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Case report

Periocular ecthyma gangrenosum with *Pseudomonas septicemia* in an infant: A case report☆

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ABSTRACT

Ecthyma gangrenosum is a skin manifestation of fatal septicemia. We report a case of periocular ecthyma gangrenosum, which is an uncommonly infected area and rarely reported in infants. A 1-month-old female infant with periocular ecthyma gangrenosum presented with a high-grade fever and acute left medial canthus of the eyelid swelling and erythema. Hemoculture at 6 h confirmed *Pseudomonas aeruginosa* infection. Intravenous and topical antibiotics were administered. Daily dressing of the wound and noninvasive bedside escharectomy were performed. Cosmetically acceptable scar was achieved without additional surgery. The patient was considered to have congenital neutropenia due to persistent neutropenia and severe skin and mucosal infections in her first year of life. Noninvasive debridement of the wound reduces the risk of exposure keratitis, lacrimal drainage pathway damage, and the need for further surgical reconstruction. The cause of compromised immunity in infants with ecthyma gangrenosum should be investigated, and intensive follow-up is recommended.

Introduction

Ecthyma gangrenosum is a rare skin manifestation of fatal septicemia most commonly caused by *Pseudomonas aeruginosa* and opportunistic bacteria. Initial lesions' characteristics are painless red patches in infected areas that rapidly transform into hemorrhagic blisters or gangrenous ulcers with black eschar and surrounding erythematous halo [1–3]. Ecthyma gangrenosum usually occurs in critically ill and immunocompromised adult patients. In pediatric cases, it may affect healthy children or children with congenital immunodeficiency [4–6]. Herein, we report a case of periocular ecthyma gangrenosum, which is an uncommon infected area and rarely reported in infants. The author was HIPAA complaint and adhered to the tenets of Helsinki.

Case presentation

A 1-month-old healthy female infant presented with a high-grade fever, acute eyelid swelling, and erythema in the left medial canthus. The mother noticed redness and yellowish discharge in her infant's left eye for two days. She dropped breast milk into the injected eye owing to her belief that breast milk could treat eye infections; however, the eyelid swelling worsened. Ophthalmic examination revealed swelling and erythema in the upper and lower left eyelid and medial canthal area, with yellowish discharge. There was negative mucoid reflux discharge upon lacrimal sac compression. The conjunctiva and anterior segment of the left eye appeared normal.

On the first day of hospitalization, the patient had a high-grade fever of 39.5 °C. The complete blood count test showed a white cell count of 12.1 $\times$ 10^9 cells/L (normal 3.7–10.0 $\times$ 10^9 cells/L) and an absolute neutrophil count (ANC) of 484 cells/μL. Gram staining of the eye discharge revealed numerous polymorphonuclear cells with a few gram-positive and gram-negative bacilli. We diagnosed the patient with preseptal cellulitis and acute dacryocystitis. The empirical treatment consisted of intravenous ceftriaxone 50 mg/kg/day and ophthalmic moxifloxacin eye drops. However, the progression of redness and swelling in the medial canthus of the eyelid continued. Necrotic tissue developed in 12 h after hospitalization with prolonged high-grade fever. The intravenous ceftriaxone dose was increased to 100 mg/kg/day, and intravenous clindamycin 100 mg/kg/day was added.

Twenty-four hours after admission, the redness in the medial canthal

Abbreviations: ANC, absolute neutrophil count.

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area progressed into a black eschar and gangrene ulcer that extended to the upper and lower left eyelids. The vital signs were unstable, with tachycardia, hypotension, and delayed capillary refill associated with periocular worsening, and the patient had septic shock. Rapid progression of periocular ecthyma gangrenosum is shown in Fig. 1.

Non-contrast computed tomography of the brain and orbital area was performed, and the antibiotic was switched to intravenous piperacillin/tazobactam (400 mg/kg/day piperacillin) and vancomycin (60 mg/kg/day). The complete blood count test on the second day after admission showed a white cell count of 10.1×10^9 cells/L (normal $3.7–10.0 \times 10^9$ cells/L) and a decrease in ANC to 101 cells/μL. A hemoculture at 6 h presented *Pseudomonas aeruginosa,* consistent with the eye discharge swab for culture, and confirmed the pathogen of septicemia. Non-contrast computed tomography revealed a left periorbital soft tissue swelling in the medial part of the left orbit without bony destruction or abscess formation. The left intraconal region was preserved, and no extension to the surrounding sinus or central nervous system was shown (Fig. 2).

On the third day after admission, we performed surgical debridement and examination under general anesthesia. After escharectomy in the medial canthal area, we found necrotic tissue involving the medial regions of the upper and lower eyelids, upper and lower puncta, caruncles, and conjunctival fornix. A medial orbital rim incision was performed, and necrotic tissue was found without capsulated pus formation. The necrotic ulceration extended deep into the pre-septal region of the orbit. The left medial rectus muscle was restricted using a force duction test. We considered minimizing the eyelid and lacrimal passage defects using noninvasive debridement and escharectomy. Intravenous antibiotics were switched to ceftazidime (150 mg/kg/day) and metronidazole (30 mg/kg/day) based on the susceptibility aerobic culture test results and continued for 3 weeks. The topical antibiotics were switched to fortified ceftazidime and amikacin eye drops. Daily dressing of the wound and noninvasive escharectomy were performed, and the wound improved without additional surgery, as shown in Fig. 3.

After sepsis was controlled, we investigated the persistent neutropenia by checking the patient's immunological status. Bone marrow aspiration revealed no arrested maturation at the promyelocyte stage. The absolute lymphocyte counts revealed total, CD4 + , and CD8 + T cell numbers within the normal interpretation. Humoral immunity, NK-cell (CD3-, 16 +, and 56 +), B cells (CD19 +), IgG, IgA, and IgM levels were normal. Complement and neutrophil function, total complement (CH50), and phagocytosis levels were normal. The patient was discharged one month after admission, and congenital neutropenia was considered. We initiated antibiotic prophylaxis with trimethoprim/sulfamethoxazole (5 mg/kg/day TMP) and itraconazole (10 mg/kg/day).

One month later (two months after onset), the medial part of the left eyelid had a small defect that was cosmetically acceptable without reconstruction. The lacrimal drainage system was normal, with no epiphora, as shown in Fig. 3(f). During the coronavirus disease pandemic, the patient was lost to follow-up. After 6 months, the patient had severe diarrhea with perianal gangrene and died of septic shock.

Discussion

Periocular ecthyma gangrenosum is an uncommon cutaneous infection of the eye, characterized by a rapidly progressive necrotizing skin lesion. It is caused by *Pseudomonas aeruginosa, Staphylococcus aureus, Serratia marcescens, Klebsiella spp, Aeromona hydrophilia,* and some fungi. Seventy percent of ecthyma gangrenosum cases are *Pseudomonas aeruginosa*-associated and usually occur in immunocompromised patients [1, 2]. Cases have been reported in adult patients with secondary neutropenia after chemotherapy for hematologic malignancies and immunosuppressive status. Cases in children with primary immunodeficiency, congenital agammaglobulinemia, chronic antibiotic use, and occasionally in healthy children have also been reported [4–6,8].

In the present case, a 1-month-old infant presented with medial canthus redness and a history of breast milk dropped into her eye. No evidence of abnormal lacrimal passage was observed after birth, such as dacryomucocele or congenital nasolacrimal duct obstruction. Progression of distinctive areas from indurated papules to black eschar was seen within 12 h after admission, and clinical septicemia appeared within 24 h of the skin lesion progression. This may be explained by pseudomonal invasion to the blood vessels through elastase, protease, and exotoxin A enzyme activities, which was disseminated and caused the concurrent bacteremia. Early surgical debridement of the necrotic tissue is recommended to eradicate the pathogen, promote wound healing, and ensure proper local antibiotic irrigation. In some cases, ecthyma at the periorbital area could erode and fragment the nasal bone, lacrimal bone, ethmoid, and maxillary sinus, and expose the eyeball [3]. In the present case, noninvasive debridement was performed to avoid large cutaneous defects and lacrimal injuries. The eschar was removed and irrigated daily with fortified ceftazidime and amikacin eye drops. Wound improvement in the child was acceptable without requiring any reconstructive surgery.

Periocular ecthyma gangrenosum in infants has rarely been reported and is usually associated with primary immunocompromised status [4–6,8]. Skin lesions begin as indurated papules and then rapidly progress to black eschar and clinical worsening due to *Pseudomonas septicemia*. Early recognition, appropriate antibiotic treatment, and debridement are crucial to reduce morbidity and mortality. Noninvasive debridement reduces the risk of lacrimal system damage and the need for surgical reconstruction in the future.

The evaluation of immunological function is important, especially in defining the possibility of primary immunodeficiency and preventing recurrent infections. Immunological tests performed in this patient did not reveal cellular or humoral complement deficiency or neutrophil dysfunction. Considering the persistent neutropenia over a one-month period with periocular ecthyma gangrenosum ulcer and pseudomonal septicemia, we postulated that congenital neutropenia is most likely to be the cause, although the arrest of promyelocyte stage in the bone marrow was not observed [7]. We prescribed prophylactic antibiotics and planned a follow-up. The patient was lost to follow-up during the coronavirus disease spread. The patient was reported to have recurrent severe infections (diarrhea with perianal gangrene and sepsis), which supported our postulate that the patient was immunocompromised.

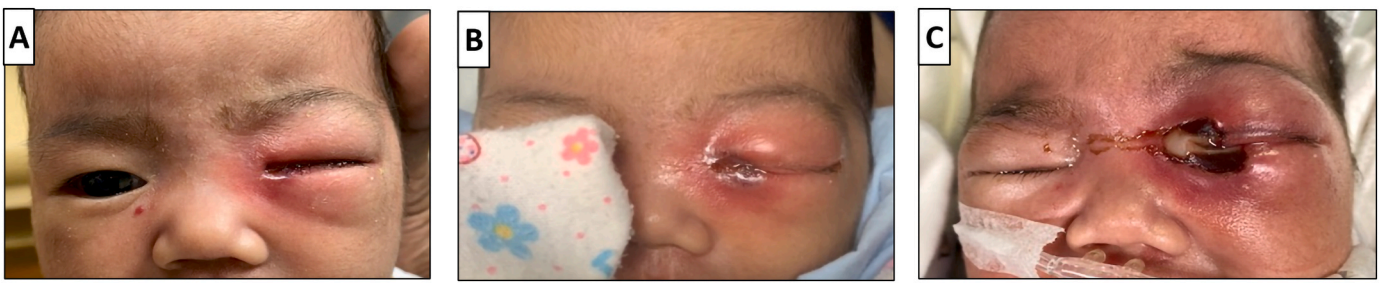

Fig. 1. Development of periocular ecthyma gangrenosum (A) at admission, (B) 12 h after hospitalization, and (C) 24 h after hospitalization.

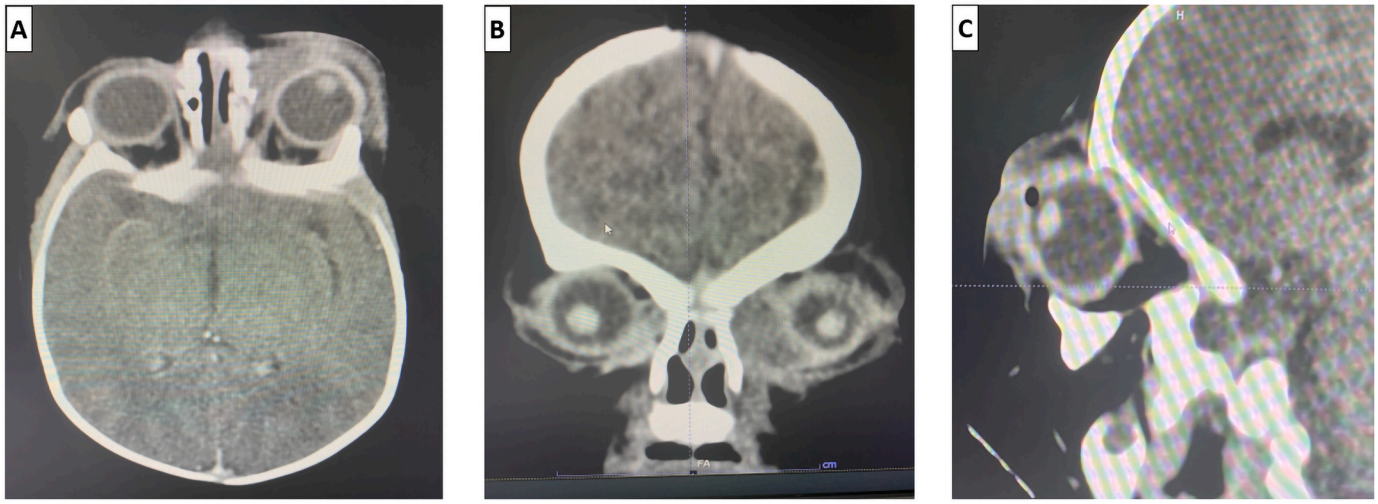

Fig. 2. Non-contrast computed tomography revealed left periorbital soft tissue swelling and haziness in the pre-septal region and medial part of left orbit.

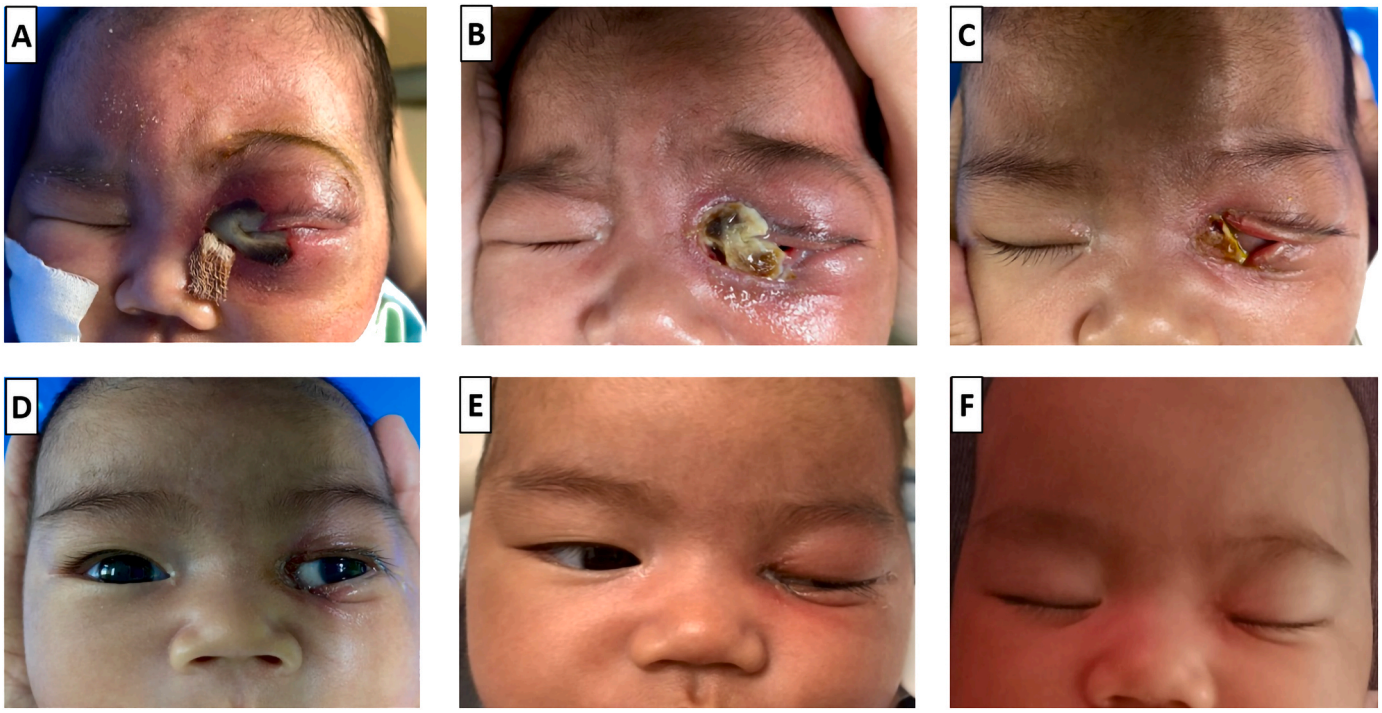

Fig. 3. Progression of the wound after noninvasive escharectomy at (A) 1 day, (B) 1 week, (C) 2 weeks, (D) 3 weeks, (E) 1 month, and (F) 2 months.

Investigation for immunodeficiency and intensive follow-up are recommended for infants with ecthyma gangrenosum.

Ethics approval and consent to participate

Ethics approval and consent to participate was approved by the research Ethics Review Committee of Queen Sirikit National Institute of Child Health.

Consent

Consentfor publication Parentsgave the consent for publication. Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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CRediT authorship contribution statement

Supawan Surukrattanaskul: Data curation, Writing – original draft, Conceptualization. **Rosana Pittayapongpat:** Visualization, Supervision, Resources. **Nutsuchar Wangtiraumnuay**: Writing – review

& editing, Data curation, Supervision.

Declaration of Competing Interest

All authors had no financial disclosure.

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Authors contributions

All authors attest that they meet the current ICMJE criteria for Authorship. SS and RP analyzed and interpreted the patient data regarding the periocular gragrenosum. SS and NW reviewed literature and was a major contributor in writing the manuscript. All authors read and approved the final manuscript.

Competing interests

Supawan Surukrattanaskul, Rosana Pittayapongpat, and Nutsuchar Wangtiraumnuay declare that they have no competing interests.

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