

Ecthyma Gangrenosum in Immunocompromised Patient without Detectable Bacteremia – A Case and Discussion

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Abstract

Ecthyma gangrenosum (EG) is typically pathognomonic of *Pseudomonas aeruginosa* bacteremia among immunocompromised patients, particularly with underlying malignancy. Recently, other pathogens and clinical histories have been implicated, challenging the classic picture of patients with EG. The cutaneous findings in patients follow a pattern of lesion progression from indurated pustules and hemorrhagic vesicles evolving to necrotic ulcers with central black eschar and surrounding erythema. While lesions typically occur on the perineum or lower extremities, their presence has also been described elsewhere.

Herein, we describe a case of an immunocompromised man with chronic lymphocytic leukemia and multiple myeloma actively undergoing chemotherapy presenting with EG and sepsis symptoms but without detectable bacteremia who responded to broad spectrum empiric antibiotic coverage including double anti-pseudomonal coverage.

It is important to consider EG even in cases without all of the classic presenting symptoms and manifestations of *P. aeruginosa* bacteremia given a potentially broad range of etiologies for this disease. As seen in this patient, clinical outcomes for patients without bacteremia may be better.

Introduction

Ecthyma gangrenosum (EG) is a rare condition, classically a cutaneous complication of *Pseudomonas aeruginosa* bacteremia among immunocompromised patients -- particularly those with hematologic malignancies.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 can be fatal in a large portion of cases, 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 detectable bacteremia who responded to broad empiric antibiotic coverage.

Case

A 70 year-old-man with type II diabetes mellitus, a four year history of chronic lymphocytic leukemia, and a recent diagnosis of multiple myeloma on lenalidomide and bortezomib presented with fever (102.1 Fahrenheit), tachycardia (heart rate 107), borderline neutropenia (absolute neutrophil count 1527 at 46.3% of white blood cells on admission), and several day history of self-reported “bug bites” to his right forehead with localized itching and progressive right periorbital swelling. Examination revealed an area of hemorrhagic vesicles on the right forehead surrounded by exquisitely tender erythema and edema. Eye movements were unimpaired and nonpainful. Similar smaller and newer pustules and hemorrhagic vesicles with surrounding

erythema were also seen on the patient's abdomen (Figure 1). The large forehead lesion progressed from hemorrhagic discharge to a large ulcer with central black eschar (Figure 2). Black eschar was also seen on a lesion on the abdomen (Figure 3).

An investigative workup was notable for right-sided periorbital cellulitis on maxillofacial CT, unremarkable urinalysis, hypocalcemia, hypoalbuminemia, and elevated CRP. The patient was promptly treated with empiric intravenous vancomycin and piperacillin/tazobactam given clinical criteria for sepsis were satisfied. Meropenem was later added for double coverage of pseudomonas for two days before transitioning to iprofloxacin. Initial and repeat blood cultures demonstrated no growth. Additionally, his home dexamethasone, lenalidomide, and bortezomib were held per his oncologist. 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. Vital signs improved and the skin lesions ultimately began improving prior to discharge. The patient was ultimately discharged on a 3-week course of ciprofloxacin and was instructed to continue holding immunosuppressive medications until a follow-up visit with his oncologist.

Important differential diagnoses considered were Sweet syndrome and disseminated varicella. Sweet syndrome was not favored due to clinical morphology of the lesions, a non-neutrophilic predominance, and symptoms presenting in the setting of systemic steroid therapy. Varicella was not considered 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 has been 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 manifestations, highlighting uncommon features – periorbital involvement and absent bacteremia.^{3,5,6} The multiple lesions of this patient 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 progressed from hemorrhagic vesicles and pustules to necrotic ulcers with central eschar and surrounding tender erythema.³ Additionally, this patient's positive outcome highlights the improved prognoses among non-bacteremic patients with EG.⁴

Given the potential for serious morbidity, it is important to consider EG in a broad range of patients and start empiric antibiotics coverage early.

Disclaimer

"This research was supported (in whole or in part) by HCA and/or an HCA affiliated entity. The views expressed in this publication represent those of the author(s) and do not necessarily represent the official views of HCA or any of its affiliated entities."

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