



Brief Report

Orbital implant exposure after Acanthamoeba panophthalmitis

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ABSTRACT

Purpose: Acanthamoeba is a protozoa that can lead to severe ocular disease and sequelae. Although intraocular Acanthamoeba infection is rare, the following case demonstrates an unusual presentation of recurrent Acanthamoeba infection in a 30 year old contact lens wearing male.

Observations: After presenting with recurrent Acanthamoeba keratitis and undergoing various treatments, the patient developed nodular scleritis, which evolved into panophthalmitis, and ultimately, required enucleation. Eight months post-operatively, the patient developed orbital implant exposure secondary to persistent Acanthamoeba infection and underwent removal of the implant and aggressive, systemic treatment involving a multispecialty care team. He then underwent placement of a dermis fat graft and had no signs of persistent infection at the time of last follow-up, which was 24 months after placement of the dermis fat graft.

Conclusions: and Importance: To the authors' knowledge, this is the first known case of Acanthamoeba infection causing orbital implant exposure. Persistent infection should be considered in Acanthamoeba patients who have undergone enucleation and have orbital implant exposure. Better knowledge regarding the pathogenesis and extracorneal complications of this challenging disease may improve patient care and outcomes.

1. Introduction

Acanthamoeba is a ubiquitous free-living protozoa that can lead to destructive ocular sequelae and significant ocular pain. Initially described in association with ocular trauma, the use of contact lenses has increasingly been associated with Acanthamoeba infection. The authors report a case of recurrent Acanthamoeba keratitis, which evolved into nodular scleritis and retinitis requiring enucleation, and ultimately caused orbital implant exposure. To the authors' knowledge, this is the first reported case of orbital implant exposure secondary to Acanthamoeba infection.

2. Case report

2.1. Collection and evaluation of protected patient health information was HIPAA-compliant

A healthy, 30-year-old, male, soft contact lens wearer was referred to the University of Iowa for evaluation of a persistent epithelial defect of the right eye (OD). He had been unsuccessfully treated with topical tobramycin-dexamethasone and trifluridine. On initial presentation,

visual acuity (VA) was 20/300 OD. He had 360° of ciliary flush and a 5 × 6 mm central epithelial defect (Fig. 1A). His exam was otherwise unremarkable. He was diagnosed with herpes simplex keratitis (by direct fluorescent antibody testing) and Acanthamoeba keratitis (by confocal microscopy) and started on hourly topical chlorhexidine, topical moxifloxacin, and oral acyclovir. Corneal cultures for aerobic, anaerobic, and acid-fast bacteria were negative.

Two weeks later, he developed a ring infiltrate and hypopyon. He was treated with intensive topical steroid and continued on topical anti-amoebic therapy. After one month, the epithelial defect persisted, and pain increased despite mild improvement in vision (20/125) and inflammation. Hourly propamidine-isethionate was added, and therapeutic penetrating keratoplasty (PKP) was performed. Histopathology confirmed the presence of amoebic cysts in the host (Fig. 1B). Despite continuation of topical anti-amoebic agents, overwhelming amoebic infection was noted in the graft. After six months, despite maximum anti-amoebic treatment, the graft infection persisted, and keratouveitis worsened. Intravenous pentamidine 200 mg daily and oral prednisone 60 mg daily were initiated (under the guidance of the infectious disease team) for two weeks before a second PKP was performed. A recurrent ring infiltrate was noted. Hourly topical voriconazole and oral

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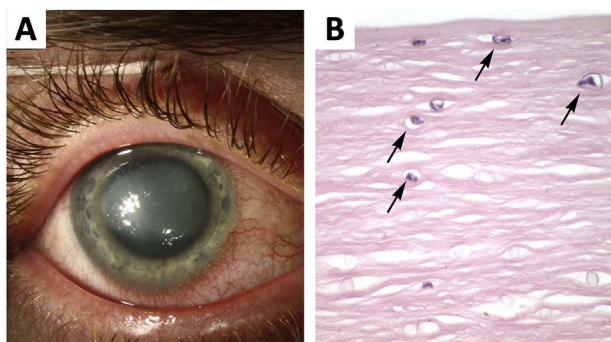


Fig. 1. 1A: At presentation, exam showed 360° of ciliary flush and a 5 × 6 mm central epithelial defect. 1B: Histopathology confirmed the presence of amoebic cysts in the host cornea. Arrows indicate cysts. (Hematoxylin and eosin stain, 300x).

fluconazole 200 mg daily were added; a third PKP was performed. Recurrent amoebic cysts were noted in the third graft. Additionally, a cataract formed, vision deteriorated to hand motions, and three scleral nodules were noted superiorly. Fifty milligrams of daily intravenous caspofungin was started, and the patient underwent planned sclerokeratoplasty, extracapsular cataract extraction, and placement of an intraocular lens. The decision was made to place a lens since the infection was presumably confined to the cornea, the patient had been pre-treated systemically, and the scleritis was thought to be a sterile immune response. Pathology confirmed this; there were no organisms in the corneoscleral button, yet the patient's nodular scleritis persisted, which was consistent with the presumption that the scleritis was a sterile immune response. Due to this presumed sterile yet destructive scleritis, relatively aggressive immunosuppressive therapy with Mycophenolate mofetil (1 g BID) was started under the guidance of the rheumatology team; oral steroids were simultaneously tapered.

One year following presentation, the patient was correctable to 20/25; the graft was clear and compact. After completion of the oral steroid taper, scleral nodules and anterior uveitis recurred. The rheumatology service recommended resuming oral prednisone and adding oral cyclosporine. Ultrasonography of the nodules did not show purulence, so incision and drainage was deferred. A 4 mg dose of sub-conjunctival triamcinolone was injected adjacent to the most prominent nodule. Soon after, the patient noted vision loss (VA 20/70). A dilated exam showed a sector of retinal whitening and hemorrhage (Fig. 2A and B). Given the duration of systemic immunosuppression, recent localized steroid injection, and new retinitis on exam, there was concern for viral retinitis. Immunosuppressive agents were discontinued. A vitreous biopsy was performed with intravitreal injection of vancomycin, foscarnet, and voriconazole. To treat possible viral retinitis, three intravitreal foscarnet injections were given at two-day intervals. Valganciclovir replaced acyclovir. His vision worsened to count fingers at 1 foot, and there were new and expanding regions of retinitis. The vitreous cultures and laboratory studies were negative for a viral etiology.

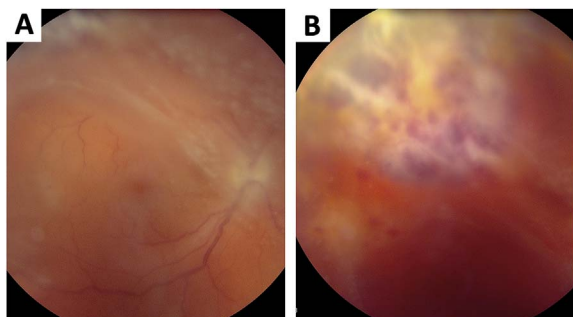


Fig. 2. 2A: Dilated exam showing a sector of retinal whitening and hemorrhage. 2B: Magnified view of the retinal whitening and hemorrhage consistent with retinitis.

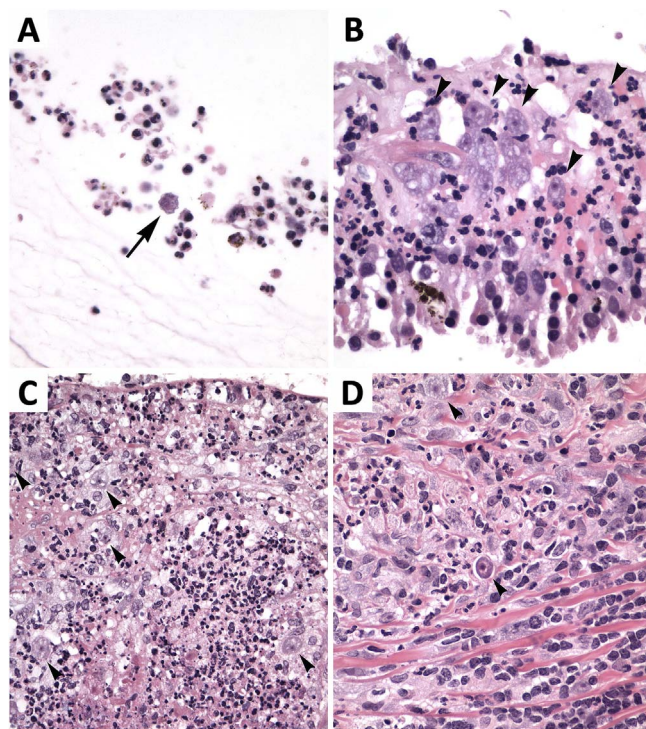


Fig. 3. 3A–D: Ocular pathology demonstrating Acanthamoeba panophthalmitis with organisms in the vitreous (A, 200x), retina (B, 300x), choroid (C, 200x), and sclera (D, 300x). Arrows indicate cysts. Arrowheads trophozoites. (Hematoxylin and eosin stain).

Brain MRI and lumbar puncture were performed to rule out central nervous system (CNS) involvement of Acanthamoeba or other infectious process—these returned within normal limits.

Due to poor visual prognosis and concern for systemic spread, the patient was admitted for intravenous caspofungin, and an enucleation was performed two days later at which time Medpor SST-EZ® orbital implant - was placed. Ocular pathology demonstrated Acanthamoeba panophthalmitis with organisms in the vitreous, retina, subretinal space, choroid, and sclera (Fig. 3A–D). The nerve was free of inflammation, and no acanthamoebae were noted beyond the lamina cribrosa. The patient initially did well post-operatively (Fig. 4A) but developed an exposure of the orbital implant approximately 8 months after enucleation. One month later, he underwent repair of exposure with scleral patch graft and biopsy of the conjunctiva adjacent to the exposure. The pathology showed acute and chronic inflammation with necrosis and the presence of Acanthamoeba organisms. The patient underwent removal of the orbital implant the following week with further biopsies of the patient's conjunctiva, tenon's capsule, and orbit. No organisms were identified on pathology. There was concern that



Fig. 4. 4A: Post operative month 1 status post enucleation prior to fitting for ocular prosthesis. 4B: Final follow-up following dermis fat grafting and placement of ocular prosthesis.

organisms could be present within his orbit due to the previous finding of Acanthamoeba organisms in the conjunctiva, and no implant was placed.

Due to concern for CNS involvement or possible spread, the Infectious Disease Service started the patient on miltefosine under the guidance of the Center for Disease Control. Five months after removal of the orbital implant, the patient underwent placement of a dermis fat graft with tarsorrhaphy and did well post operatively with no signs of persistent infection at the time of last follow-up (Fig. 4B), which was 24 months after placement of the dermis fat graft.

3. Discussion

Acanthamoeba species are ubiquitous free-living protozoa⁷ that exist in both a resilient cyst form and an infective trophozoite form.⁸ Intraocular infection, and particularly retinitis, of Acanthamoeba is rare.^{1–6} This case demonstrates an unusual presentation of recurrent Acanthamoeba infection presenting as nodular scleritis and panophthalmitis, masquerading as viral retinitis, and finally causing orbital implant exposure. Histopathology revealed organisms throughout the eye, including the choroid, which has not been previously reported. The authors are not aware of any case of Acanthamoeba infection causing orbital implant exposure in the literature. Specific treatment of intraocular dissemination of Acanthamoeba is not well established, and despite aggressive measures surgically and with topical, oral, intravenous, and intravitreal anti-amoebic agents, the infection rapidly progressed. This case demonstrates the aggressive nature of Acanthamoeba infection. Early diagnosis, particularly with confocal microscopy, as well as appropriate therapy are keys to a good prognosis,⁹ while late diagnosis, poor initial visual acuity, and large infiltrates are associated with surgical intervention, and subsequently, with worse visual outcome.⁸ In this case, the patient was properly diagnosed with acanthamoebic keratitis at the time of presentation and aggressive anti-acanthamoebic therapy was immediately initiated.

4. Conclusion

A relatively recently recognized infectious entity, Acanthamoeba can be difficult to diagnose and difficult to treat.^{8,9} Appropriate and successful management often requires a coordinated, multi-disciplinary approach, which in this case involved the cornea, retina, pathology, and oculoplastic services as well as the rheumatology and infectious disease services. Persistent infection should be considered in Acanthamoeba patients who have undergone enucleation and have orbital implant exposure. Better knowledge regarding the pathogenesis and extraocular complications of this challenging disease may improve patient care and outcomes.

Patient consent

The patient consented to publication of the case in writing.

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Conflict of interest

The following authors have no financial disclosures: MSB, ACM, NAS, RCA.

Authorship

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

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Appendix A. Supplementary data

Supplementary data related to this article can be found at <http://dx.doi.org/10.1016/j.ajoc.2018.01.043>.

References

- Burke JP, Webber SK, Kerr-Muir MG, Talbot JF, Parsons MA. Acanthamoeba polyphagia panophthalmitis. *Cornea*. 1992 May;11(3):274–275.
- Davis MJ, Packo KH, Epstein RJ, Grostern RJ, Cohen JA. Acanthamoeba endophthalmitis following penetrating keratoplasty for Acanthamoeba keratitis. *Arch Ophthalmol*. 2010 Apr;128(4):505–506.
- Jones BR, McGill JI, Steele AD. Recurrent suppurative kerato-uveitis with loss of eye due to infection by Acanthamoeba castellanii. *Trans Ophthalmol Soc U K*. 1975 Jul;95(2):210–213.
- Jones DB, Visvesvara GS, Robinson NM. Acanthamoeba polyphagakeratitis and Acanthamoeba uveitis associated with fatal meningoencephalitis. *Trans Ophthalmol Soc U K*. 1975;95(2):221–232.
- Moore MB, McCulley JP, Newton C, et al. Acanthamoeba keratitis: a growing problem in soft and hard contact lens wearers. *Ophthalmology*. 1987;94(12):1654–1661.
- Moshari A, McLean IW, Dodds MT, Damiano RE, McEvoy PL. Chorioretinitis after keratitis caused by Acanthamoeba: case report and review of the literature. *Ophthalmology*. 2001;108(12):2232–2236.
- Bouheraoua N, Gaujoux T, Goldschmidt P, Chaumeil C, Laroche L, Borderie VM. Prognostic factors associated with the need for surgical treatments in acanthamoeba keratitis. *Cornea*. 2013;32:130–136.
- Illingworth CD, Cook SD. Acanthamoeba keratitis. *Surv Ophthalmol*. May-Jun 1998;42(6):493–508.
- Dart JKG, Saw VPR, Kilvington S. Acanthamoeba keratitis: diagnosis and treatment update 2009. *Am J Ophthalmol*. 2009;148:487–499.