on medications.

Clinical Examination:On examination, solitary ulcers measuring 22*15cm with irregular borders, undermined edges, and a violaceous border with purulent discharge was noted on left tight posteriorly extending to medial aspect also. The patient experienced severe pain, and the lesions were surrounded by an area of erythema. Systemic symptoms such as fever and malaise were absent.

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Investigations:Laboratory investigations revealed an elevated white blood cell count and inflammatory markers with high blood sugars. Wound cultures were positive for bacterial, and negative for viral, and fungal infections. Histopathological examination of a biopsy from the ulcer edge demonstrated a dense neutrophilic infiltrate without evidence of infection or vasculitis, supporting the diagnosis of pyoderma gangrenosum.

Management: As the patient had uncontrolled diabetis she was referred to physician for further evaluation and optimization. Immunosuppressive therapy, including systemic corticosteroids and azathioprine, was initiated to control the inflammatory response. Topical wound care, with regular dressing changes and compression therapy, was employed to promote ulcer healing. A multidisciplinary team involving dermatologists, physicians,surgeons managed the patient effectively.

Outcome: With the combined approach of systemic immunosuppression, optimized diabetes management, and meticulous wound care, the patient showed significant improvement within six weeks. The ulcers began to heal split skin graft was done to cover the large raw area. and the patient's pain decreased substantially. Regular follow-up visits were scheduled to monitor the response to treatment and adjust therapy as needed.

Case report 3

A 25-year-old male presented with a six-week history of painful ulcerations on his lower extremities. The lesions initially appeared as blebs that eventually ruptured with seropurulent discharge for which he was taking ayurvedic treatment but it progressed to large ulcer with fowl smelling greenish purulent discharge. The patient had no co-morbidities.

Clinical Examination:On examination, a large ulcer with irregular borders, undermined edges, and with necrotic scab was found over the anterior aspect of left leg extending from knee to mid leg.surrounding are showed local rise of temperature with erythema. The patient experienced severe pain and was not able to walk properly due to contracture as result of immobilisation. No systemic symptoms such as fever and malaise

Investigations:Laboratory investigations revealed an elevated white blood cell count and inflammatory markers. Wound cultures were negative for bacterial, viral, and fungal infections. Histopathological examination of a biopsy from the ulcer edge demonstrated a dense neutrophilic infiltrate without evidence of infection or vasculitis, supporting the diagnosis of pyoderma gangrenosum.

Management:Immunosuppressive therapy, including systemic corticosteroids and azathioprine, was initiated to control the inflammatory response. Topical wound care, with regular dressing changes and negative pressure wound therapy, was employed to promote ulcer healing. A multidisciplinary team involving dermatologists,physiotherapist, surgeons and plastic surgeon helped in patient management

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Outcome:following systemic immunosuppressive therapy and meticulous wound care, the patient showed significant improvementin 4 weeks. As the ulcers began to heal,split skin graft was done .physiotherapy seesions were given to improve his gait.

Conclusion

Pyoderma gangrenosum poses a diagnostic challenge due to its variable presentation and lack of specific laboratory markers.

All our three cases presented to us with non healing ulcers over period 4 weeks and there was delay in initiating the appropriate immunosuppressive therapy as timely diagnosis of pyoderma gangreosum could not be done.

All our patients were treated with repeated wound debridment which made the PG more bad and increased the ulcer surface area.

Hence when in doubt about a non specific,non healing ulcer its better to keep PG as one of the differential in mind. Timely diagnosis with skin biopsy and initiation of immunosuppressive therapy with meticulous wound care can reduce the morbidity and mortality.

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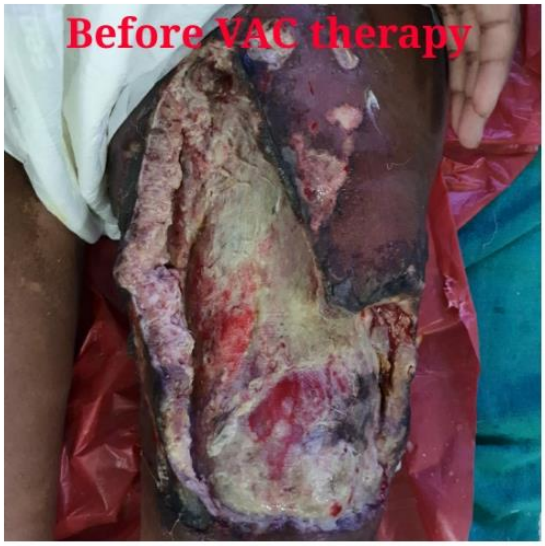

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