Periocular Necrotizing Fasciitis Causing Posterior Orbitopathy and Vision Loss: How to Manage?

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Abstract
Necrotizing fasciitis (NF) is a rare, rapidly progressive bacterial infection. Periorbital NF may spread from the eyelid into the posterior orbit. Extent of the infection is critical in planning surgical debridement. A diabetic 70-year-old man presented with a black wound and severe pain in the left periorbital area following a mild trauma. Clinical findings were consistent with NF involving the eyelids, temporal and malar regions. In addition, he had proptosis, diffuse ophthalmoplegia, and central retinal artery occlusion, suggesting deep orbital involvement. Computed tomography showed soft tissue abnormalities in the anterior orbit. The patient was successfully treated with subcutaneous debridement, antibiotherapy, and metabolic support. Periorbital NF may be complicated with posterior orbital cellulitis-like symptoms and retinal vascular occlusions, possibly because of remote vascular thrombi induced by bacterial toxins. This clinical manifestation should be distinguished from true bacterial invasion of the posterior orbit, which may require more aggressive surgical treatments such as exenteration.

Keywords: Necrotizing fasciitis, central retinal artery occlusion, cellulitis, posterior orbitopathy, vision loss, treatment

Introduction
Necrotizing fasciitis (NF) is a severe infection characterized by necrosis of the subcutaneous tissues spreading through the fascial planes. Although it rarely occurs in the periorbital region, the eyelid infection can rapidly spread into the posterior orbit and cervicofacial area and may result in blindness and death if untreated. The mainstay of treatment is early and complete surgical debridement of the infected tissues. Bacterial invasion of the posterior orbit, orbital cellulitis, can cause signs such as proptosis, ophthalmoplegia, and vision loss and requires treatment by orbital exenteration. However, in some cases, these symptoms may not be the result of true cellulitis. Such a case is presented herein, and this paradoxical condition, which may be critical for surgical planning, is discussed.

Case Report
A 70-year-old man presented with a black wound involving the eyelids and severe periorbital pain on his left side. The patient had fallen and hit the left side of his face on the ground 4 days previously. He had a 5-year history of diabetes mellitus. The patient was afebrile, fatigued, and in distress from the periorbital pain. Black, necrotic shells were noted on the left upper and lower eyelids. Two other oval, seminecrotic lesions were seen in the temporal and malar regions. The perilesional
Skin was erythematous, firm, and tender to palpation (Figure 1A). Lifting the upper eyelid with a Desmarres retractor revealed proptosis, restriction of eye movements in all directions, diffuse chemosis, and corneal edema. Visual acuity was 20/50 in the right eye and light perception in the left eye. A relative afferent papillary defect (RAPD) was present in the left eye, and central retinal artery occlusion (CRAO) was detected on slit-lamp fundoscopic examination. Mild nonproliferative diabetic retinopathy was noted in the other eye.

Laboratory studies were significant for leukocytosis (19,800/mL), neutrophilia (85%), hyperglycemia (580 mg/dL), and high C-reactive protein (92 mg/L). Urine analysis was positive for glucose and negative for ketone bodies. Computed tomography (CT) showed diffuse soft tissue thickening, fascial plane blurring, and gas collection in the anterior periorbital regions (Figure 1B). Wound swabs were taken and empirical treatment with ampicillin-sulbactam (6 g/day, intravenous) and ciprofloxacin (1200 mg/day, intravenous) was initiated. The swab culture grew *Staphylococcus aureus*, *Streptococcus parasanguinis*, and *Enterobacter cloacae*.

At surgery, the necrotic shells were elevated with a sub-brow incision in the upper eyelid and a subciliary incision in the lower eyelid. All foul-smelling, necrotic tissues were excised up to the viable edges (Figure 1C). The operative field was copiously irrigated with povidone-iodine and 3% hydrogen peroxide solutions. The eyelid margins and ischemic (purplish) skin areas were not removed. After surgery, the patient experienced arterial hypotensive episodes that responded to fluid resuscitation in the intensive care unit. He was transferred to the ophthalmology ward 3 days later and received hyperbaric oxygen therapy (2.5 atmospheres absolute, 2 hours) in the following 10 days. Histologic findings were consistent with NF (Figure 1D).

Postoperatively, periorbital swelling and extraocular muscle motility improved gradually. Corneal scarring secondary to cicatricial lagophthalmos developed, and visual acuity remained at the level of hand motion. In the late period, upper and lower eyelid reconstructions were performed with skin grafts to correct cicatricial eyelid retractions (Figure 1E). During a follow-up of 11 months, no other complication occurred.

**Discussion**

Suspected necrotizing infections are emergent conditions, and a delay in diagnosis and treatment is associated with worse results. Most patients are taken to surgery based on clinical suspicion, and intraoperative findings play a critical role in making the diagnosis and determining the extent of surgical debridement.

Orbital exenteration surgery is indicated in some periorbital NF cases to overcome the infection, just as severe extremity NF requires amputation. In a review of 94 patients with periorbital NFs, rates of blindness, exenteration, and mortality were 13.8%, 7.4%, and 8.5%, respectively. In two recent, large series, orbital exenteration rates were 2.5% (n=1/40) and 17.6% (n=5/17). One of the prognostic factors for mortality is blindness, and the others are toxic shock, polymicrobial infection, and facial involvement. In two studies, emergent orbital exenteration was performed to control the infection in 7 (58.3%) of a total of 12 patients with NF who had posterior orbitopathy signs (such as vision loss, proptosis, and ophthalmoplegia). The etiology of vision loss was specified as central retinal or ophthalmic artery occlusion because of the spread of infection to the retrobulbar orbit in 5 patients. Recently, there have been reports of other patients with periorbital NF who underwent exenteration without any signs of orbitopathy other than vision loss.

In contrast, other periorbital NF cases were also described in the literature, which were successfully treated with only local/subcutaneous debridement, despite vision loss and other posterior orbitopathy signs. Two well-described patients initially presented with CRAO and developed signs of orbital cellulitis within 12 to 24 hours. In these cases, surgical debridement limited to subcutaneous and preaponeurotic fat tissues was sufficient for complete resolution of the NF. In a study of 40 eyes, orbital involvement presented as motility problems in 12 eyes, proptosis in 8 eyes, and RAPD in 9 eyes. Although orbital involvement resulted in poor visual prognosis in 5 eyes, only 1 eye required exenteration surgery.

CT and magnetic resonance studies can help diagnose NF as well as indicate the extent of infection. Typical CT findings include thickening, fluid collections and, more specifically, gas bubbles in the soft tissues. Of the 9 patients who

**Figure 1.** (A) Preoperative view of the patient with periorbital necrotizing fasciitis. (B) Computed tomographic scan shows gas collection in the preseptal area and soft tissue thickening in the periorbital and temporal regions. (C) Typical intraoperative appearance of the necrotic tissues. (D) Diffuse necrotic inflammation involving the subcutaneous muscle and fibroadipose tissue layers, and heavy, diffuse infiltration of neutrophils and macrophages into the subcutaneous tissues (hematoxylin-eosin, x40 and x200). (E) The patient’s appearance after eyelid reconstruction.
underwent exenteration due to periorbital NF in four different reports, only 2 had radiological images.\textsuperscript{3,6,7,8} In these cases, CT images demonstrated soft tissue thickening in the anterior orbit and were no different from those in the present case and other non-exenterated periorbital NF cases.\textsuperscript{9,10,11,12} No retrobulbar involvement was present in any CT images in the literature.\textsuperscript{3,5,8,9,10,11,12} One recent report of an exenterated case does not include a radiologic image, but it states that orbital CT demonstrated preseptal cellulitis, without intra- or retro-orbital involvement.\textsuperscript{7}

How can clinical signs indicating posterior orbitopathy (such as vision loss, proptosis, diffuse ophthalmoplegia, CRAO, and RAPD) be explained when only the anterior orbit is infected? It can be assumed that infection foci that are too small to be radiologically visible can reach the posterior orbit. If this were the case, subcutaneous debridement limited to the anterior orbit would likely be inadequate to control fasciitis, and exenteration would be required for definitive treatment in almost all cases with posterior orbitopathy. When treatment is only based on antimicrobial therapy and support, mortality approaches 100%.\textsuperscript{1} Compartment syndrome associated with orbital edema or superinfection with vascular infiltration have been suggested to explain retinal arterial occlusions.\textsuperscript{9,11}

Experimental NF models show that streptococcal toxin injection can trigger an immune-mediated platelet activation and thrombus formation.\textsuperscript{14} Microthrombi develop at both the local site of infection and distant areas. Soft tissue blood flow decreases regionally within minutes, depending on the toxin dose. Toxin-induced arterial occlusion mediates the rapid ischemic destruction of tissue, continued expansion of the bacterial niche, and thwarting of the host immune response.\textsuperscript{15} Vision loss, ophthalmoplegia, and CRAO may not be caused directly by bacterial invasion of the deep orbit, but by the vaso-occlusive effect of bacterial toxins released from the eyelid infection.

During surgery, it is possible to recognize the signs of NF and determine the infection margins macroscopically. Therefore, in the subgroup of patients with periorbital NF and posterior orbitopathy, when there is no retrobulbar involvement on radiologic studies, surgery can be initiated using subcutaneous debridement in the eyelid. If viable tissue is not reached in the aponeurotic fat and fasciitis signs persist into the posterior orbit, the surgery may be converted to an exenteration procedure. Thus, it may be possible to avoid unnecessary surgical morbidity in some cases.

\textbf{Ethics}

\textbf{Informed Consent}: Obtained.

\textbf{Peer-review}: Externally peer reviewed.

\textbf{Authorship Contributions}

Surgical and Medical Practices: B.Y., H.S., FT, Concept: B.Y., H.S., FT, Design: B.Y., H.S., Data Collection or Processing: B.Y., H.S., FT, Analysis or Interpretation: B.Y., H.S., FT, Literature Search: B.Y., H.S., Writing: B.Y., H.S., FT.

\textbf{Conflict of Interest}: No conflict of interest was declared by the authors.

\textbf{Financial Disclosure}: The authors declared that this study received no financial support.

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