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Ecthyma Gangrenosum in Patient with Bone Marrow Aplasia: A Case Report and Review of the Literature

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Abstract

Background: Ecthyma gangrenosum (EG) is an infrequent and discernible cutaneous disease caused by *Pseudomonas aeruginosa.* In situations where it is associated with septicemia in debilitated patients, the prognosis is usually unfavorable. **Objective:** In this case, we aim to verify risk factors, clinical, bacteriological and therapeutic characteristics of ecthyma gangrenosum and we review the literature to highlight the features of this rare condition and discuss the role of early diagnosis and treatment. **Case Report:** We describe the clinical case of a 4-year-old male with bone marrow aplasia who was presented with characteristic skin lesions of EG and developed sepsis later. **Conclusion:** EG is a cutaneous disease characterized by its aggressive nature. The presence of delayed diagnosis and therapy, along with sepsis, is closely linked to a high mortality rate. Treatment is empirically founded on an aggressive initial approach.

Keywords

Ecthyma Gangrenosum, Child, Bone Marrow Aplasia

1. Introduction

EG represents a distinctive dermatological manifestation associated with a potentially fatal systemic infection due to *Pseudomonas aeruginosa*. This lesion was first mentioned by Barker and was later noted by other authors to be pathognomonic of *Pseudomonas septicemia* [1]. It arises subsequent to bacteremia or subsequent to a primary skin lesion [2]. Less frequent organisms that are responsible are *Pseudomonas cepacia*, *S. aureus*, *E. coli*, *Klebsiella*, *N. meningiti-*

dis, *Aspergillus* species, *Aeromonas* species [3]. Lesions occur due to bacteremia results in to disseminated infectious vasculitis which is characterized by erythematous macules, papules, or nodules with a central hemorrhagic vesicle or bulla that ruptures resulting in a perforated indurated ulcer surrounded by raised oedematous margins with central necrosis and eschar formation. The occurrence of EG in infant is uncommon and primarily observed in the septicemic manifestation. It represents a distinctive dermatological presentation found in individuals with compromised immune systems, specifically those afflicted by malnutrition, malignant disease, or severe hematologic disease, as well as individuals who have received antibiotic or immuno-suppressive therapy [1].

We report the case of a 4-year-old child with bone marrow aplasia in whom the diagnosis of Ecthyma gangrenosum was made. This case highlights the features of this rare condition and discusses the importance of an early, aggressive and multidisciplinary approach.

2. Clinical Observation

We present the case of a 4-year-old male child with bone marrow aplasia, he was admitted to our Pediatric department with 2-days history of fever, oral ulcers on his upper lip, cough and weight loss. He had poor nutritional history.

On admission, he was febrile (39.1˚C), pale, pulse rate was 112/min, a respiratory rate was 26/min and blood pressure was normal. Arterial oxygen saturation (SaO_2) was 100%. Fine inspiratory rales were present on auscultation. At skin level, multiple purple necrotic lesions were observed on the dorsal surface of the left hand, exhibiting distinct stages of evolution. These lesions manifested as rounded, ulcerated, necrotic papules and nodules, each characterized by a central crust Also he had a necrotic ulceration occupying the upper lip extending to the palate then towards the nasal septum (**Figure 1**).

Laboratory tests (**Table 1**) showed pancytopenia (1200 white blood cells/mm^3) with an absolute neutrophil count of 200 cells/mm^3, anemia (8.3 g/dL hemoglobin), and thrombocytopenia (9000 platelets/mm^3). Bleeding time was normal. Erythrocyte sedimentation rate (ESR) was 126 mm/h, elevations in C-reactive protein CRP at 241 mg/l and procalcitonin at 4.1 ng/ml.

Table 1. Our patient's biological findings upon admission.

Laboratory parameter	Initial value	Reference range
White blood cells (*G/L*)	1.2	4.5 - 11
Neutrophils (G/L)	0.2	2.5 - 7
Lymphocytes (G/L)	0.45	1 - 4.8
Platelet count (G/L)	9	150 - 450
Hemoglobin (G/dl)	8.3	10.5 - 13.8
CRP (mg/L)	241	0.2 - 7.5
procalcitonin (ng/ml)	4.1	0.15 - 0.50
ESR (mm/h)	126	10 - 20

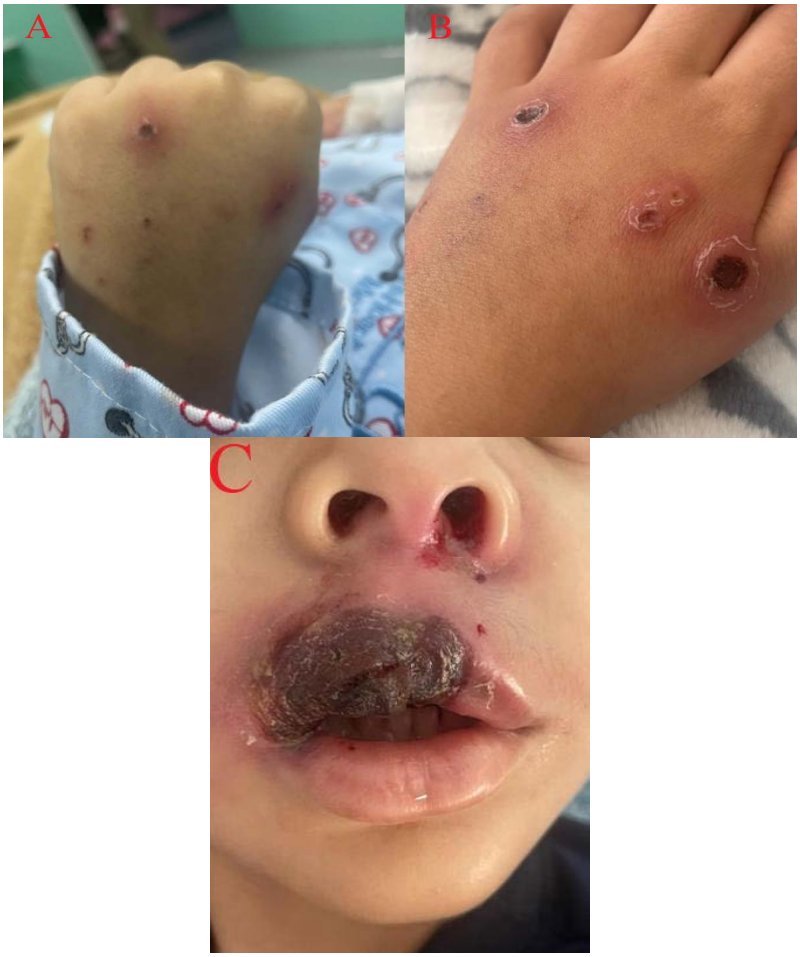

Figure 1. (A) (B) Multiple purple necrotic lesions were visible on the dorsal surface of the left hand; (C) Initial oral lesions of ecthyma gangrenosum. The lesions developed into blackish crusted grangrene in the upper lip.

Serology tests were negative for HIV, herpes simplex virus, cytomegalovirus, Epstein-Barr virus, and mumps/measles/rubella.

Blood culture was positive for *Staphylococcus aureus* repeated samples were taken with mycological and bacteriological studies in order to identify the germ in question and adjust the treatment, returning in favor of *Klebsiella pneumoniae*, *Candida albicans.*

A chest radiography was performed and returned with no abnormalities. The CT neck chest-abdomen-pelvis showed two necrotic collections of the nasal cavity and upper lip responsible for a loss of substance whose most likely caused by infectious origin.

The initiation of Intravenous antibiotic treatment comprising amikacin + imipenem took place at the beginning of hospitalization. According to the results of the repeated mycological and bacteriological studies intravenous voriconazole was also added. The patient received blood and platelet transfusion. Yet, there was no significant improvement of the lesions.

Finally, excision of necrotic plaque was performed (**Figure 2**).

Figure 2. Tissue defects after debridement.

Histopathologic examination of the tissues sampled intraoperatively demonstrated an inflammatory cell infiltrate. Culture for causative bacterial organisms was suggested streptococcal or staphylococcal infection. Antibiotic treatment was continued but our patient experienced a severe decline, resulting in the failure of multiple organs. Despite an intensive resuscitation, progression was rapidly fatal. The patient died secondary to sepsis.

3. Discussion

Ecthyma gangrenosum (EG) is a skin lesion that commonly presents in combination with sepsis caused by *Pseudomonas aeruginosa*, a result of subcutaneous thrombotic vascular lesions. In the etiology of EG it is possible that other bacterial strains may also have an impact: Gram-positive cocci, such as *Streptococcus pyogenes*, *Staphylococcus aureus* and Gram-negative cocci: *Neisseria gonorrhoea* or Gram-negative bacteria: *E. coli*, *Citrobacter freundii*, *Klebsiella pneumoniae*, *Morganella morganii*, *Burkholderia cepacia*, *Pseudomonas stutzeri*, *Serratia marcescens*, *Xanthomonas maltophilia*, *Aeromonas hydrophila*, *Chromobacterium violaceum*. Additionally, instances of EG induced by fungi have also been documented: *Candida albicans*, *Aspergillus fumigatus*, *Fusarium solani*, and *Mucor pusilus* [4]. Kanehiro *et al.* have documented that in Japan, staphylococcal infection is responsible for 60% of cases of EG, while the remaining cases are attributed to *streptococcal* and *P. aeruginosa* infections, in descending order of prevalence [5]. There are two postulated mechanisms identified in the literature that describe the pathogenesis of EG. In the first form, bacteria from a primary infection originating in the genitourinary, respiratory, or gastrointestinal tract travel hematogenously, disseminating through the vasculature to the skin. In the second manifestation, a cutaneous abnormality emerges and microbial infiltration takes place at the precise location of the abnormality, consequently resulting in subsequent septicemia. Our patient was initially presented with fever, and

respiratory symptoms, characteristic of the bacteremic form and the blood cultures were positive. Although the majority of individuals afflicted with EG possess a background of immunodeficiency, the prevailing elements predisposing to this condition are typically acute myeloid leukemia, chronic myeloid leukemia, Hodgkin disease, leukopenia, immunosuppression, chemotherapy, and lymphoma [6].

EG is a rare (with a prevalence of 1.3% - 13%) that has the ability to impact any anatomical location. It typically manifests as painless erythematous macules that become indurated, bullous, or pustular and soon become necrotic, covered by a gray-black eschar surrounded by an erythematous halo. It may also start as a nonspecific rash. Clinically, EG most frequently presents as hemorrhagic bullae or necrotic ulcers that have progressed from painless macules or papules. Often the presenting lesion will exhibit an erythematous halo The patient may experience other constitutional symptoms, along with fever, depending on the severity of the infection and their immune status. Diarrhea and gastrointestinal upset are also frequently linked symptoms [7]. It is crucial to observe these manifestations at an early stage in order to promptly initiate the appropriate antibiotic treatment. Qualitative neutrophil defects represent significant risk factors for sepsis. EG lesions typically manifest before the results of the blood and lesion cultures facilitating the prompt administration of appropriate antimicrobial therapy without delay. On histologic examination, EG arising from *P. aeruginosa* and that arising from streptococcal or staphylococcal infection can be distinguished from each other.

In cases of EG associated with *P. aeruginosa* a distinguishing feature is the presence of hemorrhagic necrosis with little cellular reaction, fibrin thrombus formation, and numerous eosinophilic bacteria in the blood vessel wall and perivascular space are characteristically observed. Conversely, in cases of EG caused by streptococcal and staphylococcal infections, a widespread infiltration of inflammatory cells is generally observed, although the absence of bacteria in the lesions is notable.

Thus, the histopathologic findings in our patient suggested a streptococcal or staphylococcal infection or both rather than *P. aeruginosa*, which was in accord with the clinical manifestations and laboratory results [7].

Nevertheless, many antibiotic regimens have been used, and there are no specific treatment recommendations in the literature. The management of EG consists of three distinct stages. The first stage involves the prompt administration of empirical antibiotic therapy as soon as infection is suspected. Subsequently, once the causative agent has been identified, aggressive antibiotic or antifungal treatment is administered. Finally, surgical excision is often necessary, because EG manifests as a necrotizing soft-tissue lesion [8]. It is important to note that our patient's treatment plan was devised in accordance with these three stages.

Antibiotic therapy with spectrum for *Pseudomonas aeruginosa* involves aminoglycosides, third- and fourth-generation cephalosporins, β-lactam antibiotics, and broad-spectrum penicillins. Prognosis relies on the host and on the degree

of immunosuppression. The disease exhibits a substantial mortality rate. For individuals with EG and septicemia caused by *Pseudomonas*, it ranges from 38% - 77%, whereas for patients without sepsis, it is estimated to be approximately 15% [8]. The presence of neutropenia at the time of diagnosis is the most significant prognostic factor for mortality in cases of invasive forms [9].

Ecthyma gangrenosum is an uncommon infective vasculitis. That could indicate a poor prognosis for immunocompromised patients.

Considering elevated rate of mortality, it is imperative to ensure early detection and expeditious effective treatment.

4. Conclusion

In summary, early detection of EG aids in the provision of adequate empiric antibiotic treatment at an early stage of sepsis and improves prognosis. Surgical excision may be beneficial if no improvement was noted via antibiotic treatment.

Consent

Written informed consent was obtained from the patient parents for publication of this case report and accompanying images.

Author Contributions

All authors contributed to the conduct of this work. All authors also declare that they have read and approved the final version of the manuscript.

Conflicts of Interest

The authors declare no conflict of interest.

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