Pyoderma gangrenosum - a rare skin disease

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Abstract

Pyoderma gangrenosum (PG) is an idiopathic, inflammatory, ulcerative disease of unknown etiology. Both topical and systemic corticosteroids are commonly used for the ulcer of PG, but these ulcers are often intractable despite treatment.

A 26-years old man presented with multiple painful ulcerated lesions on both legs which was appeared spontaneously 6 months back and he was treated with dapsone and prednisolone for this problem.

Re-epithelization and healthy granulation tissue developed with this treatment which indicates dapsone with systemic steroid is one of the useful way of treatment of ulcers in a patient with PG.

Key word : Pyoderma gangrenosum.

Introduction

First described in 1930, pyoderma gangrenosum is a non-infectious neutrophilic dermatosis[1]. This condition has an idiopathic form and it may associated with some underlying diseases such as inflammatory bowel disease, arthritis, haematological disease, human immunodeficiency syndromes and solid tumours[2,3]. There is also an idiosyncratic form that can be triggered by certain drugs[4].

The classic skin lesion of pyoderma gangrenosum usually begins with folliculocentric pustules or fluctuant nodules with an inflammatory halo, and expands peripherally to form an ulcer with sharply circumscribed violaceous raised edges[1]. Lesions typically affect the lower extremities and the trunk[4]. The varied appearance of these ulcers has led to their recent clinical classification, which includes four prototypic forms of pyoderma gangrenosum:

- ulcerative
- pustular
- bullous and
- vegetative.

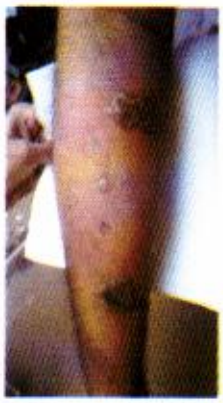
Only the vegetative form has no common association with underlying systemic disease. Each form may turn into another form and become ulcerative[4,5]. The diagnosis does not depend on histological biopsy findings and is especially challenging in its initial clinical form. A clinico pathological approach is required to make the diagnosis and to exclude other ulcerative processes causing dermal neutrophilia [4].

Usually the first line of treatment of pyoderma gangrenosum is systemic corticosteroid with treatment of underlying cause[5]. Many therapeutic approaches have been reported with inconsistent results[5].

Case report

A 26-years old male came to the out patient department of BSMMU with multiple painful ulcers on both legs for last 6 months. Initially he noticed a pustular lesion on left lower leg. Gradually he developed ulcer which is sharply circumscribed, undermined and painful. In course of time he developed multiple similar types of lesions on both legs.

Examination of the skin revealed multiple well defined rounded and oval ulcers covered with thick crusting. On palpation ulcers are tender. Pathergy test appeared positive.



Multiple ulcers covered with thick crusting and rolled border on the lower leg of the patient

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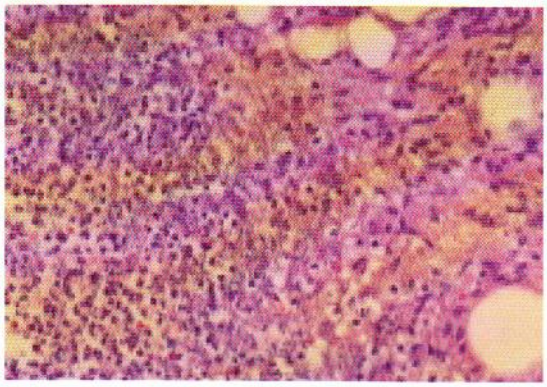
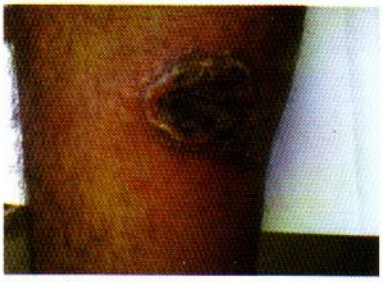
As pyoderma gangrenosum can be associated with HIV infection, an HIV test was performed and found to be negative. Other diagnostic tests such as chest X-ray, abdominal ultrasound and a double contrast barium enema, performed in order to exclude any underling diseases, revealed no abnormalities.

The lesional skin biopsy demonstrated a dense diffuse neutrophilic infiltrate with a mixture of lymphocytes, plasma cells, histiocytes and occasional foreign body giant cells that extended to the subcutis.

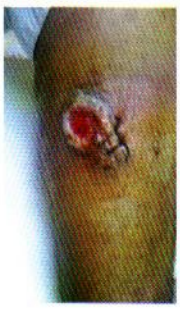


Histopathology (lesional skin biopsy) the dermis shows dense infiltrate of acute and chronic inflammatory cells, fibrosis and features of vasculitis

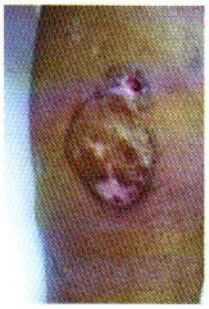
Then we started treatment with sysemic steroid and Dapsone. Gradually the patient improved and completely cured within 3 months.



A well defined ulcer covered with thick crust and prominent rolled border on lower leg



Same ulcer with red granulation tissue



Same ulcer with atrophic and dyspigmented plaque indicate healed ulcer after 6 weeks therapy with prednisolone and dapsone

Discussion

Since its first description in1930 the aetiology of pyoderma gangrenosum has remained obscure[1] Pyoderma gangrenosum is a rare, chronic, destructive, ulcerating skin disorder of unknown etiology. Complete and sustained resolution of the lesion is known to occur when the associated systemic disease is treated and cured. Many hypotheses have been proposed, but attention has focused principally on immune abnormalities and alterations in cell-mediated immune response[6]. It is thought that pyoderma gangrenosum may be the result of a hyperergic (or hypersensitive) reaction of the immune system due to an altered, exaggerated and uncontrolled inflammatory response to specific and non-specific stimuli, leading to a neutrophilic vasculitis, characterized by perivascular deposition of immunoreactants, mainly immunoglobulin M (IgM), C3 and fibrin, with direct immunofluorescence. Neutrophils appear to play a key role in the pathogenesis of pyoderma gangrenosum. This is evidenced by the fact that the disease responds to therapies that have antineutrophilic activity. In some patients defective cell-mediated immunity has been identified, including: defective leukocyte adhesion glycoproteins; defective neutrophilic chemotaxis and intracellular killing of microbial pathogens; selective anergy (immune unresponsiveness) to bacterial or fungal antigens; and E hypergammaglobulinaemia. In 40 to 50 percent of cases, pyoderma gangrenosum occurs in patients with no known associated systemic disease, and its occurrence is assumed to be sporadic.[7,8]

When pyoderma gangrenosum is associated with systemic disease, the therapeutic approach should also address the underlying disorder[4,9]. Treatment of lesions usually involves systemic treatment, together with local therapies[5,9]. Systemic treatments include steroids such as prednisolone 40-120mg/day until healing[10,11,6] Although prednisolone is a first-line treatment, it is not consistent in treating the

condition successfully and sometimes the high doses of prednisoloe is required which is associated with significant side effects. Therefoe, other drugs such as- Cyclosporine, Dapsone may use to minimize the side effects of steroid. Cyclosporine (6mg/kg) is a common therapy used either alone or in combination with steroids[12,13]. Systemic antibiotics have also been used, including rifampin, tetracycline, vancomycin and mezocillin[6,7]. Thalidomide has also been shown to be effective, especially in genital pyoderma gangrenosum[14].

We manage this patient successfully with Dapsone and Prednisolone therapy for three months.

References

1. Su WP, Davis MD, Weenig RH, Powell FC, Perry HO. Pyoderma gangrenosum: clinicopathologic correlation and proposed diagnostic criteria. Int J Dermatol 2004; 43(11): 790-800.
2. Powell FC, Schroeter AL, Su WP, Perry HO. Pyoderma gangrenosum: a review of 86 patients. Q J Med 1985; 55(217): 173-86.
3. Rafael MR, Fernandes CM, Machado JM, Rodrigues PA, Cardoso OJ, Afonso A, et al. Pyoderma gangrenosum or leukaemia cutis? J Eur Acad Dermatol Venereol 2003; 17(4): 449-51.
4. Crowson AN, Mihm MC, Magro C. Pyoderma gangrenosum: a review. J Cutan Pathol 2003; 30(2): 97-107.
5. Reichrath J, Bens G, Bonowitz A, Tilgen W. Treatment recommendations for pyoderma gangrenosum: an evidence-based review of the literature based on more than 350 patients. J Am Acad Dermatol 2005; 53(2): 273-83.
6. Wines N, Wines M, Ryman W. Understanding pyoderma gangrenosum: a review. MedGenMed 2001; 3(3): 6.
7. Fuenfer MM, Olson GE, Polk HC Jr. Effects of various corticosteroids upon the phagocytic bactericidal activity of neutrophils. Surgery 1975;78(1):27-33.
8. Kucan JO, Robson MC. Influence of topical steroids on bacterial proliferation in the burn wound. J Surg Research 1978;24(2):79-82.
9. Powell FC, Su WP, Perry HO. Pyoderma gangrenosum: classification and management. J Am Acad Dermatol 1996; 34(3): 395-409; quiz 410-2.
10. Barriere H, Litoux P, Stalder JF, Berger M, Delahaye M. Pyoderma gangrenosum associated with congenital hypogammaglobulinemia. Ann Dermatol Venereol 1979; 106(8-9): 695-6.
11. Caproni M, Pestelli E, Fabbri P. Le immunoglobuline endovenose in dermatologia. G Ital Dermatol Venereol 2003; 138: 377-83.
12. Elgart G, Stover P, Larson K, Sutter C, Scheibner S, Davis B, et al. Treatment of pyoderma gangrenosum with cyclosporine: results in seven patients. J Am Acad Dermatol 1991; 24(1): 83-6.
13. Fradin MS, Ellis CN, Voorhees JJ. Management of patients and side effects during cyclosporine therapy for cutaneous disorders. J Am Acad Dermatol 1990; 23(6 Pt 2): 1265-73; discussion 1273-5.
14. Faver IR, Guerra SG, Su WP, el-Azhary R. Thalidomide for dermatology: a review of clinical uses and adverse effects. Int J Dermatol 2005; 44(1): 61-7.