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Ecthyma Gangrenosum in an Immunocompromised Patient without Detectable Bacteremia

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Introduction

- **Ecthyma gangrenosum (EG)** is a rare condition, classically a cutaneous complication of *Pseudomonas aeruginosa* bacteremia among immunocompromised patients, particularly in those with hematologic malignancy.¹
- The lesions of EG follow a particular pattern, starting as painless gray macules or papules with surrounding erythema, evolving into **pustules or hemorrhagic bullae**, and ultimately evolving into **necrotic ulcers** with central, black eschar surrounded by tender erythema.¹
- Patients are **typically acutely ill**. The presentation of bacteremia and EG is **often fatal**, but timely treatment with intravenous antibiotics can improve mortality.²
- In this report, we present a case of an immunocompromised man with a history of hematologic malignancy presenting with **EG with sepsis symptoms but without bacteremia** responding to empiric antibiotic coverage.

Images



Figure 1. Pustules with surrounding erythema on abdomen. Characteristic lesion enlarged in pop-out picture



Figure 2. Necrotic ulcers with central black eschar and surrounding erythema on right forehead (2a) and abdomen at waistline (2b).

Case

Clinical History

- A 70 year-old-man with type II diabetes mellitus, CLL, and a recent diagnosis of multiple myeloma on lenalidomide and bortezomib presented with fever (102.1F), tachycardia (HR 107), borderline neutropenia, and several days of self-reported “bug bites” to his right forehead.
- He reported localized itching and evolving periorbital swelling.

Physical Examination

- Right forehead: Hemorrhagic vesicles with exquisitely tender erythema/edema. Eye movements unimpaired and painless.
- Abdomen: Similar smaller, newer pustules and hemorrhagic vesicles with surrounding erythema (Figure 1).
- Forehead lesion progressed from hemorrhagic discharge to a large ulcer with central black eschar (Figure 2a). Black eschar was also seen on a lesion on the abdomen (Figure 2b).

Laboratory Workup and Treatment

- Maxillofacial CT: right-sided periorbital cellulitis
- Chemistries: hypocalcemia, hypoalbuminemia, elevated CRP
- The patient was promptly treated with empiric vancomycin and piperacillin/tazobactam given clinical criteria for sepsis were satisfied. Initial and repeat blood cultures remained negative.
- The lesions were not biopsied or cultured, given the high clinical suspicion of ecthyma gangrenosum based on the clinical examination and history details, and appropriate management had been initiated.

Clinical Outcome

- Vital signs improved and the skin lesions ultimately began improving prior to discharge.
- The patient was discharged on a 3-week course of ciprofloxacin and was instructed to continue holding immunosuppressive medications until a follow-up visit with his oncologist.

Differential

- Differential diagnoses included both **infectious** (disseminated varicella) and **non-infectious** causes (Sweet Syndrome)
- **Sweet syndrome** was not favored due to clinical morphology of the lesions, non-neutrophilic predominance, and progressive symptoms presenting in the setting of systemic steroid therapy.
- **Disseminated varicella** was not favored due to the generally well-appearing patient, lack of widespread distribution of lesions, and the present of hemorrhagic vesicles and pustules as opposed to clear vesicles as the primary lesions.

Discussion

- While EG was once considered to be pathognomonic of *P. aeruginosa* sepsis, a 2015 review found case reports demonstrating **clear cases of EG in the presence of other bacteria or in the absence of sepsis** altogether.³
- Approximately 9% of cases were non-bacterial, 15% involved the head or neck, and bacteremia was much less likely in non-pseudomonas cases.³ Notably, **patients without the characteristic bacteremia have improved prognoses** compared to their more traditional counterparts.⁴
- This case presents an example of the spectrum EG clinical manifestations, highlighting uncommon features - periorbital involvement and absent bacteremia.^{3,5,6} The multiple lesions and sepsis symptoms indicated disseminated disease.
- Improvement in the setting of empiric antibiotics coverage suggests a **potential bacterial pathogen that may have been missed on multiple cultures**.
- Consistent with what is seen in most cases, the **lesions progress** from hemorrhagic vesicles and pustules to necrotic ulcers with central eschar and surrounding tender erythema.³

Conclusion and Key Learnings

- It is important to consider EG in a **broader range of cases** beyond just *P. aeruginosa* bacteremia.
- Focus should be on **early empiric antibiotics**.
- Outcomes for patients without bacteremia appear to be better.

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