



PYODERMA GANGRENOSUM

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Pyoderma gangrenosum (PG) is an uncommon, recurrent and chronic skin disease characterized by its distinct chronic ulcer. Initial lesions may resemble small bug bites or papules that eventually develop deep ulceration and chronic wounds. Since the clinical manifestations vary based on the individual and underlying cause, a biopsy may be required to make a definitive diagnosis.

The name pyoderma gangrenosum is a misnomer; it has neither an infectious nor gangrenous etiology. Pyoderma gangrenosum is commonly associated with systemic disease. Approximately 50% of patients with PG have underlying conditions like inflammatory bowel disease (ulcerative colitis and Crohn's disease), arthritis, myeloproliferative disorders (myelocytic leukemia, hairy cell leukemia), and PAPA syndrome (pyogenic arthritis, pyoderma gangrenosum and acne). PG most commonly presents in middle age women, which parallels the demographic afflicted with autoimmune diseases.

Although a strong association is seen with underlying systemic or immunologic diseases, approximately 25-50% of cases remain idiopathic. Currently, experts attribute PG to failing neutrophilic function, or a humoral and cell-mediated immunity dysfunction; however these findings have not been consistently demonstrated.

One of the most unique cutaneous features of PG, found also in Behcet's disease, is known as pathergy. Pathergy is a condition where the appearance of an exaggerated skin lesion arises secondarily to minor trauma such as scratches, bumps and needle pricks. Although both PG and Behcet's disease demonstrate pathergy, the lesions seen in PG are often responsive to topical steroids, whereas those in Behcet's are not.

The classic ulcerative lesion in PG typically originates as an innocent appearing tender papule or pustule surrounded by an erythematous base. This sentinel lesion rapidly enlarges and eventually undergoes central necrosis resulting in an ulcer. The number of lesions may vary from a single, well-defined ulcer to greater than a dozen, which may coalesce encompassing large areas of the body. Other variants of PG include the bullous, pustular, and superficial granulomatous forms.

No uniform or specific therapy exists for PG, and therefore treatment regimens can vary among dermatologists. Guiding factors in choosing a therapeutic approach include size, number, depth, rate of expansion and any associated underlying disease. The mainstay of treatment has traditionally been local and or **systemic corticosteroids**, which aim at reducing the inflammatory nature of the wound. In cases demonstrating resistance to corticosteroids, both **tacrolimus** and **cyclosporine** have been used.

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