Pyoderma Gangrenosum: A Case Study

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Background

Pyoderma gangrenosum (PG) is a necrotizing, ulcerative skin disorder first described in 1930. Poorly understood, it poses a challenge in terms of both diagnosis and treatment. However, familiarity with this disease may help minimize the difficulty this illness presents.

Etiology:

- The etiology of PG is presently unknown.
- Theories for the cause of PG focus on demonstrated abnormalities in neutrophil behavior. Proposed theories include:
  - Deficiency of adherence protein, resulting in abnormal migration of neutrophils
  - Intracellular metabolic oscillations resulting in abnormal tracking of neutrophils
- Drug induced PG described, due to iodide, bromide, isotretinoin, colony-stimulating factors, propylthiouracil, and alpha-2b-interferon

Characteristics and Clinical Appearance:

- Demonstrates pathergy phenomenon (action at site of injury)
- Typically begins as discrete pustule or pustules surrounded by erythema
- May have fever, arthralgia, malaise, and myalgias
- Pustule breaks down into deep, red or purple ulceration. Center of ulcer is liquefying, and border may be covered by hemorrhagic blisters.
- Can be slow-growing or rapidly progressive
- 50-75% of cases associated with an underlying illnesses (See Table I)
- Four types of PG lesions:
  - Typical PG (found most commonly on legs, usually in conjunction with IBD)
  - Atypical or bullous PG (found more commonly in patients with hematologic disease)
  - Vegetative PG (found on back, painless lesions are superficial)
  - Malignant PG (found on head or neck, may have neurologic signs from nerve involvement)
- PAPA Syndrome – Autosomal dominant condition characterized by Pyogenic sterile Arthritis, Pyoderma gangrenosum, and Acne.

Demographics:

- Most common in people aged 25-54
- More common in women
- No racial association described
- Incidence difficult to determine, because of rarity of the disease

Diagnosis:

- Made by exclusion (See differential in Table II)
- Histopathology varies depending on type of PG, but most commonly shows massive neutrophilic infiltration, hemorrhage, and epidermal necrosis.
- Biopsy usually sterile, unless secondary infection present

Management:

- Supportive treatment for patient combined with local and systemic therapies, depending on extent of disease
- Local therapy consists of injections into the lesions. Most common agent used is triamcinolone hexacetonide. Other local treatments reported successful include cyclosporine, topical 5-aminosalicylic acid, topical sodium cromoglycate, topical nitrogen mustard, and topical nicotine via transdermal patches.
- Systemic therapy most commonly consists of prednisone therapy, 40-120 mg/day until ulcer is completely resolved, followed by tapering of therapy. IV pulse therapy has also been used. Other systemic agents reported to be successful include cyclosporine, dapsone, minocycline, methotrexate, and plasmapheresis with cyclophosphamide.
- Surgical debridement is contraindicated because of the pathergic nature of PG.

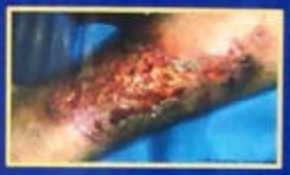

TABLE I

Illness Associated with Pyoderma Gangrenosum
A. Inflammatory Bowel Disease (esp. UC)
B. Arthritis (commonly polyarticular or psoriatic arthritis)
C. Monoproteinemias (esp. benign IgA type)
D. Hematologic malignancy (leukemia, polycythemia vera, myelofibrosis, multiple myeloma)
E. HIV
F. Autoimmune hepatitis
G. Behcet's disease

TABLE II

Differential Diagnosis of PG-Like Lesions
A. Infectious process
B. Malignancy
C. Vasculitis
D. Insect bite
E. Vascular insufficiency
F. Halogenoderma
G. Anticoagulant-associated skin necrosis
H. Sweet's syndrome
I. Factitious ulcers

Case

Chief Complaint

Patient is a previously healthy 35 year-old male presenting with a 2 month history of a progressively spreading ulcer on his left leg.

History of Illness

Two weeks prior to the onset of the ulcer, patient injured his left shin, resulting in a small abrasion. He recalls no other trauma to his leg. The site of injury became tender and slowly began to grow and ulcerate. The ulcer continued to grow more painful, and began to drain a cloudy yellow substance. Patient recalls no fever, malaise, myalgias or arthralgias. Patient reports no other symptoms.

Physical Exam

Patient's vital signs were within normal limits, with temperature at 37° C and BP at 120/78 mm Hg. There is a 2 cm ulcer on the left leg with a central areas of necrosis and a 5 cm area of erythema surrounding the ulcer.

Management

The patient had received several courses of antibiotics as an outpatient, including cephalexin, amoxicillin-clavulanate, and ofloxacine-ear. The ulcer showed no improvement, however. He was admitted to the hospital, where he underwent two surgical debridements. Cultures were obtained for AFB and fungi, but came back negative. Bacterial cultures grew S. epidermidis. Histology revealed necrotizing lipodrus. The patient was treated with courses of ceftazidin and ciprofloxacin, but the ulcer showed no improvement. At this point, a presumptive diagnosis of PG was made, which was confirmed through an outpatient dermatology consult. The patient was treated with 60 mg prednisone daily, and showed impressive improvement within 8 weeks. Prolonged dressings, Silvadene were also used to aid in the healing of the ulcer and were also able to minimize the patient's pain. Indeed, pain medication was unnecessary. The patient was scheduled for a colonoscopy, but was subsequently lost to follow up.

Conclusion

Our patient's diagnosis was made based on clinical presentation. Three biopsies were performed, which did not reveal a diagnosis, but were important to exclude other possibilities. Ideally, the patient would have undergone a colonoscopy because of the high incidence of IBD in patients with PG. Indeed, had an associated illness been apparent in this patient, the clinical picture may have been clearer.

Our case well illustrates the difficulty PG can present a clinician. The long differential diagnosis for ulcers of this sort can lead one to choose a treatment which actually exacerbates the disease. Familiarity with PG is so important because the clinical picture can be very confusing. With a confident grasp of the disease and its associations, a physician will be well prepared to meet and conquer this challenging illness.