Pyoderma Gangrenosum Involving the Foot

A Case Report

HOWARD KASHEFSKY, DPM*
ELVIRA CALLAHAN, DPM†
USHA C. RUDER, MD‡
PATRICK J. GRISAFI, DPM§
MICHAEL P. DELLACORTE, DPM||

Pyoderma gangrenosum is a rare and destructive inflammatory skin disease. The authors present a report of a patient with a classic case of pyoderma gangrenosum involving the foot. The diagnosis was made on the basis of clinical presentation and progression of the disease after differential diagnoses of common conditions were excluded. A brief overview of the disease process, its treatment, and its correlation with ulcerative colitis is provided. (J Am Podiatr Med Assoc 89(3): 137-140, 1999)

Pyoderma gangrenosum was first described by Brunsting et al[1] in 1930. Pyoderma gangrenosum is a rare, destructive inflammatory skin disease in which a painful nodule or pustule breaks down to form a progressively enlarging ulcer with a raised, tender, undermined border. There may be one or several lesions associated with the disease. In 10% to 50% of cases, the disease is a purely cutaneous disorder.[2, 3] In the remainder, lesions are associated with a systemic disease, such as ulcerative colitis, Crohn's disease, arthritis, or hematologic disorders. It was once believed that 50% of patients with pyoderma gangrenosum had ulcerative colitis; this proportion is now thought to be much lower, approximately 10%.[4] By contrast, the association with Crohn's disease has increased.[5] The etiology of pyoderma gangrenosum remains unknown. Lesions in the early stage are always sterile, but later in their course a variety of organisms have been found in cultures of the ulcers. Lesions associated with pyoderma gangrenosum demonstrate the phenomenon of "pathergy," or the development of new lesions following minor trauma. This explains the exaggerated and uncontrolled inflammatory responses that often accompany pyoderma gangrenosum.

*Submitted during third-year residency, Catholic Medical Center of Brooklyn and Queens, New York. *Mailing address:* 6213 N Hills Dr, Apt E, Raleigh, NC 27609.

†Submitted during first-year residency, Catholic Medical Center of Brooklyn and Queens, New York.

‡Director, Pathology Clinical Services, Catholic Medical Center of Brooklyn and Queens, New York.

§Chief, Division of Podiatry, and Director of Podiatric Medical Education, Catholic Medical Center of Brooklyn and Queens, New York.

||Director of Podiatry, Mary Immaculate Hospital, New York; Director, Primary Podiatric Medicine Program, Catholic Medical Center of Brooklyn and Queens, New York.

Clinical Manifestations

The lesions of pyoderma gangrenosum may begin as deep, painful nodules or as superficial, hemorrhagic pustules. Eventually these lesions develop into irregular ulcers with raised inflammatory borders and a spongy, necrotic base.[6] Pustular or hemorrhagic exudate may accompany the lesion. The margins are erythematous and the bases usually contain some necrotic material. The lesion may be very superficial, but more often than not, it will invade the fat and even the fascia. Lesions are usually solitary, but they may gather into a cluster, which will eventually become a single large lesion. Pyoderma gangrenosum may occur in patients of any age, and the condition

may last for weeks or months. It may affect any body part, although it has a predilection for the lower extremities, buttocks, abdomen, and face. The ulcer may be aggressive and cover the whole body part in days, or it may be slow-growing and appear quiescent.

Healing begins in the center of the lesion, which becomes a scar. Histologic findings of pyoderma gangrenosum are variable and nonspecific. Under a microscope, older lesions may show pseudoepitheliomatous hyperplasia in the margins of the ulcer, foreign body–type granuloma formation, capillary proliferation, and fibrosis. There are no specific laboratory tests to make a definitive diagnosis of the disease, and histopathology is only suggestive; therefore, the diagnosis of pyoderma gangrenosum is based on its clinical presentation and its course.

Treatment

In cases in which pyoderma gangrenosum is associated with a systemic disorder, treatment must be designed to heal both pyoderma gangrenosum and the systemic disease. Topical treatment is usually not sufficient to treat any type of pyoderma gangrenosum. Local wound care, however, is very important because it is essential in preventing infection. Any type of surgical procedure must be discouraged, as it will trigger new lesions because of the pathergy phenomenon.

Systemic corticosteroids in high doses represent the most effective treatment for pyoderma gangrenosum. This type of treatment causes side effects in about 50% of patients.[7] Pulse therapy with very high doses of methylprednisolone (1 g per day for 5 days) has been reported to stop progression of pyoderma gangrenosum lesions without any side effects.[8] Some sulfa drugs have been successful in treating pyoderma gangrenosum lesions, but not all patients respond to this type of treatment.[6] Sulfa drugs in combination with corticosteroids, especially in initial phases of the disease, may be necessary. It has been reported that 300 mg per day of clofazimine stops progression of lesions in 1 to 2 weeks and leads to complete or partial healing in 2 to 5 months.[1,9] There are usually fewer side effects with clofazimine than with corticosteroids.

Course and Prognosis

Pyoderma gangrenosum is a very unpredictable disease. Spontaneous healing may occur, but as old lesions heal, new ones may develop. The disease may lie dormant for a very long time and then reappear following trauma or surgery, or for no apparent reason. When associated with ulcerative colitis, pyoderma gangrenosum may parallel its course. The prognosis of pyoderma gangrenosum is good, but there may be considerable scarring following the disease.

Case History

A 58-year-old woman presented to the emergency department at the Catholic Medical Center of Brooklyn and Queens with the chief complaint of a painful "blister" on her right foot. The patient stated that the lesion was not responding to oral antibiotics and local wound care. She reported that the pain had appeared suddenly, 3 days before presentation, and was becoming worse. The symptoms had became so debilitating that the patient was unable to walk.

The patient's medical history included pneumonia, pancreatic abscess, gastrointestinal bleeding, ulcerative colitis, mild asthma, and anemia (the patient had refused transfusions in the past). Her current medications were cephalexin monohydrate, metronidazole, and a low dose of prednisone. Her surgical history included colonoscopy and colostomy. The patient denied having any allergies. Her social history and family history were unremarkable.

Physical examination revealed that the patient had a temperature of 99.6°F, stable vital signs, and a body weight of 240 pounds. The patient was alert and well oriented to her surroundings. Examination revealed no abnormal findings of the head, mouth, neck, lungs, or neurologic system. The patient did, however, have a protuberant abdomen with a tender right-lower quadrant. Lower-extremity physical examination revealed normal vascular, orthopedic, and neurologic systems for both limbs. The dermatologic examination revealed a blackened, fluid-filled sac on the dorsolateral aspect of the right foot (Fig. 1). The lesion was 1 cm in diameter and was surrounded by a 1-cm erythematous rim. Severe tenderness was elicited on palpation of the area.

Laboratory studies revealed the following values: white blood cell count, $14.1 \times 10^3/\mu l$; hemoglobin, 12.3 g/dL; hematocrit, 38.3%; blood urea nitrogen, 15 mg/dL; creatinine, 0.9 mg/dL; glucose, 162 mg/dL; hemoglobin A_{1c}, 6.4 g/dL (near normal); prothrombin time, 11.8 sec; partial thromboplastin time, 23.1 sec; and erythrocyte sedimentation rate, 146 mm/h. The antinuclear antibody and hepatitis profile test results were negative. Foot radiographs were unremarkable. A culture taken from aspiration of the lesion during the initial physical examination revealed no bacterial growth. Additionally, there was no bacterial, fungal, or acid-fast growth from multiple biopsy sites tested 1 week later. Blood cultures were also negative. The phenomenon of pathergy was noted 24 to 48 hours after the biopsy (Fig. 2). Pathology findings of the



Figure 1. Pyoderma gangrenosum at initial presentation.



Figure 2. The pathergy phenomenon following tissue biopsy.

foot biopsy revealed pseudoepitheliomatous hyperplasia of the epidermis with organized granulation-tissue proliferation (Fig. 3). Pathology results of the colon biopsy revealed an active phase of mucosal ulcerative colitis.

An exhaustive process of exclusion of common conditions related to the patient's symptoms, based on the clinical presentation and course of the disease, resulted in a diagnosis of pyoderma gangrenosum. The patient's ulcerative colitis and pyoderma gangrenosum improved with an increase in dosage of corticosteroids. Her long-term treatment consisted of prednisone and sulfasalazine. The patient was prescribed oral antibiotics upon discharge from the hospital to prevent a secondary infection.

The patient did not return for follow-up as directed. Six months after the initial presentation, the patient was hospitalized for unrelated health problems. The patient was examined and interviewed at her bedside by the hospital's podiatric physicians. The patient stated that she had completed the course of antibiotics prescribed for her during her last hospital stay and was still taking the steroids. She said that her right foot was asymptomatic. The neurologic, vascular, and orthopedic examinations of the right foot revealed normal findings. Dermatologic examination revealed a large area of hyperpigmented scarring on the dorsolateral aspect of the right foot (Fig. 4).

Discussion

This case presentation is unique because it documents a rare disorder, pyoderma gangrenosum, from initial presentation to resolution of the disease. The

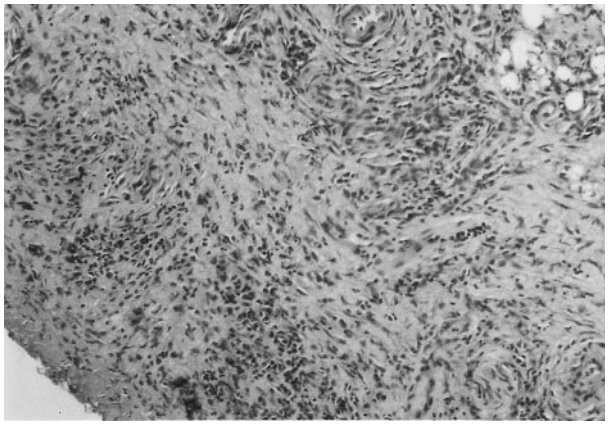


Figure 3. Capillary proliferation and nonspecific chronic granulomatous inflammation (H&E, ×40).



Figure 4. There was considerable scarring at the 6-month follow-up.

definitive diagnosis of pyoderma gangrenosum was made after care was taken to exhaust all other possible causes of this lesion. The diagnosis was made by observation of clinical correlation to the disease process. This included an initial presentation of a painful sterile nodule that eventually ulcerated and displayed the pathergy phenomenon after biopsy. Nonspecific findings were histologically compatible with the disease process. In this case, pyoderma gangrenosum finally responded to high-dose steroid treatment. It is important for the podiatric physician to be aware of such conditions because they are so rarely encountered.

References

1. Brunsting LA, Goeckerman WH, O'Leary PA: Pyoderma (ecthyma) gangrenosum and rheumatoid arthritis. Arch Dermatol **22:** 665, 1930.
2. Newell LM, Malkinson FD: Pyoderma gangrenosum [commentary]. Arch Dermatol **118:** 769, 1982.
3. Hickman JG, Lazarus GS: Pyoderma gangrenosum: a reappraisal of associated systemic diseases. Br J Dermatol **102:** 235, 1980.
4. Levitt MD, Richie JK, Lennard-Jones JE, et al: Pyoderma gangrenosum in inflammatory bowel disease. Br J Surg **78:** 676, 1991.
5. Schoetz DJ, Coller JA, Veindenheimer MC: Pyoderma gangrenosum and Crohn's disease. Dis Colon Rectum **26:** 155, 1983.
6. Perry HO, Brunsting LA: Pyoderma gangrenosum: a clinical study of nineteen cases. Arch Dermatol **75:** 380, 1957.
7. Holt PJA, Davies MG, Saunders KC, et al: Pyoderma gangrenosum: clinical and laboratory findings in 15 patients with special reference to polyarthritis. Medicine (Baltimore) **59:** 114, 1980.
8. Johnson RB, Lazarus GS: Pulse therapy: therapeutic efficacy in the treatment of pyoderma gangrenosum. Arch Dermatol **118:** 76, 1982.
9. Thomsen K, Rothenborg HW: Clofazimine in the treatment of pyoderma gangrenosum. Arch Dermatol **115:** 851, 1979.