Sterile but Serious – Pyoderma Gangrenosum A Case Study

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Introduction

Medicine is seeing a greater dependence on diagnostic tests in both the inpatient and outpatient setting. While physicians continue to use clinical knowledge to come to a diagnosis, there is a desire to obtain the confirmation in the form of a test. This is one such case where all the tests results are negative, there are no significant radiological findings, and the pathology slides are sterile. The diagnosis was suggested by a seasoned physician who was studying for his recertification. He happened upon a question with a presentation similar to this case.

Case Summary

The patient is a 41-year-old female with no past medical history who presented with a left leg wound. 4 days prior she had hit her leg against a chair resulting in a small bump. The wound increased in size and started to have a burning sensation. She denies any fever, chills, joint pain, rash, vomiting, recent travel, fatigue, or muscle aches.

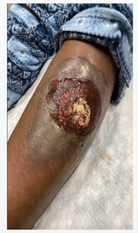
Physical Exam

General: No acute distress.
Respiratory: Clear to auscultation bilaterally.
Cardiac: Tachycardic, regular rhythm, no murmurs auscultated.
GI: Soft, non-tender, positive bowel sounds, no hepatosplenomegaly.
MSK: Lower: normal ROM in hips, knees, ankles. On the anterior of the patient's left lower extremity is an ulcer. At the center is a 2-3 cm erythematous flesh exposures regions, that is limited to the subcutaneous layer. Surrounding that is a 7-6c m region that is surrounded by grey sloughing skin. Around that is another 13 cm region of edematous indurated skin that is mildly erythematous.
Neuro: 5/5 strength in UE and LE bilaterally. Normal gait and coordination
Extremities: 2+ dorsalis pedis pulses bilaterally.

Labs and Imaging

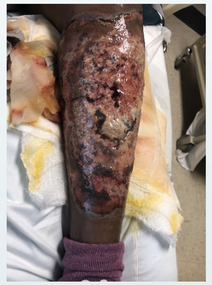
	Result	Reference
WBC	26.1	3.5-11.0
Hgb	11.7	12.0-16.0
Hct	34.9	36.0-48.0
Platelet	264	130-400
Neutro	23.9	1.5-5.2
Lymph	1.1	1.2-4.2
Mono	1.1	0.2-1.0
Eos	0.0	0.0-0.5
Baso	0.0	0.0-0.2
Sodium	138	135-145
Potassium	3.4	3.5-5.0
Chloride	109	97-109
CO2	20	22-30
Glucose	197	65-99
BUN	22	6-22
Creatinine	0.78	0.60-1.30
CRP	28.90	0.00-0.30
Lactate	2.6	0.4-2.0

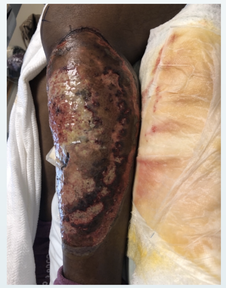
MRI:
Diffuse skin and subcutaneous fat edema consistent with cellulitis. No evidence of deep muscle or fascial involvement. No evidence of bone bruise, osteomyelitis or fracture. No evidence of localized abscess collection or hematoma.



Day 1

Day 8





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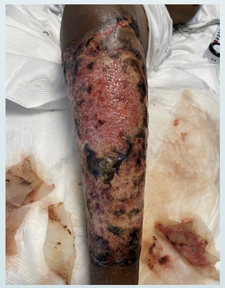


Initial Assessment and Treatment

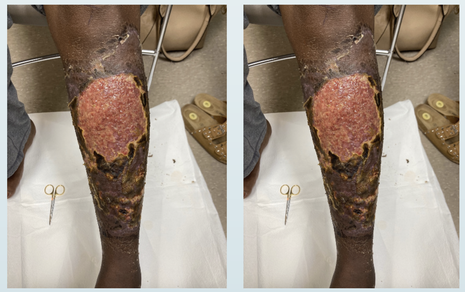
Patient was started on broad-spectrum antibiotics and had the wound debrided by surgery twice during her stay. MRI performed showed neither fascial or deep tissue involvement, ruling out necrotizing fasciitis. Wound and blood cultures were sterile. Biopsy report stated pathology was inconclusive and notably sterile. Patient's cellulitis worsened, covering most of her left leg and she remained tachycardic with leukocytosis.

Final Diagnosis

Eventually the patient was diagnosed with pyoderma gangrenosum. This diagnosis explained the sterile culture, lack of response to antibiotics, elevated neutrophil count, and worsening condition with debridement. Patient was started on antibiotics and wound started to granulate and heal.

2 weeks after initiating steroids

4 weeks after initiating steroids



Discussion

Pyoderma gangrenosum is a rare, painful, inflammatory skin disease caused by a dysregulated immune response that presents many challenges for the physician. It is not only difficult to diagnose, it is often is misdiagnosed leading to delayed treatment.

The lesions often start as unassuming pustule which can affect any area of the body but is commonly found on the lower limbs. The lesion enlarges and erodes in a short period of time resulting in a rapidly progressing, painful, marginated ulcer with seropurulent drainage. Wound culture is sterile and grows no bacteria. In a well-meaning attempt to clear necrotic tissue for wound healing, surgical debridement exacerbates the lesion. Wound biopsy reveals inflammation with an abundance of neutrophils often suggesting a neutrophilic dermatosis. There is no specific laboratory or histopathological test that can diagnose pyoderma gangrenosum making it a diagnosis of exclusion.

Management strategies include topical corticosteroids for mild lesions but the mainstay of treatment remains or oral systemic corticosteroids. Given the high dosage and lengthy interval required of steroid therapy, precautions should be taken including bone density evaluations. For refractory cases, TNF-alpha inhibitors such as infliximab can also be considered. The main focus of treatment is not only anti-inflammatory, wound healing, pain control, but diagnosing and treating associated diseases.

Pyoderma has a strong relationship with other systemic autoimmune diseases such as inflammatory bowel disease and rheumatoid arthritis, The next steps for the patient is to be screened early for associated comorbid conditions.

While this was a diagnosis of exclusion, this case challenged the traditional medical algorithm of wound equals antibiotics and debridement. This case emphasized the importance of depending on our clinical acumen, discussing difficult cases with colleagues, and continued lifelong learning.

References

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