Received: 18 March 2019 | Revised: 12 June 2019 | Accepted: 16 June 2019

DOI: 10.1002/ccr3.2314

CASE REPORT

Clinical Case Reports Open Access WILEY

Localized ecthyma gangrenosum without sepsis in a neutropenic patient with a myelodysplastic syndrome—Refractory anemia with excess blasts type 2

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Abstract

The diagnosis of ecthyma gangrenosum should be evoked in front of maculopapular lesions rapidly evolving to necroting ulcers, particularly in the presence of prolonged neutropaenia or other hematological malignancies.

KEYWORDS

ecthyma gangrenosum, myelodysplasia, neutropenia

1 | INTRODUCTION

Ecthyma gangrenosum (EG) is a rare cutaneous infection most commonly associated with *Pseudomonas aeruginosa* bacteraemia. The infection typically occurs in immunocompromised or neutropenic patient. We report the case of a patient with myelodysplastic syndrome, refractory anemia with excess blasts type 2, who developed a localized EG without bacteraemia.

Ecthyma gangrenosum is a rare cutaneous infection most commonly associated with *P aeruginosa* bacteraemia.[1-3] The infection typically occurs in immunocompromised or neutropenic or critically ill patients and warrants prompt diagnosis and treatment.

We report the case of an EG in a patient with myelodysplastic syndrome—refractory anemia with excess blasts type 2 (MDS EB1).

2 | CASE PRESENTATION

A 74-year-old man with a SMD EB1 treated by blood transfusion support, developed 2 days ago, two papular and erythematous cutaneous lesions, on both the left leg and thigh. These lesions were painful, evolving in one week into nodular lesions with ulceration and central necrosis (Figure 1). No causal factors were found. The patient was in fair condition, and he had no fever. Blood tests demonstrated a stable anemia and a neutropenia (0.2 G/L) as well as an elevated CRP (216 mg/mL). Cultured swabs were positive to *P aeruginosa*. Different blood cultures were negative, consistent with nonbacteraemic EG. Treatment included intravenous amoxicillin-clavulanic acid (1 g × 3/d for 14 days). Within a few days, the lesions were much less painful and significant healing was noted (Figure 2).

3 | DISCUSSION

To our knowledge, there are few cases of nonbacteriemic cutaneous localized EG in patients with myelodysplastic syndrome—refractory anemia with excess blasts type 2. Indeed, myelodysplasia is often associated with lymphoma or acute leukemia and EG is then septicemic.

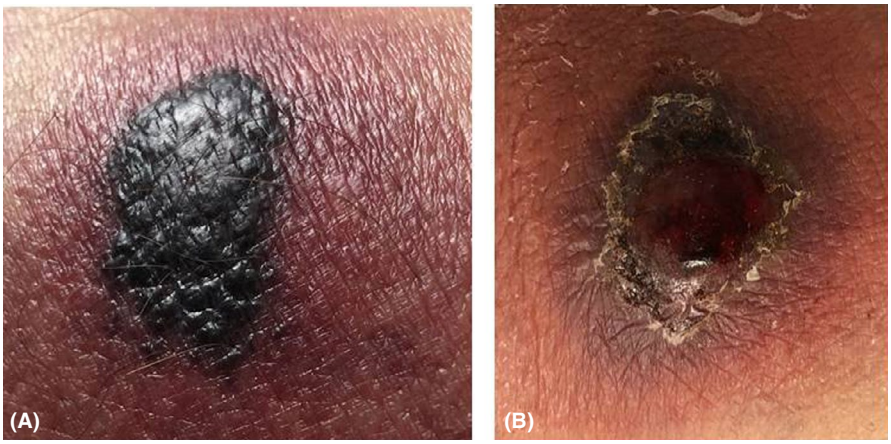

FIGURE 1 initial presentation on left thigh (A) and left leg (B)

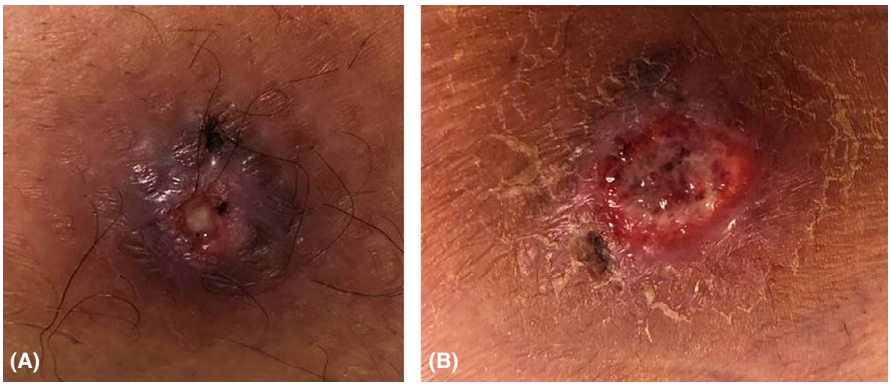

FIGURE 2 clinical improvement after 7 d of antibiotics (A and B)

Ecthyma gangrenosum is a rare cutaneous manifestation of *Pseudomonas* infections.[1-3] *Pseudomonas aeruginosa* is considered as an opportunistic bacteria that rarely causes infection in immunocompetent subjects.[4,5]

Pseudomonas aeruginosa is a nonfermentative, mobile, Gram-negative rod able to produce hydrosoluble pigments (eg, pyocyanin) and different extracellular enzymes (eg, collagenase, elastase, and phospholipase), exotoxins, and endotoxins. EG usually occurs in immunocompromised patients, especially in diseases involving abnormal neutrophil function or in patients with granulocytopenia.

The lesions typically begin with bullae or hemorrhagic pustules that evolve into ulcerations and eschars surrounded by an erythematous halo. The diagnosis is based on the macroscopic aspect as well as on the clinical evolution of the lesions, blood cultures, skin biopsies, and bacteriological analysis of tissue specimen.

Differential diagnosis includes anthrax, erythema multiforme, Herpes infection, Sweet syndrome, and pyoderma gangrenosum.

The presence of *P aeruginosa* is a strong argument for the diagnosis, but several other bacterial and fungal pathogens have also been involved,[5,6] including Gram-negative bacteria (*Aeromonas hydrophila*, *Escherichia coli*, *Citrobacter freundii*, and *Serratia marcescens*), *Gram-positive cocci* (*Staphylococcus aureus* and *Streptococcus pyogenes*), *Gram-negative cocci* (*Neisseria gonorrhea*), and *fungi* (*Aspergillus* sp, *Fusarium solani*, *Candida* sp, and *Scytalidium dimidiatum*).

Of the 167 published EG cases recently analyzed, *P aeruginosa* was detected in 123 (73.65%), and other bacterial etiologies were detected in 29 cases (17.35%). Among the 123 EG cases with *P aeruginosa* etiology, sepsis was described in 72 cases (58.5%) and an absence of septicemia was reported for 51 cases (41.5%).[7] Two distinct clinical presentations have been described: a cutaneous localized form[5-8] and a septicemic form with a poor prognosis and a high mortality rate, around 60% in contrast to 15% for localized EG.[5,6]

Classically, hematological seeding from the gastrointestinal, respiratory, and genitourinary tracts to the skin results in ulcer formation. Nonbacteraemic EG[5-8] may result from direct inoculation of the skin, or possibly through undetected low-grade transient bacteraemia.

Although empirical systemic antibiotics should include antipseudomonal coverage and wound care, there are no specific guidelines in the literature. Bodey et al[9] reported an overall cure rate of 67% for patients receiving appropriate antibiotics but only 14% for those receiving inappropriate antibiotics. Moreover, this retrospective

analysis of *Pseudomonas* bacteremia cases indicated that in cases where optimal antimicrobial therapy was delayed (24-48 hours), the healing rate decreased from 74% to 46%. Surgery may be necessary in case of important necrosis, or abscess. In the event of important neutropaenia, the addition of granulocytic colony-stimulating factor should be discussed.[10]

4 | CONCLUSION

The diagnosis of EG should be evoked in front of maculopapular lesions rapidly evolving to necroting ulcers, particularly in the presence of prolonged neutropaenia or other hematological malignancies.

CONFLICT OF INTEREST

No potential conflict of interest was reported by the authors.

AUTHORS CONTRIBUTION

All authors: contributed to all aspects of the manuscript.

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How to cite this article: Zanella A, Faure C, Merle C, Charollais R, Aubin F. Localized ecthyma gangrenosum without sepsis in a neutropenic patient with a myelodysplastic syndrome—Refractory anemia with excess blasts type 2. *Clin Case Rep*. 2019;7:1754–1756. https://doi.org/10.1002/ccr3.2314