Pyoderma Gangrenosum: An Inside Job

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ANNALS CASE
“A 23-year-old woman with a history of arthralgias and inflammatory bowel disease presented to the emergency department (ED) with a progressive, painful, atraumatic leg lesion. When first noticed 2 weeks before, it was coin-sized and slowly grew, with ulceration. She was treated with antibiotics and then oral steroids and cyclosporine, but the lesion rapidly expanded—up to 2.5 cm of wound margin extension daily—to now involve most of her lateral lower leg. On examination, the patient was afebrile, with normal vital signs. Initial laboratory study results were unremarkable, and she was admitted for aggressive inpatient wound evaluation and management. Biopsy supported her presumptive diagnosis of pyoderma gangrenosum [PG]."1

Quick! What’s the first thing that comes to mind when you see these images? That sand pit monster on Tatooine, where Jabba the Hutt tries to throw in Luke Skywalker? No, not a Star Wars fan? Well, we suppose it does look more like an unroofed abscess blossoming with fresh maggots."2

SO YOU HAVE AN ULCER. UM...

Ulcers are such a frustrating conundrum in emergency medicine because, honestly, they’re repulsive, there’s often no immediate relief for the patient, and the differential diagnosis seems, well, endless. Sure, our brains are triggered to instantly recognize certain bizz-buzz associations with ulcer presentations, such as necrotizing fasciitis, mucormycosis, or acute embolic disease, to name a few. But if you’re like us, we have quite a lot of brain-fart moments.

Whether your mind is drawing a blank or is tied up in a mental cat’s cradle, let’s first start with some basic dermatologic concepts and then go over a simple algorithm that will help organize your thought process the next time you see an ulcer.

SKIN BASICS
There are 3 components critical to maintaining a functioning skin barrier: an intact epidermis, a healthy dermis, and an adequate vascular supply. If there is derangement to any one of these components, your skin barrier is effectively compromised and you are prone to skin breakdown and ulceration. We’ll come back to this concept in a little bit, but ask yourself these 2 questions the next time you see an ulcer: is the cause internal or external, and if the cause is internal, is it infectious, inflammatory, or neoplastic?

STEP 1: INSIDE JOB OR OUTSIDE HACK?
The first step into investigating the ulcer cause is to find out whether the skin was broken into from the outside, whether self-induced (eg, trauma from chronically picking at the skin, pressure causing skin breakdown) or not (eg, injected venom from a brown recluse spider bite).2 Clinical context should help identify or exclude those causes. That being said, a really good “picker” can tell a good story and occasionally fool you.

STEP 2: “INSIDE JOBS”—INFECTION, INFLAMMATION, OR NEOPLASM?
For inside jobs, we go through the “infection, inflammation, or neoplasm” algorithm.

Infection
There are many infectious processes that can result in ulcerations. These range in severity and can affect any part of the functioning skin system: epidermis, dermis, or intravascular. Although bacteria (eg, Treponema pallidum, Streptococcus) and viruses (eg, herpes, Coxsackie) are the
first things popping into our minds, we should remember the often-overlooked causes: bacterial (eg, *pseudomonas*, anthrax), fungi (eg, mucormycosis, coccidioidomycosis), parasites (eg, leishmaniasis, amoebiasis, schistosomiasis), and mycobacteria.

**Inflammatory**

Most inflammatory conditions causing ulceration do so by affecting the vasculature. A compromised vascular supply causes a breakdown of the epidermal and dermal layers. Inflammation of the blood vessels leads to a vasculitis, and vasculitis of the medium-sized vessels (eg, polyarteritis nodosa, microscopic polyangiitis, Wegener’s granulomatosis, rheumatoid vasculitis) can lead to ulceration. Ulceration can also be caused by embolic or thrombotic processes within the vessels. Without blood supply, the dermal and epidermal layers will necrose. Although embolic and thrombotic phenomena are technically not inflammatory, these will be considered here for the sake of simplicity.

**Neoplasm**

Commonly, nonmelanoma skin cancers, such as basal cell carcinoma and squamous cell carcinoma, will cause the skin to ulcerate. In addition, cutaneous lymphomas can cause ulceration of the skin. Practically speaking, though, if you’re suspecting malignancy, you should be referring the patient for a skin biopsy to make the definitive diagnosis.

**The Leftovers**

After having gone through all of that, what we’re left with are the rare and weird cases. Remember the triad of intact epidermis, healthy dermis, and adequate vasculature? Well, when your immune system goes haywire and sends a bunch of neutrophils to the dermis, causing inflammation, ulceration can occur because of the overwhelming inflammation and edema. These are the neutrophilic dermatoses: PG, Sweet’s syndrome, and Behçet’s disease. Let’s just say that these conditions are rare, very difficult to diagnose, and most often not diagnosed in the ED.

**IF IT TAKES A DERMATOLOGIST 10 LAB TESTS, A BIOPSY, AND TISSUE CULTURES…**

Specifically in regard to this case, PG is a rare condition in the family of neutrophilic dermatoses, resulting in ulceration of the skin, significant pain, morbidity, and disfigurement. Classically, PG is associated with underlying inflammatory bowel disease, polyarthritis, or a hematologic malignancy. The associated images demonstrate the typical evolution of a pustule to the classic deep, cribriform ulcer with an undermined, violaceous border. PG often occurs on the lower extremities and peristomal sites, but can occur anywhere. And, although these nasty-looking lesions may exude a lot of purulence, some requiring several dressing changes a day, PG’s name belies the fact that it is neither infectious nor gangrenous. As you can imagine, infection often tops the differential diagnosis and patients typically have received multiple courses of antibiotics without improvement, along with unremarkable wound culture and laboratory test results. In fact, the diagnosis of PG is classically taught to be a “diagnosis of exclusion.” All other potential causes of the patient’s ulcer must be excluded before a final diagnosis can be made. This causes a lengthy and, by definition, fruitless search for the truth. Whew! Thank goodness for outpatient providers.

**PATHERGY …SAY WHAT?**

There is one thing that clinically stands out, though. Pathergy is an exaggerated skin response, causing blistering or ulceration, after a minor trauma. It can be seen in a few inflammatory conditions, namely, PG and Behçet’s disease, both neutrophilic dermatoses. Although pathergy can be quite troublesome to a patient, it can be a useful pearl when PG is presumptively diagnosed. Because the diagnosis of PG can be “elusive,” the presence of pathergy can be a serendipitous finding that only an astute clinician may notice. Pathergy will manifest approximately 24 to 48 hours after a small injury, such as a needle stick. So if a patient with known inflammatory bowel disease comes to the ED and shows an ulcer at the site of a blood draw from the day before, you may have clinched your diagnosis of PG. Furthermore, pathergy is the reason debridement is not recommended for patients with PG because it can actually worsen the condition!

**A WORD ABOUT TREATMENT**

As you can imagine, suspected PG should be managed by a dermatologist and wound care specialist. Some mild cases can be managed with topical or intralesional steroids, but long-term treatment often includes a variety of immunosuppressants, including oral steroids, cyclosporine, mycophenolate mofetil, or tumor necrosis factor-α inhibitors.

**BACK TO THE CASE**

There are a few hints in this case that led us to the presumptive diagnosis of PG: the characteristic appearance and progression of the lesion, along with the important red flag of a history of inflammatory bowel disease. Once other diagnoses were excluded and biopsy results supported the PG diagnosis, the patient’s immunosuppression treatment

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Annals of Emergency Medicine 117

DeClerck et al  
EM:RAP Commentary
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**IMAGES IN EMERGENCY MEDICINE**

(continued from p. 115)

**DIAGNOSIS:**

*Pyoderma gangrenosum.* Biopsy supported her presumptive diagnosis of pyoderma gangrenosum. Her immunosuppression treatment was escalated to methylprednisolone, cyclosporine, and azathioprine, with prompt wound improvement (Figure 3).

Pyoderma gangrenosum is a rare, noninfectious, ulcerative, neutrophilic skin disease characterized by infiltration and destruction of tissue.1-4 Lesions mimic infection, occur most often in adults, typically involve the lower extremities, and are associated with conditions including malignancy, inflammatory bowel disease, hepatitis, and rheumatoid arthritis.1 Lesions can rapidly progress, and delayed treatment may lead to wound complications and increased likelihood of surgical intervention.2 Its diagnosis is largely one of exclusion, assisted by clinical criteria—major diagnostic criteria include presence of a painful, necrolytic, cutaneous ulcer expanding daily at 1 to 2 cm or greater, and exclusion of other causes of ulceration.1-3 Lesion biopsy may support the diagnosis, and treatment includes immunosuppressive agents and supportive wound management.3 Invasive procedures such as irrigation and debridement rarely improve outcomes.4

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