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Life-threatening periocular pseudomonal necrotizing fasciitis in an immunocompetent infant

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A B S T R A C T

Purpose: Necrotizing fasciitis, a severe soft tissue bacterial infection, is uncommon in the periocular region due to its rich blood supply. This report highlights a rare case in an immunocompetent infant.

Observations: A 10-month-old immunocompetent female exhibited fever, irritability, and right eyelid swelling post-fall. Despite initial treatment with cephalexin for presumed cellulitis, her condition rapidly deteriorated, suggesting necrotizing fasciitis. She stabilized after receiving broad-spectrum intravenous antibiotics and aggressive surgical debridement but later required orbital exenteration due to extensive tissue necrosis.

Conclusions and Importance: Periocular necrotizing fasciitis is exceedingly rare, particularly in immunocompetent individuals. Prompt diagnosis and treatment are critical to mitigate morbidity and mortality.

Necrotizing fasciitis (NF) is a life-threatening bacterial infection involving soft tissues with overall mortality as high as 34 %.[1] Infection leads to rapid necrosis of subcutaneous tissue and muscle fascia and can swiftly spread to adjacent tissues, escalating to sepsis.[2] NF commonly emerges following penetrating skin injuries, though it can also, albeit rarely, develop after blunt trauma.[3] The infection primarily occurs in the lower and upper extremities, as well as the perineum.[4] NF seldom affects the head and neck area, particularly the periocular region, due to the region s rich collateral blood supply, which typically confers some degree of protection.[5–7] It is in general a rare disease, and even less commonly occurs in the pediatric population.[4] The typical bacteriology for NF represents a spectrum of gram positive bacteria, most commonly Group A streptococci and viridans streptococci.[3,5–8]

This report details a rare and insidious presentation of periocular necrotizing fasciitis that followed blunt head trauma in a previously healthy 10-month-old female infant.

A 10-month-old girl presented with a one-week history of fever and irritability, which evolved into right eyelid swelling (Fig. 1) two days after an unwitnessed fall. Initially treated with cephalexin at another facility, her condition worsened within 24 hours. Blood and urine cultures tested positive for *Pseudomonas aeruginosa*.

Upon transfer to our tertiary hospital, she exhibited severe sepsis and liver failure. The pediatric Intensive Care Unit team initiated treatment with vasopressors and a regimen of broad-spectrum intravenous (IV) antibiotics, including vancomycin, cefepime, clindamycin, and gentamicin. In the setting liver failure she became fluid overloaded, and she appeared diffusely edematous on physical exam. Eschar formation over the right medial canthus raised suspicion of necrotizing fasciitis (Fig. 2). A complete corneal epithelial defect without infiltrates or hypopyon was also noted in the same eye. There was no vitritis on B-scan ultrasound.

She was taken emergently to the operating room for debridement. During the operation, it was observed that the superficial tissue planes from the brow to the eyelid crease, and from the nose to about 2 cm past the lateral canthus, were necrotic. The orbicularis and surrounding tissues were noted to be grossly ischemic. Tissues were removed until a bleeding edge was encountered and the wound was copiously irrigated with normal saline. The infection appeared to extend beyond the hairline within the superficial temporalis fascia and temporalis muscle. Due to the extensive loss of upper and lower eyelid anterior lamella, a complete temporary tarsorrhaphy was performed.

Tissue specimens demonstrated an accumulation of bacterial forms, and necrosis of epidermis, dermis and fibroadipose tissue. Bacterial culture of the periocular tissues and cornea identified an abundance of *Pseudomonas aeruginosa*. The patient was continued on IV tobramycin and piperacillin/tazobactam. The cornea was treated with fortified

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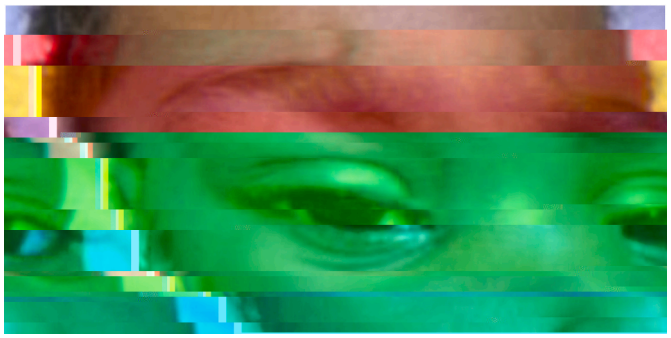
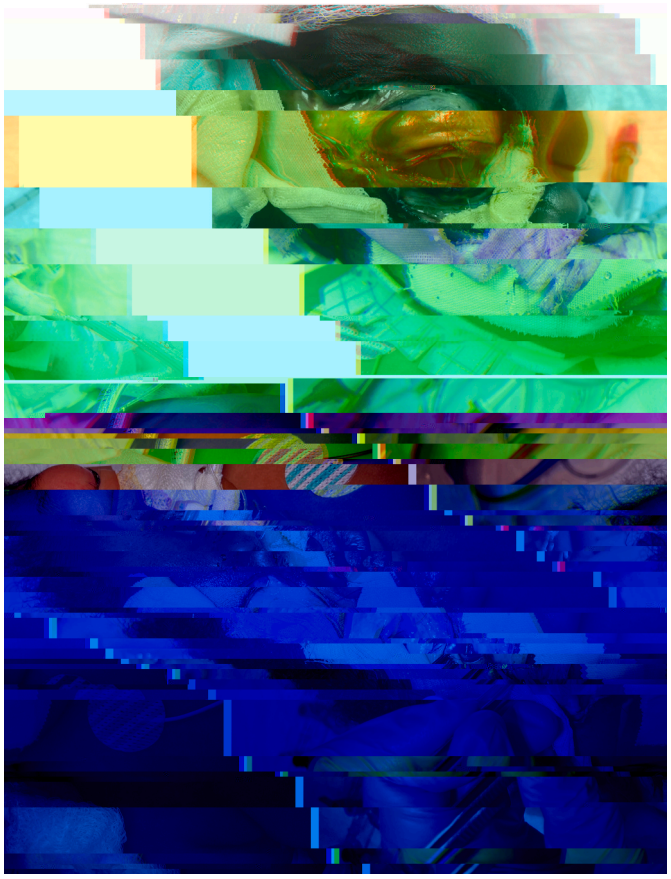
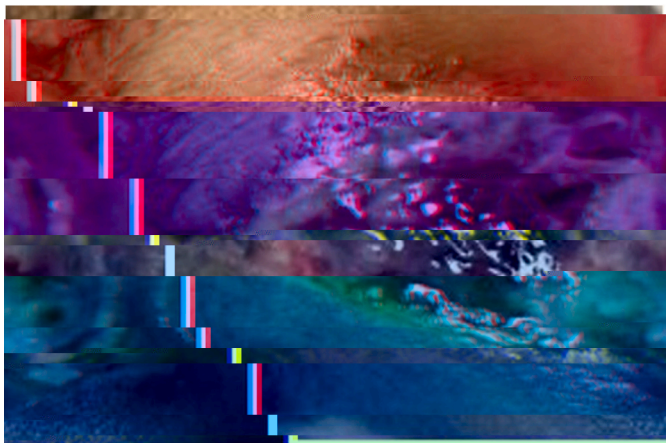


Patient prior to hospital admission.

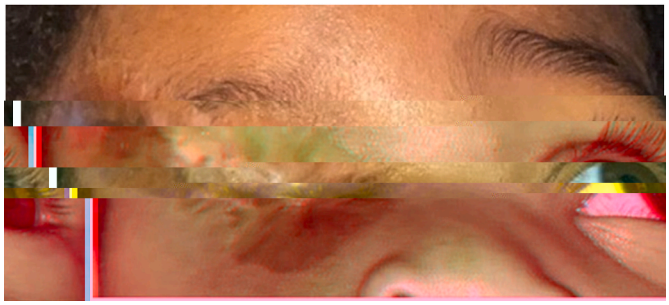
Patient upon transfer to tertiary hospital.



Limbus to limbus corneal epithelial defect and hypopyon.



Ischemia of anterior orbit.



Post-operative month 6.

topical tobramycin, ceftazidime, and prednisolone. Over the ensuing two days the patient s systemic condition stabilized and she was weaned off vasopressors. However, four days later she developed new corneal infiltrates and a hypopyon (Fig. 3). The patient was examined serially twice daily. The decision to return the patient to the operating room for additional debridement was made in dialogue with the primary medical team and was based on the patient s systemic condition as well as the examined tissues. A Morgan lens was placed to improve delivery of the topical medications (Fig. 4).

Her cornea continued to deteriorate, and she was taken to the operating room for examination, additional surgical debridement of the periocular tissues, and placement of an Integra dermal regeneration template. Corneal cultures from this session identified *Candida parapsilosis*. She was weaned off prednisolone drops and started on topical amphotericin drops.

Her cornea deteriorated further and by 10 days, there was a complete limbus to limbus corneal infiltrate and a corneal perforation. Previously perfused eyelid tissue was noted to be ischemic. There was notably poor perfusion of conjunctiva and Tenons. She was taken again to the operating room for a penetrating keratoplasty. During this procedure it was noted that there was further necrosis of the eyelid tissue including the posterior lamella, leading to inadequate coverage of the ocular surface. Therefore, a conjunctival and Tenons flap was fashioned over the cornea. Within 20 days, the conjunctival graft had necrosed leaving the cornea exposed.

The patient was again taken to the operating room to further debride the periocular tissues. The anterior orbit was noted to be necrotic (Fig. 5). There was minimal viable conjunctiva. The superior rectus was divided and advanced over the cornea to provide coverage. An Integra was placed over the entire periocular region and cornea. Given the presence of necrotic orbital tissue, low visual potential and delayed wound healing, a subtotal exenteration was recommended. On hospital

Reported cases of periocular necrotizing fasciitis attributable to *Pseudomonas aeruginosa*.

Reference	Age	Immune Status	Trauma	Sepsis	Debridement
Ganesh 2007	2 weeks	Leukocyte adhesion deficiency	No	No	No
Steinkogler 1988	6 weeks	Congenital cellular immunity defect	No	Yes	No
Scheepers 2010	16 months	HIV/AIDS (CD4 count of 254 x 10^6/litre)	No	Yes	No
Yeh 2019	14 years	Chronic immunosuppression due to systemic lupus erythematosus	No	No	Yes
Rodriguez-Gonzalez 2013	53 years	Small cell lung carcinoma	No	No	Yes
Hulten 2009	60 years	HIV/AIDS	No	No	No
Lattman 1998	62 years	Neutropenia	No	No	No
Lee 2021	62 years	Chronic Immunosuppression due to kidney transplant	No	No	Yes
Poitelea 2005	68 years	Immunocompetent	No	No	Yes
Mutamba 2013	68 years	Leukemia	Yes	No	Yes

HIV/AIDS, human immunodeficiency virus/acquired immunodeficiency syndrome.

day 50, the patient underwent exenteration. At post-op month 6 the patient s orbit was fully epithelialized without evidence of sino-cutaneous fistula (Fig. 5). At last follow up 1 year post-op the patient was healthy and had met all milestones.

NF presents significant diagnostic challenges due to its aggressive nature and the subtlety of its early symptoms. The infection predominantly affects fascia and subcutaneous tissue, often sparing the overlying skin initially, which can lead to underestimation of its severity. This characteristic emphasizes the need for a high index of suspicion, particularly in cases with atypical presentations where classic signs and symptoms may not be apparent.

While *Streptococcus* and *Staphylococcus* species are the more common culprits in NF, *Pseudomonas aeruginosa* presents a unique challenge. Previously documented cases of periocular Pseudomonal NF are noted in Table 1.[5–14] These cases are particularly challenging due to the pathogen s ability to create biofilms, invade the host via flagella and pili, and protect itself from complement via lipopolysaccharide (LPS), which itself can trigger a cytokine storm leading to sepsis and shock.[15,16]

Typically associated with infections in immunocompromised individuals, *Pseudomonas* s occurrence in immunocompetent patients, especially in well-vascularized areas like the periocular region, is rare.[8] In this patient an immunology workup that included a total hemolytic complement activity (CH50) assay and a granulocyte oxidative burst assay to evaluate for complement deficiency or neutrophil deficiency showed normal results. Further exome sequencing did not identify any pathological variants for primary immunodeficiencies.

The management of NF requires a dual approach involving both early antibiotic therapy and surgical debridement.[6] Antibiotics alone do not suffice, especially in cases with rapid disease progression, systemic toxicity, or bacteremia. Surgical intervention, aimed at controlling the spread of infection, becomes indispensable in such scenarios. Our case required escalating to broad-spectrum IV antibiotics and urgent surgical intervention, highlighting the utility of an aggressive treatment strategy in the face of rapid disease progression.

Diagnosing NF in infants and young children poses additional challenges. These patients may not effectively relay the presenting history or communicate their symptoms. Infants also have a body surface area approximately three times larger than adults when adjusted for body weight.[17] This characteristic makes NF especially critical, as the spread of infection can rapidly progress to life-threatening stages as was the case in this patient.[18]

The route of infection leading to the periocular region in this patient appears to be multifactorial. Prior to the appearance of any external signs, the patient was noted to be febrile and irritable. Positive blood and urine cultures indicated a possible hematogenous spread originating from a urinary tract infection. Additionally, the patient s history of blunt trauma may have created a portal of entry for the infection. These combined factors likely contributed to the development of the infection in the periocular region. This case underscores the importance of considering multiple potential sources and mechanisms in the evaluation and management of complex infectious cases in infants.

The initial surgical interventions aimed to stabilize the patient and limit morbidity. Although exenteration in his case was ultimately required due to progression of the infection and likely fungal superinfection, the primary objective surgery is to debride necrotic tissue and allow for host immune response and antibiotics to control the residual infection. Subsequent surgical plans for the right socket include orbital prosthesis when she is old enough to maintain one.

This report emphasizes the challenges in diagnosing NF in atypical regions and populations, particularly when caused by atypical pathogens like *Pseudomonas aeruginosa*. The unusual progression in an immunocompetent infant following blunt trauma coupled with the atypical and progressive periocular involvement, makes this case particularly noteworthy. It highlights the need for vigilance and emphasizes the importance of an interdisciplinary approach in management of both the local and systemic disease. Though the visual outcome was poor in the present case, it is likely that surgical therapy played a role in her systemic recovery to full function.

Consent to publish this case report has been obtained from the patient s guard in writing.

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All authors attest that they meet the current ICMJE criteria for Authorship.

Writing – review & editing, Writing – original draft, Investigation, Conceptualization. Writing – review & editing, Writing – original draft, Conceptualization. Writing – review & editing, Writing – original draft, Formal analysis, Data curation, Conceptualization.

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

None.

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