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Acute anterior necrotizing scleritis: A case report

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ABSTRACT

Necrotizing scleritis is an uncommon but potential disastrous infection to the eye. It is commonly caused by vaso-occlusive autoimmune diseases such as rheumatoid arthritis or surgically-induced, and rarely due to infections. In this article, we presented a rare case of necrotizing scleritis caused by herpes infection in an immunocompromised patient. A 49 years old, retroviral positive gentleman presented to our clinic with a painful, red right eye associated with watering, photophobia and blurring of vision. His right eye rapidly deteriorated leading to an impending perforation of the sclera despite intensive antimicrobial therapy. The patient was started on acyclovir ointment and subsequently improved remarkably salvaging the eye from the need of an evisceration. Although the visual prognosis was poor, structural integrity of the eye was achieved.

1. Introduction

Necrotizing scleritis is presumably the most severe and destructive form of scleritis. The pathophysiological mechanism is regarded as an immune complex reaction (type III hypersensitivity reaction)^[1]. Most of the time, scleritis is not caused by an infectious agent^[2] but rather trauma or surgically-induced. Here we discussed a case of necrotizing scleritis, seen in a HIV positive patient, which proved to be a challenge in management.

2. Case report

A 49 years old, HIV positive gentleman, on highly active antiretroviral therapy, presented to our clinic with a painful, red right eye. It was associated with watering, photophobia and blurring of vision for the past 3 days. The condition was in sudden onset and rapidly progressive. There were no similar episodes previously and history of fever, recent trauma or

surgery to the eye. No features of systemic autoimmune diseases such as joint pain, orogenital/skin ulcerations, rashes or photosensitivity to sunlight were presented. A systemic review of systems was grossly unremarkable. His visit to our clinic revealed a visual acuity of 6/36 in the right eye and 6/7.5 in the left eye. Intraocular pressures were 16 mmHg in both eyes. On slit lamp examination of the affected eye, there was conjunctival injection and corneal ulceration extending to the sclera on the nasal side (Figure 1). The anterior chamber showed 3+ cells with a hypopyon. Pupils were round and reactive. A fundus examination did not reveal any pathology. Examination of the fellow eye was unremarkable.

A baseline investigation of full blood count and screening for infections was normal. Autoimmune screening tests, tuberculosis workup and toxoplasma serology came back negative. Corneal scrapping from the ulcer base grew *Pseudomonas aeruginosa*. Thus, the patient was started on a regime of topical fortified gentamicin 0.9% eyedrops hourly, moxifloxacin eyedrops on an hourly basis, homatropine 2% eyedrops three times daily, oral doxycycline 100 mg/day and oral vitamin C 1 g/day. However despite this treatment, the ulceration rapidly progressed and the patient was soon found to develop a necrotizing scleritis adjacent to the corneal ulcer (Figure 2). To our dismay, the nasal sclera thinned out with underlying uveal tissue starting to bulge anteriorly with impending perforation (Figures 3 and 4). Upon further examination we noticed a

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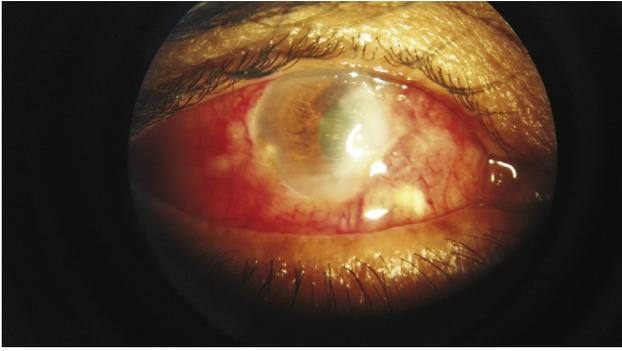


Figure 1. Corneal ulceration beginning to develop on the nasal side.

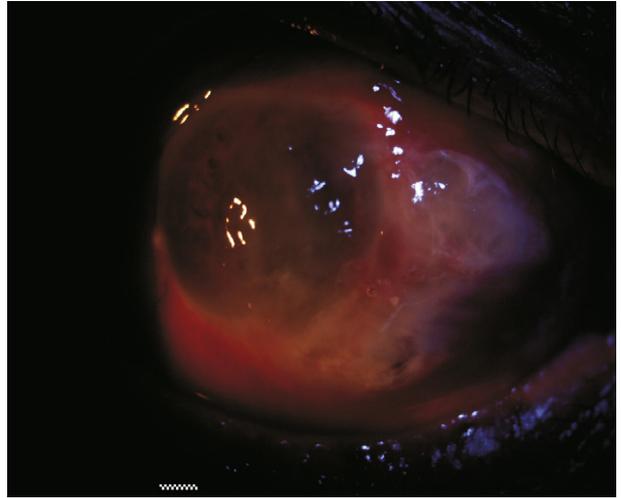


Figure 4. Impending perforation.

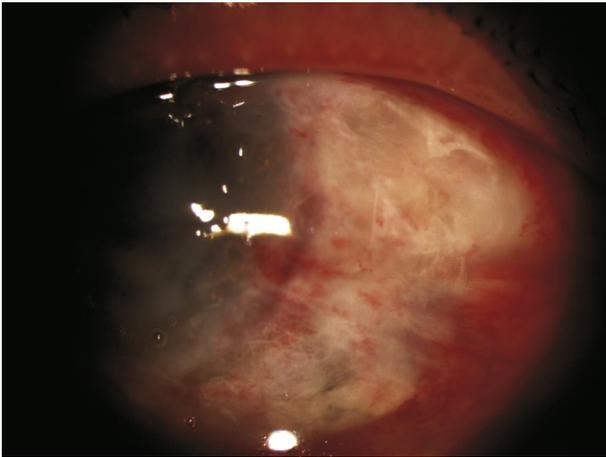


Figure 2. Necrotizing scleritis adjacent to the corneal ulcer.

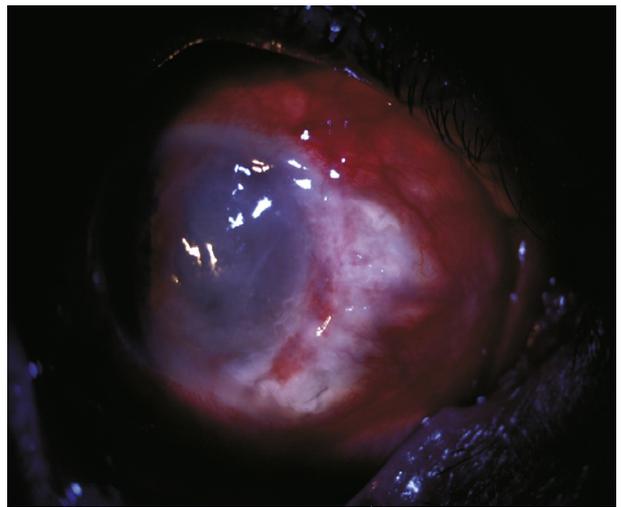


Figure 5. Rapid clinical improvement after starting ointment acyclovir treatment.

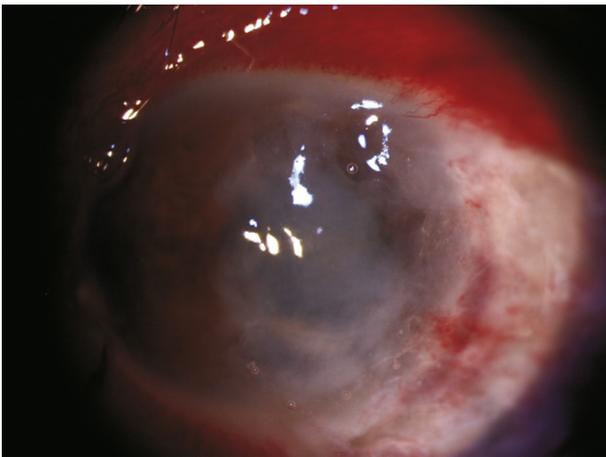


Figure 3. Progressive corneal and scleral thinning nasal side.

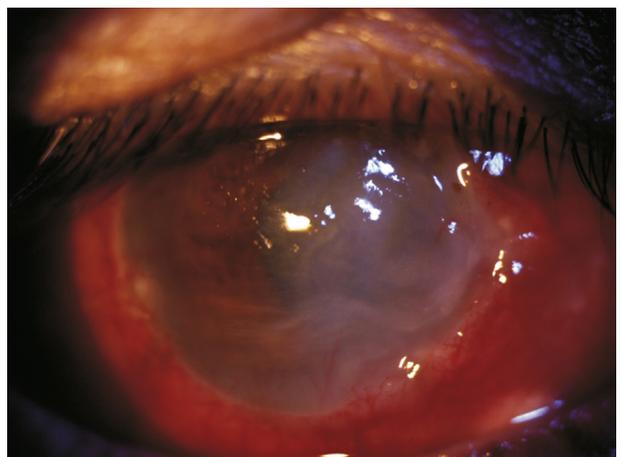


Figure 6. Post treatment resolution with residual scarring.

reduction in corneal sensation. Thus, we added ointment acyclovir to our previous treatment regimen. Soon after, a rapid clinical improvement was noted (Figure 5). Areas of scleral thinning and corneal ulceration started to resolve with fibrosis and conjunctivalization of the thinned out sclera. Subsequently, his clinical course was otherwise uncomplicated and a steady resolution of the ulcer also occurred (Figure 6). His final visual outcome was 6/60 with preservation of the ocular structural integrity.

3. Discussion

Scleritis is an inflammatory disease of the episcleral and the deeper scleral tissues. The pain of episcleritis is less severe than in scleritis^[3]. It may be classified anatomically into anterior- and

posterior-scleritis, the former being more common. Anterior scleritis can be further subdivided into nodular, diffuse and necrotizing. This condition typically presents with intense pain, thinned out white avascular areas surrounded by inflammation of the sclera and conjunctiva. Fundus examination may reveal choroidal folds, subretinal mass, disc edema, macular edema, annular ciliochoroidal detachment and serous retinal detachment^[1]. Autoimmune causes are far more common, of which rheumatoid arthritis is by far the most common systemic condition associated with scleritis. The other associated diseases are Wegener's granulomatosis, systemic lupus erythematosus, juvenile rheumatoid arthritis, polyarthritis nodosa, relapsing polychondritis, psoriasis, gout, atopy, and rosacea. The common infective causes reported are tuberculosis, syphilis, herpes zoster and herpes simplex virus^[1]. In our case, autoimmune screenings and infective screens came back negative except for the retroviral disease. Herpes infection was clinically suspected, however we were not able to isolate the virus.

Herpes zoster, which is a member of herpes virus family, is the causative agent of chickenpox and zona zoster diseases^[4]. Zona zoster is the reactivation of the latent virus long after the primary infection has resolved^[4]. This condition is characterized by eyelid necrosis, iritis, necrotizing keratitis, neurotrophic keratitis with corneal melting, extraocular muscle paralysis, internal ophthalmoplegia, cataract, chorioretinitis and optic neuritis^[5]. The pathophysiology of herpes zoster ophthalmicus remains obscure^[6]. Scleral weakening, seen in necrotizing scleritis was commonly attributed to immune complex-mediated vasculitis. However, there are reports of direct scleral invasion by herpes zoster virus^[6,7]. Our case was likely a reactivation of the virus due to immunocompromised status of the individual, a fact which the patient did not disclose to us. The condition was only found out after the positive HIV results, following which he admitted to his status and highly active antiretroviral therapy treatment. A delay in diagnosis could possibly have led to perforation and disastrous consequences.

A trial of acyclovir has been supported in cases where biopsy is not possible and suspicion of herpetic infection is strong (*i.e.*, unilateral disease and not responsive to steroids), 800 mg 3 times daily orally or if there are unequivocal signs of herpes zoster infection (*e.g.*, herpes zoster ophthalmicus). It is acceptable to use acyclovir 800 mg 5 times a day or famciclovir instead^[8]. The

response to acyclovir would depend on whether there is an autoimmune component^[8]. Amongst the findings that aroused suspicion in our case were its acute onset, failure to improve despite existing antimicrobial treatment, unilateral presentation and associated keratouveitis. Prompt administration of acyclovir based on clinical suspicion was proved vital in the prevention of scleral perforation. Steroids were not used in this case as they may delay epithelial healing^[7,8].

In conclusion, necrotizing scleritis is a disastrous and difficult condition to manage, especially in immunodeficient subjects. In cases where no response is seen to antimicrobial agents, a trial of anti-viral agents may be proved beneficial.

Conflict of interest statement

The authors report no conflict of interest.

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