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Periorbital Necrotizing Fasciitis – An Action Plan for Rapid Diagnosis and Management

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Life-threatening necrotizing fasciitis most commonly affects the limbs, perineum, and abdominal wall; however, it will rarely present in the skin and soft tissue around the eye. Although the mortality rate is lower with periorbital necrotizing fasciitis (PONF) than that associated with other presentations, saving the patient and eye requires prompt identification and treatment, preferably by a multidisciplinary team. This issue of *Ophthalmology Rounds* presents a case of trauma-induced PONF to illustrate early and effective management.

Case Summary

A 34-year-old otherwise healthy man presented with severe periorbital and facial erythema and swelling 3 days after being kicked in the face during a soccer match. One day after the injury, he noticed transient diplopia in upgaze, but denied pain, fever, or constitutional symptoms. The next day, the patient experienced an escalation of severe pain in the left periorbital area and progressive swelling of the lower eyelid and upper cheek. He sought emergency care at an outside hospital. A computed tomography (CT) scan demonstrated marked soft-tissue swelling, subcutaneous erythema, and a left orbital floor fracture with a prominent air-fluid level in the maxillary sinus. He received analgesics and a single dose of oral steroids, and was discharged.

A few hours later, the patient returned to the outside hospital with fever of 39°C, severe uncontrolled periorbital pain, and progressive swelling and erythema involving the left upper and lower eyelids and cheek with subtle blistering of the inferior eyelid skin. He was admitted and treated with intravenous antibiotics, but was discharged on no antimicrobial therapy the following day. Three days after being discharged, he reported persistent periorbital pain, dysphagia, and malaise.

At presentation to our hospital, the patient had tense erythematous swelling of the left upper and lower eyelids with extension to the cheek and lower face. There was blistering and bullae in the medial upper and lower eyelid skin with weeping serous-purulent exudate and adjacent dusky appearance, but no evidence of crepitus, anesthesia, or paresis of facial musculature (Figure 1).

Examination of the left eye was possible with Desmarres retractors and revealed conjunctival injection and chemosis. The patient's visual acuity was 20/400 in the left eye, but he had normal colour vision and no afferent pupillary defect. He had severely limited extraocular motility and his intraocular pressure was 40 mmHg. He had normal vital signs and oxygen saturation, and was afebrile. A CT scan was repeated and demonstrated marked interval increase in extensive soft-tissue swelling on the left side of the face, and inflammatory stranding within the intraconal space with thickening of the inferior rectus muscle but no abscess (Figure 2).

There were rare locules of gas in the subcutaneous tissue and orbit, but it was unclear if these originated from maxillary sinus or were generated by the infectious process. A swab of purulent material demonstrated Group A *Streptococcus* (GAS). Laboratory studies demonstrated marked leukocytosis (21 cells/mm^3) and elevated C-reactive protein (CRP; 27 mg/dL), anemia (hemoglobin 128 g/L), and elevated liver function tests. Glucose, electrolytes, and creatinine remained in the normal range. The Laboratory Risk Indicator for Necrotizing Fasciitis (LRINEC) score was 6 (*see Discussion*), which indicated a significant risk of necrotizing fasciitis.

The patient was admitted to hospital and treated with intravenous fluids, piperacillin-tazobactam, and clindamycin. Over the subsequent hours, he was monitored closely for evidence

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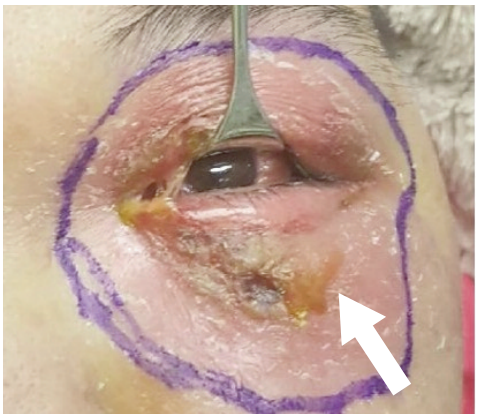
Figure 1. The patient presented to our hospital with tense swelling and erythema of the upper and lower eyelids with extensive upper and lower facial swelling. Bullae and necrosis of the medial upper and lower eyelid skin are shown (arrow). A Desmarres retractor was required to examine the chemotic eye. The erythematous region was marked to facilitate monitoring of progression.



of clinical or systemic progression. He remained afebrile with stable vital signs and there was improvement in leukocytosis and inflammatory markers within 12 hours of treatment. Periorbital erythema and edema began to recede, but the previously blistered skin evolved into necrosis. Otorhinolaryngological consultation was sought; no drainage of the affected sinuses was deemed necessary in light of the clinical improvement. Bedside debridement of the necrotic skin was performed approximately 36 hours after admission, revealing healthy orbicularis muscle. Intravenous methylprednisolone was added, which dramatically improved the periorbital edema and erythema (Figure 3).

Figure 2. Coronal computed tomography of the orbits demonstrated extensive soft-tissue swelling and subcutaneous gas on the left side of the face (arrow). A left orbital floor fracture with inflammatory stranding and a locule of gas within the intraconal space is evident. Opacification of the left maxillary sinus and ethmoid air cells is seen.

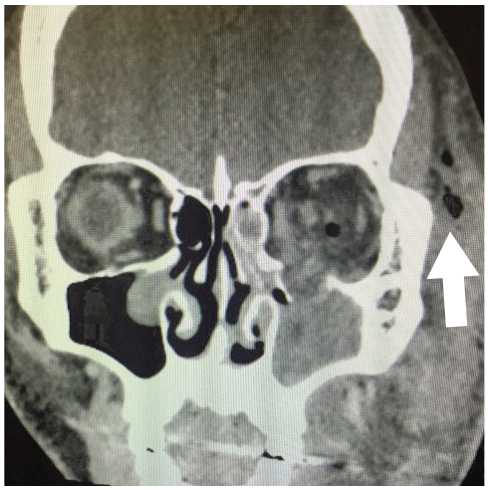
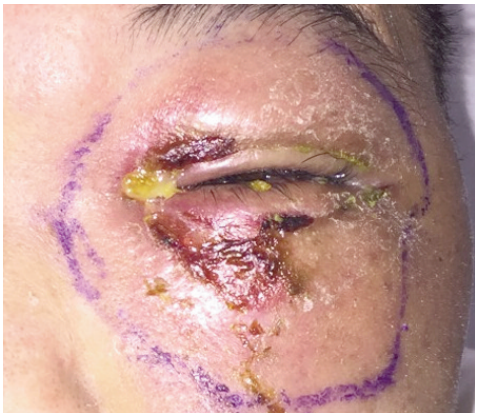


Figure 3. Three days after presentation, following treatment with intravenous antibiotics and bedside debridement, the zone of erythema and edema had receded. Healthy orbicularis muscle can be seen deep to the debrided tissue.

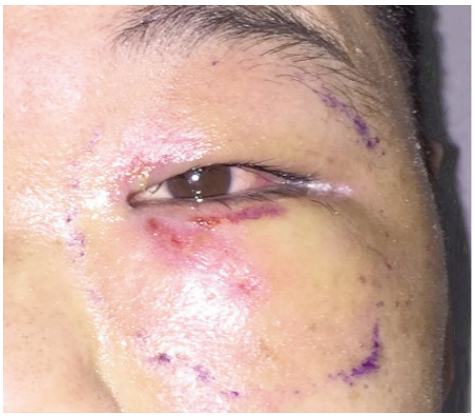


The patient was discharged from hospital 9 days after admission with markedly improved periorbital swelling and near complete re-epithelialization of the previously necrotic skin (Figure 4). Upon discharge, his visual acuity was 20/20 in the left eye with nearly normal extraocular motility and normal intraocular pressure.

Discussion

Necrotizing fasciitis is a rapidly progressive, potentially fatal, and disfiguring infection that spreads along subcutaneous soft-tissue planes and causes necrosis of the skin and soft tissue. The limbs, perineum, and abdominal wall are the most frequently involved sites, while periorbital involvement is rare. The mortality of periocular necrotizing

Figure 4. Five days after presentation, re-epithelialization of the debrided regions and further improvement in edema and erythema are seen.



fasciitis (PONF) is lower than that reported at other anatomical sites, ranging from 3%–14% in various case series.[1] Necrotizing fasciitis has been divided into 2 subgroups with unique microbiology. Type I presents in immunocompromised patients and is attributed to polymicrobial infection with mixed aerobic and anaerobic bacteria. Type II has an acute or fulminant course which may be associated with systemic shock and multiorgan failure and is associated with β-hemolytic GAS and, rarely, *Staphylococcus* species.[2] The streptococcal M protein is a virulence factor produced by GAS and may explain the increased severity of cases of necrotizing fasciitis associated with this organism.

Risk factors

Damage to the skin by trauma or surgery is the most important risk factor for the development of necrotizing fasciitis. In a series of 29 patients with PONF described by Rajak et al,[1] 64% of patients recalled a preceding injury within the past 2 weeks which involved minor trauma, a surgical incision, or soft-tissue or sinus infection. In approximately 50% of patients with GAS-associated necrotizing fasciitis, however, infection may arise deep in the soft tissues at sites of nonpenetrating trauma.[3] Transient bacteremia arising from the nasopharynx or sinuses enables organisms to reach the site of blunt trauma where released exotoxins compromise the local microvasculature and instigate the infection.[3] Chronic immunosuppression is another major risk factor for the development of PONF; 55% of patients in the Rajak et al series had evidence of chronic immunosuppression such as longstanding diabetes mellitus, solid organ or hematologic malignancy, chronic alcohol abuse, or autoimmune disease requiring immunosuppressive medication.[1]

Clinical presentation

PONF presents with a rapid onset of periorbital pain, swelling, and erythema over a few days. Early on, the skin may be pale and tense; as the infection spreads along subcutaneous fascial planes, the skin becomes cyanotic and dusky with irregular erythematous borders and serosanguinous bullae.[2] Skin and subcutaneous necrosis is pathognomonic for necrotizing fasciitis but may take hours to days to develop. Therefore, a high degree of clinical suspicion must exist, particularly when the patient describes rapid progression and pain out of proportion to findings on examination. The infection may descend to the cheek or cross through subcutaneous tissue planes to involve the contralateral periorbital skin and eyelids. Crepitus or gas formation in the subcutaneous tissues is typical of polymicrobial necrotizing fasciitis and is not typically seen in GAS-associated necrotizing fasciitis.[1,3] Ptosis, proptosis, globe dystopia, ophthalmoplegia, retinal arterial occlusion, and optic neuropathy herald orbital and possible intracranial involvement. Fever and tachycardia were present in the majority of patients in two large series of PONF. In the Rajak et al series, 21% of patients presented with evidence of systemic shock.[1] In another series of 11 patients with PONF described by Tambe et al, 1 patient was hypotensive at presentation and 2 experienced a fluctuating level of consciousness.[2]

Investigations

The patient should be admitted to a closely monitored inpatient setting such as intensive care. Vital signs must be repeated frequently to detect fever, tachycardia, tachypnea, or hypotension, which could indicate sepsis. Difficulties breathing resulting from laryngeal edema may necessitate urgent airway management. Laboratory investigations frequently demonstrate marked elevations in white blood cell count and serum CRP. Derangements in electrolyte levels, metabolic acidosis, impaired renal function, elevated creatine kinase, and liver function tests indicate organ dysfunction from sepsis. Neuroimaging with CT of the brain and orbits should be performed to assess for orbital or intracranial extension when suspected clinically. A wound swab of purulent material will enable identification of the causative organism; blood cultures should also be performed. Marking the region of affected skin at presentation can facilitate monitoring the clinical extent.

LRINEC score

Differentiating between severe cellulitis and necrotizing fasciitis may pose a clinical challenge. Evidence of sepsis such as tachycardia, hypotension, and elevated CRP (>15 mg/dL) and/or creatine kinase levels are suggestive of necrotizing fasciitis. The LRINEC is an effective 13-point clinical tool designed to differentiate necrotizing fasciitis from advanced soft-tissue cellulitis.[4] Points are ascribed for white blood cell count level, CRP, hemoglobin, sodium, creatinine, and glucose levels. An LRINEC score >5.8 is suggestive of necrotizing fasciitis (positive predictive value 57%–92%); however, lower scores do not exclude the diagnosis.[3,4] LRINEC scores must be interpreted with caution as this clinical tool may underdiagnose milder presentations of necrotizing fasciitis, periocular necrotizing fasciitis, or cases arising in the pediatric population.[2,3]

Management

A multidisciplinary approach involving infectious disease, critical care, ophthalmology, oculoplastic surgery, otolaryngology, dermatology, and plastic surgery is essential to optimize the management of patients with PONF. Appropriate medical consultation should be sought to correct metabolic derangements, initiate fluid resuscitation, vasopressor support, and further stabilization in intensive care.

Empiric intravenous antibiotic therapy should be determined in consultation with infectious disease specialists with coverage of typical organisms associated with the subtype of necrotizing cellulitis and consideration of local resistance profiles until treatment can be tailored to the results of culture and sensitivity testing. The Infectious Diseases Society of America (IDSA) guidelines recommend that polymicrobial necrotizing fasciitis be treated with vancomycin or linezolid plus piperacillin-tazobactam, a carbapenem, or ceftriaxone-metronidazole.[5] The IDSA also recommend treating GAS-associated necrotizing fasciitis with clindamycin in combination with a penicillin for 10–14 days. Intravenous clindamycin inhibits the synthesis of GAS M protein and pyogenic exotoxins. Hyperbaric oxygen may be beneficial but should not delay surgical

management.[3] The role of intravenous immunoglobulin as adjunctive therapy to neutralize extracellular toxins is controversial and is not currently recommended by the IDSA for GAS-associated necrotizing fasciitis.[3,5]

Surgical exploration and debridement

A critical aspect of the management of PONF is the early consideration of surgical exploration, debridement, and procurement of specimens for Gram stain and culture.[3] Universally accepted surgical principles in managing necrotizing fasciitis indicate that all necrotic tissue should be removed until healthy, bleeding tissue is encountered. Debridement decreases the bacterial load and subsequent production of collagenase, hyaluronidase, and M proteins, thereby limiting necrosis of adjacent tissue and minimizing systemic involvement. Survival is significantly increased among patients taken to surgery within 24 hours after admission, and early surgery may be associated with further increases in survival.[3] In cases of convincing PONF with evidence of sepsis, urgent debridement must be considered and repeated every 1-2 days until necrotic tissue is no longer present. Patients with orbital involvement may require subtotal or total orbital exenteration to obtain control of the infectious process.[6] In the series of patients with PONF published by Rajak et al, 79% of patients underwent between 1 and 5 sessions of surgical debridement 1–14 days after presentation.[1]

Some subtle differences may exist in the surgical management of PONF and necrotizing fasciitis occurring elsewhere. There is controversy about the necessity for urgent surgical debridement in all cases of PONF. Mutamba described 3 patients with PONF in whom intravenous antibiotics successfully controlled the infection.[7] Intravenous antibiotics alone were required in 17% of patients in the Rajak et al series before clinical parameters indicated improvement.[1] Superior outcomes in PONF with antibiotics alone may result from its earlier presentation, the exquisite perfusion of periocular tissues and enhanced antibiotic penetration, and the presence of the orbital septum, which acts as a natural barrier to the spread of infection.[1] Once control of the infection is obtained, performing delayed debridement may preserve periocular tissue, which is advantageous for subsequent ocular function, protection, and cosmesis. While controversial, clinicians may consider intravenous antibiotics, close inpatient monitoring, and delayed debridement for very select patients who are systemically well, present early, and have localized PONF.

Reconstruction of the periocular tissues should be delayed until after the infectious process has been controlled and the remaining tissue has had an opportunity to heal. When loss of periocular tissue has taken place, the exposed ocular surface should be managed with frequent lubricating ointment, moisture chamber, and tarsorrhaphy as needed.

Conclusion

PONF is a life-threatening condition that may present first to the ophthalmologist. PONF should be considered when a patient presents with severe pain and rapidly progressive periorbital cellulitis. A history of skin trauma or immunocompromise should heighten suspicion for PONF. Rapidly evolving cutaneous changes such as erythema, pallor, bullae, and subsequent necrosis are pathognomonic. Early diagnosis, close inpatient monitoring, fluid resuscitation, intravenous antibiotics, and consideration of early surgical debridement are essential aspects of its management and are best facilitated by multidisciplinary medical and surgical collaboration.

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