



An Ulcer That Hurts and the Hurt That Ulcerates: Pyoderma Gangrenosum- Case Report

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ABSTRACT

Pyoderma gangrenosum is an inflammatory disease of unknown etiology, commonly mistaken for an infection. Here we present a case of 50 year old female with painful ulcerative lesions with undermined edges and violaceous borders, with positive pathergy, diagnosed as idiopathic pyoderma gangrenosum after excluding associated systemic diseases.

INTRODUCTION

Pyoderma gangrenosum is a rare, non-infectious neutrophilic dermatosis which have a predisposition to pathergy. Incidence is 0.63 per 1,00,000 population ,occurs between the ages of 20 to 50 years with a female preponderance. It may be idiopathic (50%), or associated with para inflammatory (IBD, collagen vascular diseases, arthritis), paraneoplastic (associated with malignancy), hemotologic (leukemias, polycythemia), drugs , hepatitis C infection and primary biliary cirrhosis. Clinically it is classified into ulcerative, pustular, bullous and vegetative types. Diagnosis is based on typical clinical features and the exclusion of other cutaneous ulcerating diseases.

CASE REPORT

A 50 year old housewife with no significant past history or family history presented with

polymorphic vesiculo-bullous eruptions over legs, thighs, torso, breast and sub-mammary areas, gluteal region. It started as small pin head size eruptions over the neck, followed by a lesion in upper dorsal region and descended down. These papular eruptions became vesicular, later necrotic and ulcerated over next 2 days and enlarged rapidly, became deep and punched out in appearance. She remembers that whenever she tried to squeeze the vesicles, there appeared crops of vesicles around the original one and the whole area became necrotic and started to ulcerate very rapidly. She also complained of severe pain and minimal continuous bleeding from the base of ulcers. Lesions spared face, palms, soles, mucosal membranes and flexures. No h/o recent infections/drug intake/malignancy. No bowel and bladder symptoms.

On examination, her vitals were normal. General examination showed vesicles of varying ages, 0.5* 0.5*0.5 cm, multiple punched out ulcers, smallest one measuring 2*2 cm and largest one measuring 5*5 cm. Ulcers were mainly located over lower legs and torso, mainly over gluteal region and mammary and sub-mammary area. Systemic examination found to be normal.

Investigations revealed normal routine blood examination except for an ESR of 115 mmHg. LFT, RFT, lipid profile, TFT, blood sugar, serum electrolytes, urine routine were normal. Viral markers-Negative. Pus Culture was sterile. Blood Culture -Negative. ANA, RA factor, ANA profile -Negative. X ray chest/ ECG/USG abdomen-Normal. CT chest and abdomen-Normal. Colonoscopy and OGD scopy-Normal.

Biopsy of the ulcerative lesion was done. The histopathological report showed mixed inflammation with predominant neutrophils, along with lymphocytes and plasma cells, suggestive of pyoderma gangrenosum. Culture of the biopsy specimen-sterile.

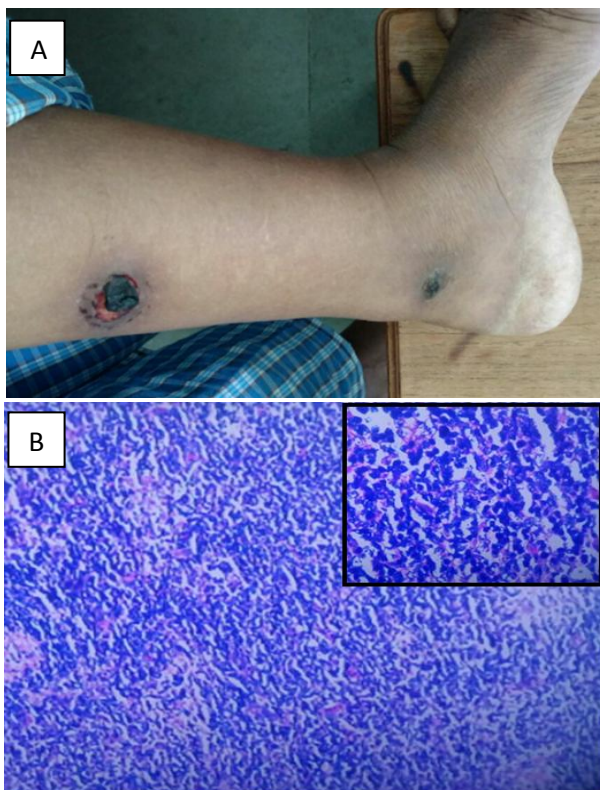


Fig.1 A, necrotic cutaneous ulcer over left leg B, Biopsy: H&E low power - mixed inflammation; (inset) high power -predominant neutrophils along with lymphocytes, plasma cells

DISCUSSION

Pyoderma gangrenosum (PG) is a primarily sterile inflammatory neutrophilic dermatosis. It was first described by Brocq in 1916 as "phagedenisme geometrique" and later named by Brunsting *et al.* The precise etiopathogenesis of PG is not well understood. However immunological factors and neutrophil dysfunction is considered to be involved in etiopathogenesis of PG.

The borders of ulcers are well defined because of their striking blue color which clearly outlined the lesions as it extended peripherally in rough, serpiginous configuration. There is undermining and necrosis of the subcutaneous tissue, the epidermis remaining as a thin, gray translucent film extending over the crater of the lesion in a ragged, irregular fashion. On the advance of the underlying process, often at the rate of 1-2 cms in 24 hrs, a zone of erythema extends as an areola into the area of normal skin. PG occurs most commonly on the lower legs, pretibial area.

The diagnosis mainly depends on recognition of the evolving clinical features and is only supported by histopathology. For classical PG, the following major criteria must be met: rapid progression of a painful necrotic cutaneous ulcer with an irregular, violaceous and undermined border and the exclusion of other causes of cutaneous ulceration. In addition, two of the following minor criteria should be met: (i) a history suggestive of pathergy or the clinical finding of cribriform scarring, (ii) systemic diseases known to be associated with PG, (iii) histopathological findings (sterile dermal neutrophilia, with or without mixed inflammation or lymphocytic vasculitis) and (iv) treatment response (e.g. rapid response to systemic corticosteroid). Massive neutrophilic infiltration ("sea of neutrophils"), in the absence of vasculitis and granuloma formation, is typical of PG.

Pyoderma gangrenosum is a dermatologic emergency because it is often rapidly progressive, causing severe local tissue destruction. It is frequently misdiagnosed as an infectious process and is treated by debridement. However, surgical

procedures or any mechanical manipulation of acute lesions induces progression of disease to the normal surrounding skin, enlargement of lesion and further tissue destruction. Therefore, it is important to recognize and treat these lesions early to avoid massive tissue destruction and loss. Corticosteroids and immunosuppressive therapy is the mainstay in the treatment of PG, given after excluding infectious process. The treatment of underlying disease may aid in healing. In patients without an identifiable associated disease, it is still possible for it to appear later; hence follow-up and evaluation are required even after the skin lesions have healed.

Our patient satisfied the diagnostic criteria with 1 major plus 3 of the minor criteria. This patient was treated with IV methyl prednisolone and other supportive measures. She showed improvement, ulcers healed with scarring and is under our follow up.

CONCLUSIONS

The clinical spectrum of pyoderma gangrenosum varies. It is like the tip of an iceberg indicating an underlying systemic disease. In our patient, all possibilities of systemic diseases were excluded and was concluded to have idiopathic pyoderma gangrenosum.