

Pyoderma Gangrenosum in a Patient With Psoriatic Arthritis

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Pyoderma gangrenosum is an autoimmune disease of the skin that causes enlarging, painful ulcers with ragged, undermined, purplish borders. Pyoderma gangrenosum is frequently mistaken for other conditions, and early diagnosis is essential to avoid disfiguring surgery and prolonged recovery. We report a case of pyoderma gangrenosum in a patient with psoriatic arthritis that was initially treated as an infected spider bite. Once the diagnosis was made, the patient recovered after 14 months of immunosuppressive therapy.

Case Report

A 34-year-old woman with a medical history notable for psoriasis, psoriatic arthritis, and fibromyalgia came to the clinic complaining of a several week history of an enlarging, erythematous ulcer on her right lower extremity. The lesion began suddenly with the appearance of an open sore that was moderately painful. There was no history of trauma or insect bite. The ulcer was initially treated as either an infected brown recluse spider bite or an infected venous stasis ulcer, and the patient was given topical mupirocin and oral cephalexin. Despite treatment, the lesion progressed in size to 6.0×4.0 cm, with central necrosis and shaggy, purplish overhanging borders (Figures 1 and 2).

The patient was referred to a dermatologist with a provisional diagnosis of pyoderma gangrenosum. She was prescribed 60 mg of prednisone and 400 mg of cyclosporine daily. Within 2 weeks the inflammation dramatically subsided, and prednisone was tapered to 20 mg daily. Within a month the prednisone was stopped, but the cyclosporine was continued. Because of insurance changes, the patient saw a new dermatologist, who discontinued

the cyclosporin and prescribed methotrexate, 12.5 mg weekly, to treat the pyoderma, psoriasis, and psoriatic arthritis. Methotrexate was tapered to 10 mg weekly and continued for 13 months. The lesion gradually healed to leave a hyperpigmented, atrophic scar.

Discussion

Pyoderma gangrenosum is a rare autoimmune disease of the skin characterized by rapidly progressive ulcers that have a unique appearance.¹⁻⁸ Although pyoderma gangrenosum can occur at any age, it most commonly affects young to middle-aged adults, with a slight female predominance.¹ The lesions of pyoderma gangrenosum typically begin as innocuous-appearing papules or papulopustules. These lesions rapidly progress within a few days to become painful ulcers with shaggy, overhanging purplish edges.¹⁻⁸ The liquefying central areas of the lesions do not form eschars and contain granulation tissue, purulent exudate, and necrotic debris. The undermined, dusky border with hemorrhagic vesiculation is virtually pathognomonic for this condition.⁸ Surrounding the ulcer is an area of induration and erythema,^{1,2} making this lesion easily confused with brown recluse spider bites or infectious causes.^{1,8}

Pyoderma gangrenosum occurs in two forms, typical and atypical. Typical pyoderma gangrenosum is found on the lower extremities 75% of the time, although a smaller percentage of cases occurs on the perineum. These lesions tend to be deeper, with the characteristics noted above. Atypical pyoderma gangrenosum occurs on the upper extremities 75% of the time, is more superficial, and is characterized more often by hemorrhagic bullous formation.^{1,7} An important aspect of either form of pyoderma gangrenosum, found in 20% to 30% of cases, is pathergy, a condition in which severe extension of ulcers can occur from even the minimal trauma associated with intravenous cannulation, biopsy, or surgical debridement.^{1,6-8} For this reason, biopsy should be avoided unless other conditions,

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Figure 1. Ulcerative lesion of pyoderma gangrenosum measuring 6.0×4.0 cm on the right medial shin of a 34-year-old woman with psoriasis and psoriatic arthritis. The erythema on the patient's foot was psoriasis.

such as deep fungal infections or vasculitis, must be ruled out.^{6–8} This caution is especially important because the histopathology of pyoderma gangrenosum is relatively nonspecific, showing chronic ulceration or a neutrophilic vascular reaction in most cases.^{1,8}

About one half of all cases of pyoderma gangrenosum are associated with other systemic diseases.¹

In keeping with its autoimmune nature, many patients have coexisting disorders, including ulcerative colitis, Crohn disease, rheumatoid arthritis, or psoriasis. There is also a strong association with hematologic malignancies. Because of these associations, it is imperative to search for signs of systemic illness, unless such an illness is already apparent.^{1,8}



Figure 2. Same lesion close up. Notice overhanging purplish border, necrotic center, and surrounding rim of erythema. This lesion was initially treated with antibiotics as either an infected brown recluse spider bite or an infected venous stasis ulcer.

Table 1. Differential Diagnosis of Pyoderma Gangrenosum.

Infection
Bacterial infection (eg, syphilitic gumma)
Mycobacterial infection
Fungal infection
Parasitic infection (eg, cutaneous amebiasis)
Viral infection (eg, chronic ulcerative herpes simplex)
Sweet syndrome
Insect bite
Brown recluse spider bite
Malignancy
Squamous cell carcinoma
Basal cell carcinoma
Cutaneous T-cell lymphoma
Halogenoderma
Iododerma
Bromoderma
Factitial ulceration
Ulcerative necrobiosis lipoidica diabetorum
Vascular disease
Venous or arterial insufficiency
Antiphospholipid antibody-associated occlusive disease
Thrombophlebitis with gangrene
Syndrome with vasculitis
Systemic lupus erythematosus
Rheumatoid arthritis
Behçet disease
Wegener granulomatosis

From Bennett et al.¹ Reprinted with permission.

As in our patient, the initial diagnosis of pyoderma gangrenosum is frequently missed in favor of an alternative diagnosis from an extensive list of differential diagnoses (Table 1).¹ Patients are often hospitalized and given high doses of broad-spectrum antibiotics, even undergoing surgical debridement, which exacerbates the lesions and prolongs recovery.^{6–8} Thus, it is important to consider pyoderma gangrenosum in the differential diagnosis among patients who come to their primary care physician with ulcerative lesions, particularly ulcers of the lower extremities that fail to heal.^{1,6–8}

Treatment of pyoderma gangrenosum is based on severity of the disease and ranges from topical corticosteroid agents and local measures to high-dose corticosteroids and other systemic immunosuppressants. Bennett and colleagues¹ recommend a stepped approach based on the stage of the ulcer. Those who have an ulcer in the inflammatory stage (as our patient did) are initially given prednisone, 1 mg/kg, with long-term follow-up. The dosage of

prednisone can be tapered rapidly by 20 mg every 3 or 4 days⁸ then tapered more slowly. Inflammation usually resolves within 6 months, complete remission occurs within 11 months, and patients typically are off steroids by 14 months.¹ Because of the numerous side effects of long-term corticosteroid therapy, a second steroid-sparing agent,¹ such as dapsone,^{1,8} cyclophosphamide,⁹ cyclosporine^{1,8,10,11} (as our patient received early), or methotrexate^{1,12} (as our patient later received) is usually added early. Patients taking steroids should be counseled regarding vitamin D and calcium supplementation, and a baseline bone densitometry should be performed. It is prudent also to consider bone antiresorptive therapy to prevent osteoporosis (eg, estrogen replacement among women, bisphosphonates, or calcitonin).^{13,14}

Conclusion

Pyoderma gangrenosum is an ulcerative autoimmune disease of the skin causing painful lesions that have a characteristic appearance. This disease is associated in 50% of cases with other systemic diseases, such as inflammatory bowel disease, rheumatoid arthritis, or hematologic malignancies, and a search should be made to rule out these comorbid conditions. Treatment of pyoderma gangrenosum involves long-term immunosuppression. To avoid potentially disfiguring surgery and to uncover important comorbid conditions, family physicians should always consider this diagnosis in patients with enlarging, painful skin ulcers that fail to respond to antibiotics or simple local measures.

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