



Bilateral *Acanthamoeba* Panophthalmitis: A rare and unique case

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ABSTRACT

Purpose: To describe a unique case of bilateral *Acanthamoeba* panophthalmitis in a 65-year-old male resulting in bilateral enucleation.

Observation: A 65-year-old man presented with a 10-year history of bilateral uveitis and scleritis, complicated by cataracts. He had undergone phacoemulsification with posterior chamber intraocular lens implantation in both eyes, left corneal transplant and pars plana vitrectomy, all without improvement in his vision and pain. Due to complete loss of vision and severe pain in his both eyes, the patient underwent bilateral enucleation. Pathologic examination of both eyes revealed severe acute, chronic, and granulomatous inflammation with abundant scar formation. Multiple large pre-retinal, choroidal, and vitreal cavitory lesions in both eyes were filled with necrotic debris, containing both *Acanthamoeba* trophozoites and cysts. These findings were consistent with a well-developed, bilateral *Acanthamoeba* panophthalmitis.

Conclusions and importance: This unique case represents the first ever reported bilateral *Acanthamoeba* panophthalmitis and illustrates the extreme complication of ocular *Acanthamoeba* infection.

1. Introduction

Acanthamoeba species are ubiquitous environmental protozoans that exist in trophozoite and cyst forms and are associated with recurrent keratitis.^{1,2} The incidence of *Acanthamoeba* keratitis is estimated to be 1–2 per 1,000,000 in the US, and approximately 85% cases are associated with poor contact lens hygiene.³ Interestingly, intraocular spread of *Acanthamoeba* is extremely rare, with only seven cases of *Acanthamoeba* endophthalmitis and two cases of *Acanthamoeba* panophthalmitis reported.^{4–11} Here, we report a case of bilateral *Acanthamoeba* panophthalmitis.

1.1. Case report

A 65-year-old man with a 10-year history of uveitis and scleritis, complicated by cataracts, had undergone phacoemulsification with posterior chamber intraocular lens implantation in both eyes, left corneal transplant and pars plana vitrectomy, all without improvement in his vision and pain. Other comorbidities included type 2 diabetes, hypertension, asthma, rheumatoid arthritis, and hepatitis C. Prior to bilateral enucleation surgery, he had completely lost vision and

experienced severe ocular pain in both eyes that was resistant to topical moxifloxacin and steroids and required oral opioid treatment. At the pre-surgical external examination, the corneas of both eyes were intensely vascularized and showed severe purulent acute inflammation involving at least the anterior chambers of both eyes. There were areas of sub-conjunctival hemorrhage and scleral necrosis (Fig. 1). Given the severity of involvement, both globes were enucleated. Eyes were sent for pathologic examination.

On gross examination, the vitreous cavities of both globes were filled with white, opaque purulent material that covered the retina and optic disc. On light microscopic examination, both globes showed severe acute, chronic, and granulomatous inflammation with abundant scar formation. The retina was detached in both globes; and multiple large pre-retinal, pre-choroidal, and vitreal cavitory lesions were filled with necrotic debris containing both *Acanthamoeba* trophozoites and cysts in both globes (Figs. 2 and 3). *Acanthamoeba* cysts and trophozoites were seen both within the corneal stroma and on the sub-conjunctival surface of the sclera. In both globes, gaps in the sclera were identified near the corneoscleral limbus and extending to the ora serrata; the intraocular uveal tissue within this region showed extensive necrosis and inflammation and abutted the large abscesses containing *Acanthamoeba*

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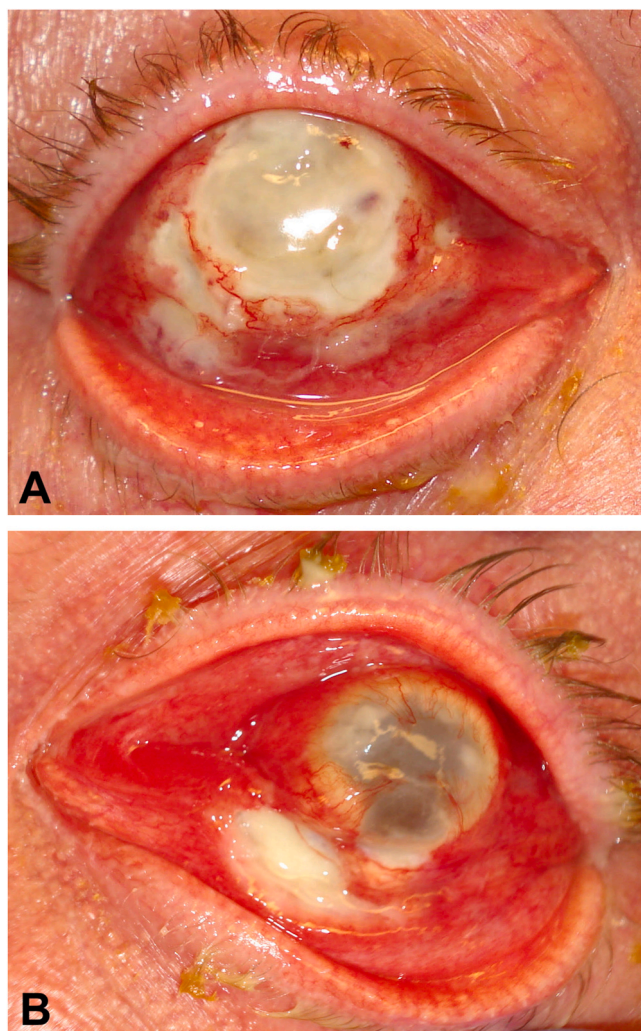


Fig. 1. External photographs of the patient's eyes. Right (A) and left (B) eyes at the pre-surgical examination show severe purulent acute inflammation mainly restricted to the corneas and anterior chambers. Both corneas are intensely vascularized, with areas of subconjunctival hemorrhage and necrosis of the sclera.

organisms. Elsewhere, the sclera showed focal scarring and chronic inflammation, but no direct invasion by *Acanthamoeba*. The pre-laminar regions in both globes were notable for chronic inflammation, but no *Acanthamoeba* organisms were present in these regions. Both optic nerves were atrophic, but without significant inflammation or fibrosis. PAS and GMS stains highlighted the presence of the amebic organisms in both eyes. These gross and histologic findings are consistent with a well-developed, bilateral granulomatous *Acanthamoeba* panophthalmitis.

2. Discussion

Given the rarity of the endophthalmic *Acanthamoeba* disease, it is unclear how the organisms extend from the corneal stroma to the posterior eye chamber. However, once the infection is established within the posterior chamber, the prognosis is devastating: the vast majority of cases result in persistent intraocular infection, eventually leading to enucleation.⁸ Interestingly in our case, the largest cavitory lesions in both globes were located in a relatively similar position, namely in the vicinity of the ora serrata. Scleral defects with severely inflamed and necrotic intraocular uveal tissue about these cavitory lesions. These scleral defects along with the associated necrosis and inflammation may suggest that the organisms gained entry into the posterior chamber through this region.

Although granulomatous meningoencephalitis is a feared complication of *Acanthamoeba* infection, only one case documents the co-occurrence of *Acanthamoeba* endophthalmitis and meningoencephalitis. This was a 7-year-old patient who had a history of wading in draining ditches and immersing himself in a cattle trough. He developed *Acanthamoeba* endophthalmitis and meningoencephalitis and later died from the infection. Pharyngeal colonization was believed to be the source of the ocular and CNS infections.¹¹

In contrast to granulomatous meningoencephalitis, immunosuppression or immunodeficiency has only been reported in two cases of *Acanthamoeba* endophthalmitis.^{5,8} Our patient had multiple comorbidities, including type 2 diabetes, asthma, and rheumatoid arthritis; he also received topical steroid therapy for his ocular infection. Although no specific risk factors have been identified that may have led to such an advanced ocular disease, it is known that corneal wound healing is significantly slower in diabetic patients. In conjunction with topical steroid treatment and altered immune response, intra-ocular penetration by *Acanthamoeba* may have been facilitated.^{12,13}

3. Conclusion

In summary, *Acanthamoeba* panophthalmitis and endophthalmitis are rare and devastating complications of *Acanthamoeba* keratitis. The

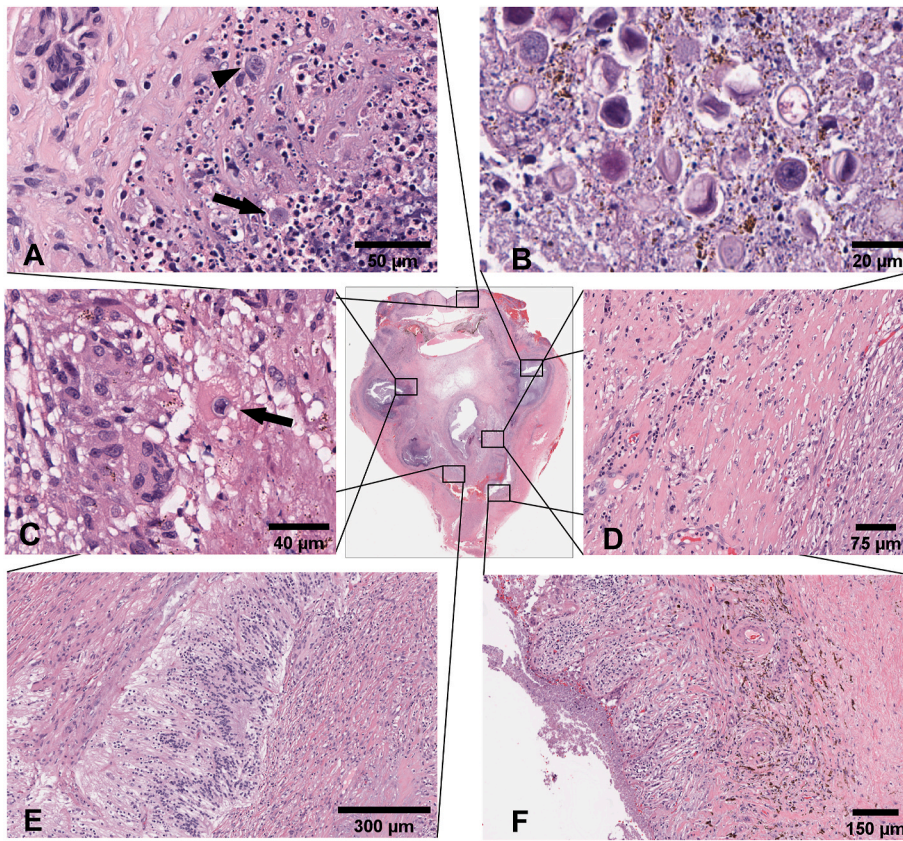


Fig. 2. Photomicrographs of the right eye (H&E). The figure shows a whole slide image of the right globe in the center and in high magnification images of different anatomic locations. (A) Acute inflammation surrounds *Acanthamoeba* trophozoite (arrow) and cyst (arrow head) forms. (B) Multiple *Acanthamoeba* organisms are demonstrated within the abscess. (C) *Acanthamoeba* cyst is seen in association with a multinucleated giant cell reaction at the edge of the abscess cavity. (D) Dense collagenous scar is seen within the vitreal cavity. (E) Dense fibrotic tissue with associated chronic inflammation surrounds the detached retina. (F) The edge of the pre-choroidal abscess cavity shows a distorted, fibrotic choroid layer with chronic inflammation.

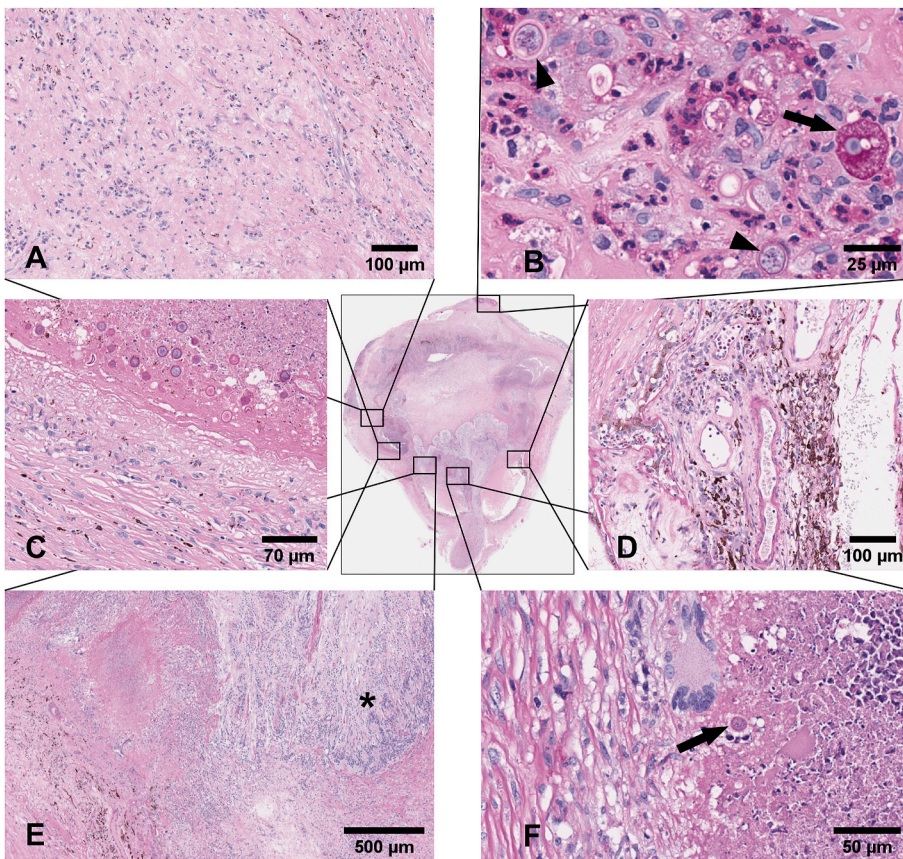


Fig. 3. Photomicrographs of the left eye (Periodic Acid-Schiff stain (PAS)). The whole slide image of the left globe in the center and in high magnification images various areas within the globe. (A) Scleral scarring and chronic inflammation. (B) *Acanthamoeba* trophozoite (arrow) and cyst (arrow head) forms in the cornea are surrounded by acute inflammation. The PAS stain highlights the abundant cytoplasm of the trophozoite and the thin rim of the cyst wall. (C) *Acanthamoeba* cysts and trophozoites at the edge of the pre-choroidal abscess, with associated multinucleated giant cell reaction at the edge of the abscess cavity. (D) Distorted choroid with chronic inflammation. (E) The abscess cavity separates detached retina (asterisk) from the choroid layer. (F) Multinucleated giant cell and *Acanthamoeba* cyst at the edge of the abscess cavity.

unique case presented here highlights the importance of prevention, early diagnosis, and treatment to avoid these complications.

Patient consent

Verbal consent to publish the case report was obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

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

Declaration of competing interest

All authors have no financial disclosures.

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References

1. Cope JR. Acanthamoeba keratitis among rigid gas permeable contact lens wearers. *United States*. 2016;123(7):1435–1441, 2005–2011.
2. Radford CF, Bacon AS, Dart JK, Minassian DC. Risk factors for acanthamoeba keratitis in contact lens users: a case-control study. *BMJ*. 1995;310(6994):1567–1570.
3. Schaumberg DA, Snow KK, Dana MR. The epidemic of Acanthamoeba keratitis: where do we stand? *Cornea*. 1998;17(1):3–10.
4. Burke JP, Webber SK, Kerr-Muir MG, Talbot JF, Parsons MA. Acanthamoeba polyphaga panophthalmitis. *Cornea*. 1992;11(3):274–275.
5. Heffler KF, Eckhardt TJ, Reboli AC, Stieritz D. Acanthamoeba endophthalmitis in acquired immunodeficiency syndrome. *Am J Ophthalmol*. 1996;122(4):584–586.
6. 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.
7. Mammo Z, Almeida DR, Cunningham MA, Chin EK, Mahajan VB. Acanthamoeba endophthalmitis after recurrent keratitis and nodular scleritis. *Retin Cases Brief Rep*. 2017;11(2):180–182.
8. Kutner A, Aldrich M, Patel S, et al. Acanthamoeba endophthalmitis during treatment for cutaneous disease in a renal transplant patient. 2018;20(2), e12843.
9. Davis MJ, Packo KH, Epstein RJ, Grostern RJ, Cohen JA. Acanthamoeba endophthalmitis following penetrating keratoplasty for Acanthamoeba keratitis. *Archives of ophthalmology*. 2010;128(4):505–506. Chicago, Ill : 1960.
10. Baker MS, Maltry AC, Syed NA, Allen RC. Orbital implant exposure after Acanthamoeba panophthalmitis. *American journal of ophthalmology case reports*. 2018;10:48–50.
11. Jones DB, Visvesvara GS, Robinson NM. Acanthamoeba polyphaga keratitis and Acanthamoeba uveitis associated with fatal meningoencephalitis. *Trans Ophthalmol Soc U K*. 1975;95(2):221–232.
12. Chen WL, Lin CT, Ko PS, et al. In vivo confocal microscopic findings of corneal wound healing after corneal epithelial debridement in diabetic vitrectomy. *Ophthalmology*. 2009;116(6):1038–1047.
13. Mutoh T, Ishikawa I, Matsumoto Y, Chikuda M. A retrospective study of nine cases of Acanthamoeba keratitis. *Clin Ophthalmol*. 2010;4:1189–1192.