

Case report

Bullous Pyoderma Gangrenosum

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ABSTRACT

Pyoderma gangrenosum (PG) is a rare neutrophilic dermatosis characterized by painful necrotic ulcerations. It affects patients between the age of 40 and 60 years. Clinically, it presents as a very painful papulopustule or bulla which rapidly progresses to form deep necrotic ulcers. Hematologic malignancies are commonly linked with bullous PG. We report a case of bullous PG who presented with multiple haemorrhagic blisters without any systemic associations.

Keywords: Dermatitis gangrenosa; Phagedenic pyoderma; Bullous PG; Neutrophilic dermatosis; PAPA syndrome

INTRODUCTION

Pyoderma gangrenosum (PG) is a sterile neutrophilic dermatosis of unidentified etiology affecting patients between 40 to 60 years (1). Women are more commonly affected than men. Pyoderma gangrenosum (dermatitis gangrenosa, phagedenic pyoderma; 2) was first described by Brocq in 1916 (3). Pain is often excruciating and out of proportion to the size of the ulcer. It is frequently associated with other systemic diseases like inflammatory bowel disease (IBD), rheumatological disorders, haematological abnormalities, connective tissue disorders. PG is often a clinical diagnosis. There is no specific HPE or laboratory findings to confirm the diagnosis.

CASE REPORT

47-year-old male patient presented to our out-patient department with complaints of fever and multiple dark coloured fluid filled lesions on both hands for 3 days. Lesions initially started on the right palm as papulopustule which rapidly spread to involve the left

palm, dorsal aspect of both hands and finger tips and were associated with severe pain. Few lesions burst on its own leaving behind erosions. There was no history of trauma, no history of any topical application, and no history of any drug intake. There was no history of joint pain, abdominal pain, diarrhoea, loss of appetite, weight loss. There was no similar complaints in the family.

Dermatological examination revealed multiple discrete well-defined tender tense haemorrhagic bullae on the palmar aspect of both hands. A single bulla was noted on the dorsal aspect of either hands. Few tender necrotic shallow ulcers of varying size with surrounding violaceous induration and crusting noted on the palmar aspect of both hands. Dactylitis of fingers was noted over both hands. No similar lesions were noted elsewhere in the body. Examination of soles, Oral mucosa, genital mucosa, scalp and nails were normal. Systemic examination was found to be normal. Differential diagnosis of bullous PG, bullous FDE, bullous contact dermatitis, erythema multiforme were made.



Fig. 1: (A) multiple bullae and necrotic ulcer and fingers showing dactylitis. (B) bulla on the dorsal aspect



Fig. 2: Resolved lesions on palmar and dorsal aspect after treatment. Skin Biopsy

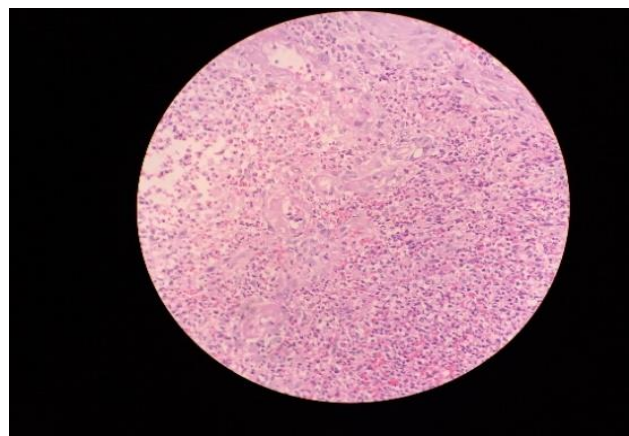
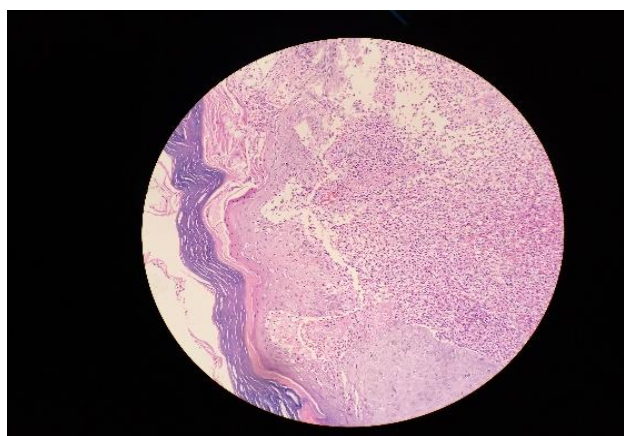


Fig. 3: Section showing hyperkeratosis, parakeratosis, exocytosis with marked inflammation

Baseline investigations like CBC, RBS, RFT, LFT, urine analysis, chest x ray, ECG, peripheral smear were done. Total counts, ANC and ESR were found to be elevated. Gram stain from the wound revealed few neutrophils with no microorganisms. Blood and skin cultures were negative.

As patient did not have any clinical symptoms like bone pain, recurrent infections, frequent fractures, loss of weight, loss of appetite and as laboratory investigations revealed absence of anemia, hypercalcemia, normal creatinine, normal albumin levels, no rouleaux formation in peripheral smear, serum protein electrophoresis was deferred.

Skin biopsy revealed stratified squamous epithelium with hyperkeratosis, parakeratosis, exocytosis and ulceration covered by inflammatory exudate. The dense inflammatory exudate extends to the deeper region with focal micro abscesses. Based on clinical presentation, diagnosis of pyoderma gangrenosum was made.

Patient was started on IV steroids, IV antibiotics to prevent secondary infection and analgesics along with local wound care. Patient showed rapid response to steroids within 3 days. IV Steroids was changed to oral steroids which was tapered slowly and stopped. Complete remission was obtained within 2 months and patient continued to be under remission during the follow up period of 4 months.

DISCUSSION

Pyoderma gangrenosum (PG) is a rare idiopathic sterile neutrophilic skin dermatosis characterized by an ulcer which is extremely painful. Lesions typically begin as tender papulopustules, nodules or plaques that rapidly evolved into shallow or deep ulcers with violaceous, undermined borders with surrounding zone of erythema. Pathergy is commonly observed in PG (4). It heals with atrophic or cribriform pigmented scars. The common sites of occurrence are lower extremities, buttocks and perineal region (5). In our case patient presented with palmar involvement which is a rare site of occurrence. Extra cutaneous sites include lungs, musculoskeletal system, spleen, eye (6-8). PG is often associated with an underlying systemic disease like IBD, hematological malignancies, rheumatologic disease.

Based on clinical morphology, there are 4 distinct variants of PG. They are classical or ulcerative type, pustular PG, bullous or pemphigoid PG and vegetative PG. Ulcerative variant is the most common type. Bullous PG is a rare variant often associated with hematological abnormalities.

Bullous PG was first described by Perry and Winkelmann in 1972 (2). It is characterized by the presence of rapidly evolving very painful superficial blisters and bulla with central necrosis, erosions and ulcerations. Upper body mainly face and upper

extremities are the commonly affected sites. This type of PG is most frequently associated with hematological malignancies such as AML, CML, myelodysplastic syndrome, monoclonal gammopathy(9-11). Among hematologic malignancies, AML is the most common type. Prognosis is poor when bullous PG is associated with leukemia. Few cases were found to have no systemic associations.

Pyoderma Gangrenosum is associated with few syndromes like PASH syndrome (Pyoderma gangrenosum, Acne, Suppurative Hidradenitis), PAPA syndrome (Pyogenic Arthritis, Pyoderma gangrenosum, Acne), PA-PASH syndrome (Pyogenic arthritis, Pyoderma gangrenosum, Acne, Suppurative Hidradenitis; 12). Differential diagnosis includes atypical Sweet syndrome, Behcets disease, vasoocclusive disorders, vasculitis, malignancy, infections, exogenous tissue injury, drugs, treponemal ulcer. PG is often misdiagnosed as an infection due to the lack of diagnostic laboratory test and histopathologic finding leading to delayed and inappropriate treatment. Other causes of ulcers should be ruled out before making a diagnosis of PG.

Treatment depends on extent, spread, associated diseases and response to the given treatment. Immunosuppression, infection and pain control forms the basis of treatment. It mainly consists of local wound care, topical therapy, systemic corticosteroids (oral/pulse IV), systemic agents (dapsone, clofazamine, colchicine, pentoxifylline, methotexate, MMF), immunosuppressants. Systemic corticosteroids form the first line of therapy. Surgical interventions should be avoided in PG due to pathergy phenomenon.

CONCLUSION

Bullous PG is a not so common variant of PG. Haematologic malignancies should be excluded in bullous type of PG. In our case patient showed good response to steroids and there was no evidence of systemic associations or hematological malignancies.

CONFLICT OF INTEREST

Authors declare no conflicts of interest.

REFERENCES

1. Bennett, M. L., Jackson, J. M., Jorizzo, J. L., Fleischer, Jr A. B., White, W. L., Callen, J. P. Pyoderma gangrenosum. A comparison of typical and atypical forms with an emphasis on time to remission. Case review of 86 patients from 2 institutions. *Medicine*. 2000 Jan 1; 79(1): 37-46.
2. Bhat, R. M. Pyoderma gangrenosum: An update. *Indian dermatology online journal*. 2012 Jan; 3(1): 7.
3. Brocq, L. Nouvelle contribution a l'etude du phagedenisme geometrique. *Ann Dermatol Syphiligr*. 1916; 6(1): 1-39.
4. Powell, F. C., Schroeter, A. L., Su, W. P., Perry, H. O. Pyoderma gangrenosum: a review of 86 patients. *QJM: An International Journal of Medicine*. 1985 May 1; 55(2): 173-186.
5. Powell, F. C., Perry, H. O. Pyoderma gangrenosum in childhood. *Arch Dermatol* 1984 Jun; 120 (6): 757-761.
6. Miserocchi, E., Modorati, G., Foster, C. S., Brancato, R. Ocular and extracutaneous involvement in pyoderma gangrenosum. *Ophthalmology*. 2002 Oct 1; 109(10): 1941-1943.
7. Krüger, S., Breuer, C., Schwarz, E. R., Piroth, W., Amotaki, B. Multiple aseptic pulmonary nodules with central necrosis in association with pyoderma gangrenosum. *Chest*. 2001 Mar 1; 119(3): 977-978.
8. Vignon-Pennamen. The extracutaneous involvement in the neutrophilic dermatoses. *Clinics in dermatology*. 2000 May 1; 18(3): 339-347.
9. Su, W. D., Davis, M. D., Weenig, R. H., Powell, F. C., Perry, H. O. Pyoderma gangrenosum: clinicopathologic correlation and proposed diagnostic criteria. *International journal of dermatology*. 2004 Nov; 43(11): 790-800.
10. Lois, M., Pizzariello, G., Olivares, L., Maronna, E. Estudio retrospectivo de pacientes con pioderma gangrenoso en un período de 20 años y revisión de la literatura. *Dermatología Argentina*. 2012 May 29; 18(2): 24-29.
11. Sakiyama, M., Kobayashi, T., Nagata, Y., Fujimoto, N., Satoh, T., Tajima, S. Bullous pyoderma gangrenosum: A case report and review of the published work. *The Journal of dermatology*. 2012 Dec; 39(12): 1010-1015.
12. Dierselhuys, M. P., Frenkel, J., Wulffraat, N. M., Boelens, J. J. Anakinra for flares of pyogenic arthritis in PAPA syndrome. *Rheumatology*. 2005; 44(3): 406-408.
13. Maverakis, E., Ma, C., Shinkai, K., Fiorentino, D., Callen, J. P., Wollina, U., *et al.*, Diagnostic criteria of ulcerative pyoderma gangrenosum: a Delphi consensus of international experts. *JAMA dermatology*. 2018 Apr 1; 154(4): 461-466.