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Pyoderma gangrenosum of the eyelid

Pyoderma gangrenosum (PG) is a rare inflammatory skin condition of unknown etiology. In rare cases, it affects the eyelid, where it can have devastating visual consequences. We describe a case of severe eyelid PG, successfully managed with aggressive chronic immunosuppression.

PG is one of the neutrophilic dermatoses—a group of dermatological diseases characterised by skin infiltrate with mature polymorphonuclear lymphocytes in the absence of an infection or true vasculitis.[1] PG has an incidence of approximately 6 per 1 million person-years in the United Kingdom. The characteristic clinical feature of PG is the formation of sterile pustules that rapidly develop into an ulcer with a purulent base that undermines its elevated, violaceous edge.[2] PG often affects the legs and trunk and is associated with chronic gastrointestinal, endocrine, or hematological conditions. We add a further case to the few reports of eyelid PG and review the literature to investigate clinical features, treatment strategies, and outcomes. We wish to highlight that cases with preservation of the eyelid posterior lamellae appear to have a good prognosis, whereas cases with lagophthalmos or orbital involvement have a poorer prognosis. In addition, we highlight that treatment with infliximab infusions and high-dose oral prednisolone has been highly successful in our case.

A 40-year-old female with poorly controlled type 2 diabetes mellitus and a 10-year history of PG presented with a painful, swollen, inflamed left upper lid, which was diagnosed as being a chalazion. The PG was usually controlled with monthly infliximab, but she had missed the infusion 3 weeks before presentation. She presented again 2 weeks later describing the lesion to have "burst" 1 day before. On examination she had a horizontal fissure through skin and orbicularis oculi muscle, at the skin crease of the left upper lid, with surrounding erythema (Fig. 1A). Additionally, she had a single 1-cm-diameter abdominal follicular skin lesion.

She was treated with oral antibiotics, but over the subsequent 2 weeks the disease process completely destroyed the left upper lid anterior lamella (Fig. 1B). The levator function, extraocular movements, and cornea were unaffected. The result of blood testing was unremarkable except for positive anti-neutrophil cytoplasmic antibody immunofluorescence with a 2+ perinuclear pattern. A computed tomography scan showed no postseptal extension of the inflammation, and swabbing of the lesion cultured

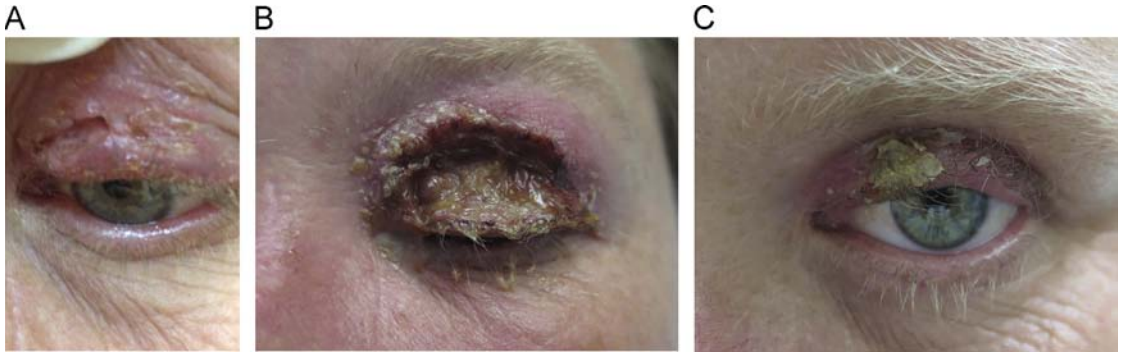

Fig. 1—(A) Left upper lid appearance at second presentation, with a horizontal upper lid fissure in the upper through the full depth of the anterior lamella. (B) Two weeks later, extensive ulceration of the anterior lamella. The ulcer has a violaceous rim and an undermined border. (C) Four weeks later, the lesion granulated and has almost completely healed with high-dose oral prednisolone and infliximab infusion.

Malassezia furfur and coagulase-negative staphylococci, which were presumed to be skin commensals. A biopsy of the edge of the lesion revealed an acute inflammatory cell exudate, abundant laminated keratin, and necrotic stratified squamous epithelium, suggestive of ruptured suppurative folliculitis with abscess formation, which is consistent with PG.[3] The patient was commenced 1 mg/kg (75 mg/day) oral prednisolone tapering by 10 mg/week and prophylactic antimicrobials, and her usual 2-monthly 5 mg/kg infliximab infusion was reinstated. The lesion granulated and scabbed, and the anterior lamella reformed steadily over the subsequent 4 weeks (Fig. 1C). She was left with 2 mm of lagophathlmos on gentle closure. The inflammation worsened again 2 months later when the dose of prednisolone dropped to 35 mg/day, necessitating a higher dose and even slower taper. The cornea and visual acuity remained unaffected throughout.

Discussion

Eyelid PG is extremely rare. Only 15 other cases have been reported, and they have a wide range of clinical presentations, treatments, and outcomes.[4–14] Our case is instructive for its dramatic response to oral steroids and for highlighting clinical features that may be predictive of a good outcome.

Eyelid PG affects a similar demographic to PG elsewhere in the body, with 9/15 (60%) eyelid cases occurring in women and the average age being 55 years (range 4–80 years).[15] The disease is more commonly unilateral (11/15 cases: 8 right, 2 left, and 1 unstated) and in the upper lid (7/15 upper, 3/15 lower, 4/15 upper and lower, and 1/15 inadequate description). In common with our case, 4/15 cases involved only the eyelid, which may reflect prompt and aggressive treatment or disease variation.[5,6,11,14] In the other 11 cases, the eyelid was part of a wider area of disease, with involvement of the scalp, cheek, temple, lateral canthus, inferior oblique, inferior rectus and lateral rectus muscles, orbital apex, sclera, anterior chamber, and maxillary sinus or nasopharynx.[4,7–13]

The initial diagnosis in our case delayed immunosuppressive treatment by 4 weeks, during which time there was complete breakdown of the anterior lamella. However, the eyelid was recovered with aggressive immunosuppression, and the corneal health and vision were preserved. Initial misdiagnosis is common in eyelid PG. Diagnostic delays of 7 days to 3 years occurred in three-quarters (9/12 in which diagnosis time is reported) of previous cases, even when the diagnosis of PG was already known, with misdiagnoses of hordoleum, stye, abscess, chalazion, herpetic infection, preseptal cellulitis, tumour, and nodular scleritis.[4–8,10–12,14] Useful clues to the diagnosis include high levels of pain inconsistent with cystic swellings and the presence of other systemic diseases; leukopaenia, aplastic anemia, ulcerative colitis, diabetes, and rheumatoid arthritis have all been reported in patients with eyelid PG.[4,7,8,11]

Oral or intravenous steroids are the mainstay of treatment for PG and, in combination with recommencement of infliximab infusions, brought about a dramatic and almost complete recovery in our patient. The precise treatment regimen will need to be case specific, but in general, aggressive early treatment is necessary and may need to be followed by chronic immunosuppression (6/15 previous cases).[4,7,10–13] Other immunosuppressants that have been used with variable success in eyelid cases include cyclosporin A, azathioprine, chlorambucil, adalimumab, cyclophosphamide, mycophenalate mofetil, sulphasalazine, and dapsone. Surgery should be avoided if possible in PG as pathergy can occur both at the disease site and at the donor site if skin grafting is used (1/8 cases).[12] If emergency corneal protection is required, we would advocate simple closure or coverage procedures with few incisions and sutures. Definitive reconstructive surgery can be considered after several months of remission, although it is not commonly needed.

Despite the dramatic appearance of our case, the preservation of the posterior lamella at the time of commencing immunosuppression was probably responsible for the good outcome. Permanent visual loss occurred in 5/15 previous cases.[7,10–13] In 4 of these 5 cases, lagophthalmos resulted in corneal perforation, necessitating eye removal in 3 cases and penetrating keratoplasty in the other.[7,11,12] In the other case, there was dense central corneal scarring without perforation.[13] Orbital inflammation followed by fibrosis resulted in impaired ocular motility in 1 case.[10] Orbital involvement of PG is a strong predictor of permanent ophthalmic complications, being present in 4/6 cases with visual or eye movement sequelae.[7,10–12] Anterior scleral inflammation (1 case) and chronic lagophthalmos were present in the other 2 cases.[11,13]

Eyelid PG is extremely rare, and easily misdiagnosed as infection or chalazion. Prompt diagnosis and institution of aggressive immunosuppression can facilitate a dramatic and complete recovery.

Disclosure: The authors declare that there are no financial, proprietary, or other conflict of interest.

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Orbital cellulitis and multiple abscess formation after strabismus surgery

A 3-year-old female underwent bilateral lateral rectus recessions of 7.0 mm for an intermittent exotropia of 35 prism diopters (PD). Her ocular history was significant for left amblyopia. She was known for a history of bilateral mastoiditis and recurrent episodes of acute otitis media. There was no history of upper respiratory tract infection before surgery.

Preoperatively, the patient's face was prepped with 10% povidone solution, and 5% povidone drops were instilled in both eyes. The lateral recti were approached through limbal conjunctival incisions, which were closed postoperatively using 8-0 polyglactin sutures. Dexamethasone and tobramycin ointments were instilled in both eyes at the conclusion of surgery. The patient was using tobramycin drops three times daily in both eyes postoperatively.

The patient presented on the third day postoperatively with left periorbital swelling, redness, and pain. She was afebrile. Best-corrected visual acuity was 20/20 in the right eye and 20/30 in the left eye, consistent with the patient's baseline status. There was a moderate left abduction deficit. Slit lamp examination revealed an abscess collection in the left temporal quadrant of the conjunctiva. There was no relative afferent pupillary defect. Dilated fundus examination was unremarkable. Magnetic resonance imaging of the orbits (Fig. 1) revealed a left lateral extraconal abscess collection, as well as inflammatory changes involving the left lateral rectus and the lacrimal gland, with enlargement of the latter. There was a small T2 hyperintense signal adjacent to the medial aspect of the left lateral bony orbit corresponding to a subperiosteal abscess, with no associated sinusitis. The infectious diseases service was consulted, and the patient was admitted to the medicine ward for intravenous pipercillin–tazobactam and vancomycin. There was no significant improvement, and the patient was taken 5 days after admission for drainage of the subconjunctival collection (Fig. 2). Cultures came back positive for methicillin-sensitive *Staphylococcus aureus*. Serial examinations revealed improvement in extraocular motility. One-week follow-up revealed a consecutive esotropia of 8 PD in the primary position at distance and near. The patient completed a 3-week course of intravenous ceftriaxone, topical erythromycin and ofloxacin, and did well on follow-up. There was slow resolution of the periorbital edema and return of full extraocular motility. At 6 months, she had a small consecutive esotropia of 6 PD.

Given the patient's history of mastoiditis and abscess formation, the possibility of innate deficiencies of the immune system was entertained. However, complement function, di-hydroamine 123 (for chronic granulomatous disease) and leukocyte surface glycoprotein (LFA-1) testing (for leukocyte adhesion deficiency) were normal.

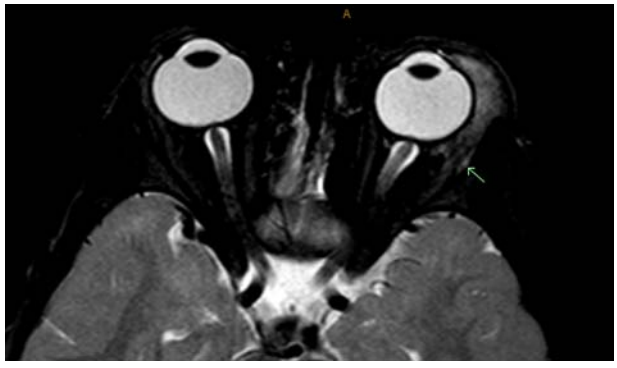

Fig. 1—T2 gadolinium-enhanced axial slice through the orbits demonstrating a small left subperiosteal abscess collection (arrow).