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CASE REPORT

WILEY

Eyelid and periorbital ecthyma gangrenosum due to *Pseudomonas aeruginosa* in an infant

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1 | INTRODUCTION

Ecthyma gangrenosum (EG) is a rare invasive cutaneous infection primarily caused by *Pseudomonas aeruginosa* in immunosuppressed patients. It is characterized by a painless necrotic ulceration with a surrounding erythematous halo and is usually associated with *Pseudomonas aeruginosa* septicemia. EG is extremely rare in the periorbital region, and there have been no other reported infant cases with orbital bone erosion.

2 | CASE REPORT

A 10-month-old infant presented with fever of 39.3°C and right eyelid swelling and erythema. In the first hospital, blood tests showed a white cell count of 1.87×10^9/L (normal [4.0-10.0] $\times 10^9$/L) and antiviral treatment was used; however, fever was not controlled. The eyelid swelling progressed and ruptured with exudation, becoming necrotic when she was transferred to our hospital on Day 5 after the onset (Figure 1A). Blood tests showed a white cell count of 0.31×10^9/L (normal [4.0-10.0] $\times 10^9$/L), red cell count of 2.78×10^{12}/L (normal [3.5-5.5] $\times 10^{12}$/L), thrombocytopenia (platelets: 52×10^9/L; normal [150-400] $\times 10^9$/L), and markedly elevated C-reactive protein (>170 mg/L; normal <8 mg/L). Noncontrast computed tomography revealed right periorbital soft tissue swelling, thickening of the inferior and medial rectus, and blurring of the medial and inferior portions of the orbital wall (Figure 2A). Magnetic resonance imaging (MRI) showed significant periorbital soft tissue swelling that involved both fat and fascia (Figure 2B). Further investigation for underlying disease included virology screens, chest X-ray, and heart and abdominal ultrasounds, all of which were normal. The parents denied a history of congenital dacryocystitis or sinusitis. However, the patient had a history of scalp ulceration following a mosquito bite at the age of 4 months, which was diagnosed as a local *Pseudomonas aeruginosa* infection.

Bone marrow biopsy results showed extremely reduced myeloproliferation and toxic particles in some mature cells of the granulocyte system. Red cells had generally normal morphology. Bone marrow suppression caused by severe infection was considered. Cultures of blood, bone marrow, and conjunctival secretions all confirmed the growth of *Pseudomonas aeruginosa*. Immunological studies showed decreased B and NK cells.

The infant was given intravenous fluids, and systemic broad-spectrum antibiotics (amoxicillin and sulbactam sodium, meropenem, and metronidazole) were started. Intravenous immunoglobulin was used, and intravenous infusions of platelets, red blood cells, and recombinant human granulocyte/macrophage colony stimulating factor were given.

On Day 12, the patient defervesced, and most of the blood parameters improved (white cell count, 1.44×10^9/L; red cell count, 3.01×10^{12}/L; platelet count, 48×10^9/L; and C-reactive protein, 57 mg/L). However, the eyelid and canthus skin rapidly necrosed despite systemic anti-inflammatory treatment. The black eschar with surrounding erythema covered half of the right eyelid, and the nasal cornea was exposed (Figure 1B).

With sepsis under control, surgical debridement and eyelid suturing were performed under general anesthesia on Day 15. The necrosis involved the entire nasal preorbital soft tissues, the superior and inferior orbital walls, and the ethmoid and maxillary sinuses. The medial nasal bone and the lacrimal bone were eroded and fragmented

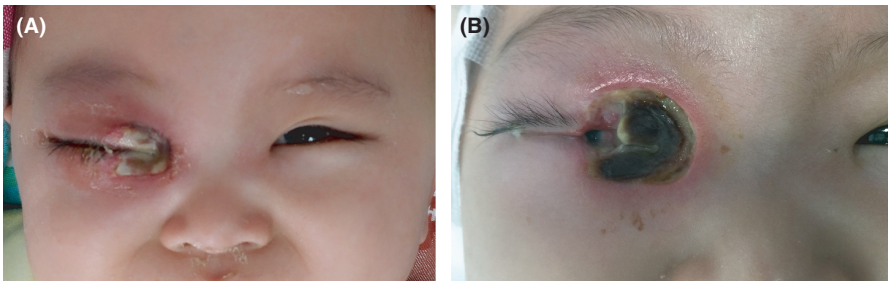

FIGURE 1 A, On Day 5 after onset, the right eyelid showed necrosis after swelling and erythema. B, The appearances on Day 12 after onset were well-demarcated central necrotic area surrounded by erythema. The black eschar is typical characteristic of ecthyma gangrenosum. The nasal cornea was exposed

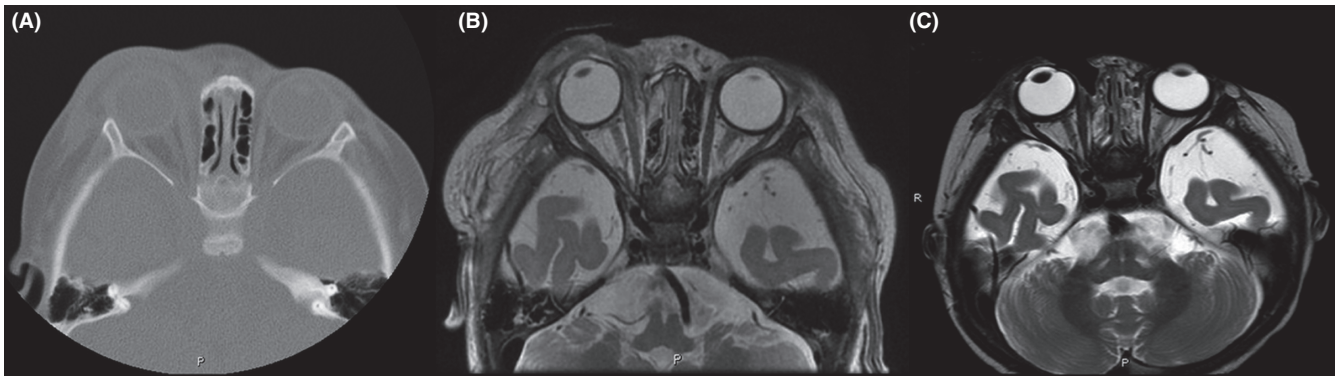

FIGURE 2 A, Noncontrast CT revealed right soft tissue swelling in the front of the orbit. The medial rectus was thickening. Orbital bone wall of media blurred. B, MRI shows significant swelling of soft tissue around the right eye, fat, and fascia. C, Five days after surgery, MRI showed improved periorbital, eyelid, and maxillofacial swelling, and low signal from the right nasal orbit

(Figure 3A). The nasal sclera was exposed, and a mild yellow scleral infiltrate was present. Histological examination showed degeneration and necrotic tissue with many inflammatory cells (Figure 3B). Within 3 days of debridement, the eyelid necrosis was controlled (Figure 4A). MRI showed improved periorbital, eyelid, and maxillofacial swelling, and low signal from the right nasal orbit (Figure 2C).

Irrigation with antibiotics (ceftazidime and tobramycin) was used for daily dressing changes until the cavity became very narrow, and systemic antibiotic treatments (meropenem, cefoperazone sulbactam, and cefpodoxime proxetil tapered) were continued for 1 month. On Day 30, the white cell count was 2.56×10^9/L; platelets, 195×10^9/L; and CRP, <8mg/L. Blood culture showed no bacterial growth. On Day 33, 18 days after debridement, superficial corneal ulceration and infiltration appeared (Figure 4B). We then placed a corneal bandage lens on the eye, and the cornea improved 1 week later. Her eyelid progressed to a relatively satisfactory appearance without additional surgery after 10 months of onset (Figure 4C). In the follow-up period, her white cell count was always below 3.0×10^9/L and intravenous immunoglobulin was used every 2 months.

3 | DISCUSSION

Ecthyma gangrenosum (EG) is a rare, distinctive skin disorder associated with potentially fatal underlying pseudomonal sepsis that usually occurs in neutropenic or immunocompromised patients.[1] It represents a bacterial vasculitis, a rare cutaneous finding manifested by bacterial invasion of blood vessel walls and subcutaneous tissue, which is observed in 1%-3% of patients with pseudomonal sepsis.[1,2] These lesions most often occur in the gluteal and perineal regions

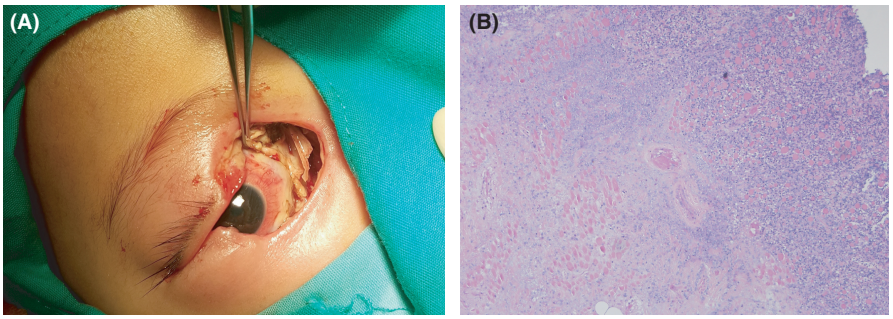

FIGURE 3 A, The nasal bone and lacrimal bone were eroded and fragmented. Sclera exposed. B, Histological examination of the removed tissues shows degeneration and necrotic tissue with many scattered inflammatory cells. (hematoxylin-eosin, ×25)

FIGURE 4 A, Three days after surgery, the wound margins were clean and nasal sclera was exposed. B, Superficial corneal ulcer and infiltration appeared repeatedly. C, Ten months after the surgery, the patient's eyelid self-repaired very well and cornea was transparent

(57%) or on the extremities (30%). Facial involvement is reported in about 6% of cases and can lead to skin defects that require reconstruction.[3] Between 1974 and 2016, only two cases of facial EG in infancy were reported: one involved the eyelid and the other, the lips.[4,5] Several adult cases involving the eyelid and orbit have been reported in patients with leukemia or chemotherapy-related myelosuppression.[6,7]

In contrast to more commonly recognized cutaneous problems associated with pseudomonal infections (eg, folliculitis, wound infection, and green nail syndrome), the appearance of EG is distinctive: small indurated vesicular papules progressing rapidly to infarcted necrotic areas with a black eschar and surrounding erythema. Biopsy usually shows nonspecific inflammation of vessels and necrosis. Diagnosis is made clinically along with a positive skin, urine, or blood culture.[8]

EG is a necrotizing vasculitis that primarily affects neutropenic patients, especially those with an underlying malignancy.[4] In children, additional associations have included congenital agammaglobulinemia, aplastic anemia, congenital megacolon, drug-related myelosuppression, and chronic antibiotic use.[9] It can also occasionally affect healthy children.[1]

Based on the history and clinical and genetic testing, leukemia, aplastic anemia, cancer, and drug-related immunosuppression were ruled out in this infant. The only laboratory finding was decreased B and NK cells, suggesting a humoral immunodeficiency. Thus, this appeared to be a case of primary immunodeficiency.

Early recognition and prompt treatment with antipseudomonal antibiotics is vital to reduce morbidity and mortality. The number of cutaneous lesions, time of initiating antibiotic therapy, and neutrophil count are directly correlated with prognosis. The mortality rate in nonsepticemic cases is generally <15%, whereas the mortality rate in septicemic cases is extremely variable but can be as high as 96%.[1,10]

The early recognition of the typical lesions of EG in the setting of orbital cellulitis and prompt, appropriate treatment are very important. Surgical debridement of all necrotic tissue is generally recommended. This case involved corneal exposure, so a therapeutic bandage lens was used to prevent exposure keratitis. Three months later, the corneal epithelium was in good condition. Reconstructive surgery is planned for the future.

CONFLICT OF INTEREST

The authors have no conflict of interests relevant to this article.

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