First Case of Subretinal Ocular Angiostrongyliasis Associated with Retinal Detachment in the United States

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Abstract

Angiostrongylus cantonensis, commonly known as the rat lungworm, is mostly found in Asia, the Pacific Basin, and the Caribbean, but is also endemic in Hawai'i, especially on the Island of Hawai'i. Ocular angiostrongyliasis is an uncommon but previously reported complication associated with permanent vision loss. This is the first reported case of ocular angiostrongyliasis involving the retina or posterior segment of the eye in the US. A 24-year-old male from Chicago visited the Island of Hawai'i, where he worked on a farm and ate a vegetarian diet. When he returned to Chicago, he became sick and was hospitalized for eosinophilic meningitis. One month later, he developed a retinal detachment which required surgical repair involving a pars plana vitrectomy. During the reattachment of the retina during surgery, a live motile was identified nematode in the subretinal space. An endolaser probe immobilized and killed the nematode, and it was subsequently extracted through the sclerotomy. Thermal scars around all retinal holes including the retinotomy site were made to stabilize the retina, and perfluoropropane gas was injected to achieve temporary tamponade. Thereafter, the patient's cerebrospinal fluid returned positive for angiostrongylus cantonensis antibodies. During extended follow-up, the patient eventually lost all vision in the affected eye due to recurrent retinal detachment. This case of ocular angiostrongyliasis demonstrates the importance of obtaining travel history from endemic areas, knowing the risk of developing eosinophilic meningitis, and understanding the risk of permanent vision loss in cases involving the retina.

Keywords

angiostrongyliasis, rat lungworm, retinal detachment, ocular, eosinophilic meningitis, case report, Hawai'i, subretinal, posterior segment

Abbreviations and Acronyms

CDC = Centers for Disease Control and Prevention
CSF = cerebrospinal fluid
MRI = magnetic resonance imaging

Introduction

Ocular angiostrongyliasis is an uncommon complication occurring in 1.2% of infections caused by *Angiostrongylus cantonensis* commonly known as the rat lungworm.[1,2] Eosinophilic meningitis has been reported in half of the human ocular angiostrongyliasis cases, and common symptoms include headache, neck pain, paresthesia, and fever.[1] *A. cantonensis* is estimated to cause 29% of eosinophilic meningitis cases in Hawai'i.[3] Parasitic infections, a major source of ocular disease throughout the world, have become increasingly more common in nonendemic areas due to increased global travel.[4]

The life cycle of *A. cantonensis* begins in the pulmonary arteries of rats where the eggs are laid by adult worms. Subsequently, the first-stage larvae migrate to the pharynx and are swallowed and excreted. The intermediate hosts, typically snails or slugs, ingest the rat feces and become infected. Ultimately, humans acquire the infection by ingestion of infective larvae found on or in snails, snail tracks, slugs, shellfish, and raw vegetables. The infective larvae migrate to the brain and mature, frequently causing eosinophilic meningitis which may lead to death.[5]

A. cantonensis has an expanding range of endemicity that includes Southeast Asia, the Pacific Islands, South and Central America, and the Caribbean.[6] Currently, Hawai'i is the epicenter for angiostrongyliasis in the United States due to the proliferation of the mollusk *Parmarion martensis*, a highly effective intermediate host that transmits the disease to humans and other susceptible animals.[7] Additionally, the high consumption of local produce and use of rainwater for harvesting likely elevates the risk of acquiring angiostrongyliasis, particularly on the east side of Hawai'i Island.[8] Between 2007 to 2017, a total of 82 cases of *A. cantonensis* infections were identified in Hawai'i.[9]

Ocular angiostrongyliasis has been reported in Asia, South America, and North America[2,10-13] The first case of ocular angiostrongyliasis in the United States was described in Miami, FL, and involved a nematode that had infiltrated the anterior chamber of the eye.[14] After a review of the existing literature, this is the first reported case of ocular angiostrongyliasis involving the retina in the United States.

Case Report

A 24-year-old male presented to a Chicago hospital with a 1-day history of fever and headaches. He also complained of painful lower extremity paresthesia, bilateral leg weakness, and significant weight loss. The patient had recently moved from Hawai'i back to Chicago following a 14-month stay in Hawai'i where he adopted a vegetarian diet and became sick while working on a farm. Previously, the patient was otherwise healthy without any significant past medical history. The cytology of blood and cerebrospinal fluid (CSF) were remarkable for eosinophilia. The brain magnetic resonance imaging (MRI) was within normal limits, but the spine MRI identified the presence of transverse myelitis. The stool sample contained no ova or parasites. The patient was diagnosed with eosinophilic meningitis and treated empirically with ivermectin and high-dose corticosteroids.

One month later, the patient suddenly developed unilateral vision loss, and a complete ophthalmologic exam was performed. Visual acuity in the right eye was limited to the ability to count the examiner's fingers, and the left eye was 20/20. Slit lamp examination revealed the absence of inflammatory cells in the anterior chamber with 1+ pigmented cells in the anterior vitreous. Dilated fundus examination revealed a rhegmatogenous retinal detachment with significant epiretinal membranes on the surface of the retina consistent with proliferative vitreoretinopathy. A retinal detachment repair involving a pars plana vitrectomy using a standard 3-port 23-gauge vitrectomy was planned. Following removal of the vitreous and the epiretinal membranes, perfluorocarbon liquid was used to reattach the retina and express out the subretinal fluid, due to the liquid's high specific gravity.

As the retina was reattached, a live motile nematode in the subretinal space inferonasal to the optic disc became visible. Transmitted heat through the endolaser probe thermal laser was used to immobilize and kill the subretinal nematode, and a retinotomy was performed to access the subretinal space. The nematode was grasped using microforceps and retrieved from the eye through the sclerotomy after removing the 23-gauge trocar [Figure 1]. A fluid-air exchange followed by air-gas exchange was performed using perfluoropropane gas (C3F8) to allow for support of the retina by intraocular tamponade temporarily. Laser was applied to the retinotomy site as well as the peripheral retina to create a thermal scar around all retinal holes, while also stabilizing the peripheral retina.

Subsequently, the patient's CSF returned positive for angiostrongylus cantonensis antibodies via testing at the Centers for Disease Control and Prevention (CDC). On extended follow-up, the patient suffered recurrent retinal detachment, which persisted despite further vitreoretinal surgeries. At most recent follow-up, the patient's visual acuity in the right eye had decreased to light perception with hypotony with an intraocular pressure of 5 mmHg with early phthisis. The unaffected left eye has normal vision with no evidence of any infection.

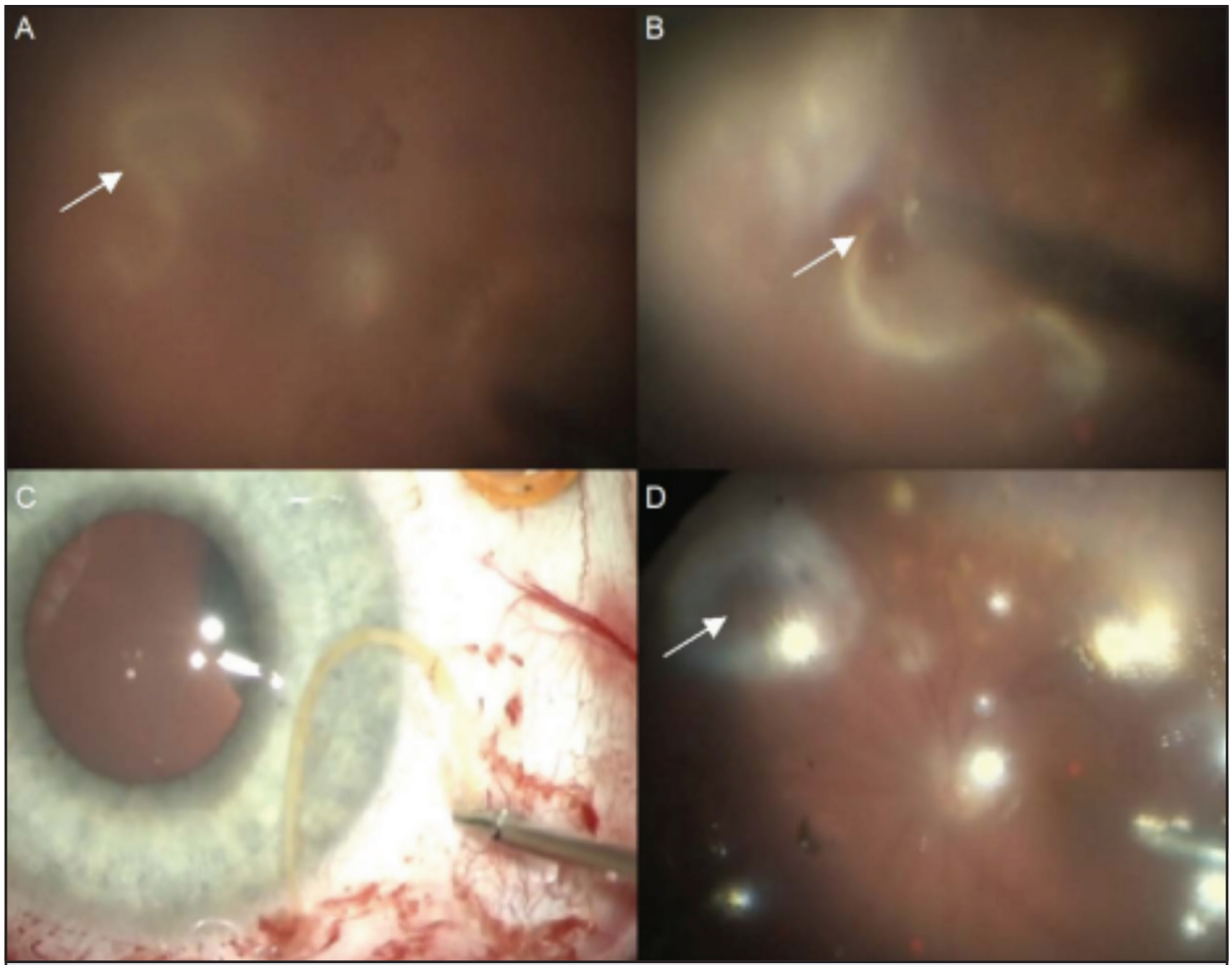

Figure 1. Intraoperative Photos of Surgical Technique. (A) Visualization of inferonasal subretinal nematode (arrow). (B) Retinotomy (arrow) made to access subretinal space. (C) Nematode on the conjunctiva following removal through sclerotomy. (D) Laser of retinotomy site (arrow) following extraction.

Discussion

Ocular angiostrongyliasis is a rare but potentially sight-threatening infection caused by *A. cantonensis*. A high index of suspicion should be maintained in patients recently diagnosed with eosinophilic meningitis who present with ocular symptoms such as vision loss. Additionally, a recent history of travel to endemic areas and high-risk dietary behaviors may further help to identify parasitic infection and *A. cantonensis* infection. Eosinophilia in the CSF has been reported in many cases, and angiostrongyliasis is one of the most common causes of eosinophilic meningitis.[15] MRI findings are typically unremarkable but may demonstrate non-specific findings such as cerebral edema, meningeal enhancement, and hyperintense signal lesions.[16] The use of highly sensitive and specific enzyme-linked immunosorbent assays is currently not widespread, although the CDC testing in this case confirmed angiostrongyliasis antibodies.[17] Ocular complications depend on the location and extent of infiltration and have included uveitis, macular edema, panophthalmitis, papilledema, optic neuritis, optic nerve compression, and orbital inflammation.[4] In case reports by Sinawat et al, fundus examination associated with optic neuritis in ocular angiostrongyliasis included abnormalities in the retinal pigment epithelium, retinal and macular edema, and subretinal tracks.[12]

The route taken by *A. cantonensis* to enter the eye is unknown, although it is postulated that the nematode may enter the eye via the optic nerve sheath, through the central retinal artery, or directly from the ocular surface. One theory is that the nematode may travel along the surface and base of the brain leading to the meningitic symptoms. Upon reaching the optic nerve, the nematode may travel between the nerve and sheath to reach the retina by moving through the lamina cribosa.[12]

Treatment of ocular angiostrongyliasis includes reducing pain and inflammation while immobilizing and removing the helminth. A 2-week course of systemic corticosteroids has been shown to significantly improve headaches, duration of headaches, and number of repeat lumbar punctures.[18] The use of anthelminthic medications to treat acute eosinophilic meningitis requires further investigation in terms of both efficacy and side-effect profile. In a study of 71 patients with acute eosinophilic meningitis, albendazole has been shown to decrease the duration of disease and reduce the use of acetaminophen without any associated serious side effects.[19] However, anthelminthics should be used with caution because necrotic parasite tissue may release toxic substances and further exacerbate intraocular inflammation.[20,21] Nevertheless, albendazole remains the anthelminthic of choice compared to other benzimidazoles due to its better penetration into the central nervous system.

The definitive treatment of ocular angiostrongyliasis involves surgical removal of the parasite. In most cases of ocular angiostrongyliasis, there is only 1 nematode in the eye, and the nematode is usually still alive.[1] The surgical technique for removal of the nematode is dependent on the location of the nematode. If the nematode is in the anterior chamber, an approach through a corneal incision has been effective at removing the nematode.[14] A patient in Japan with subretinal ocular angiostrongyliasis underwent vitreous surgery to remove the nematode by making an incision in the retina to access the subretinal space.[13] Focal laser photocoagulation to the nematode has been reported to be helpful in immobilizing and possibly killing the parasite prior to surgical removal.[20] A case report by Kanchanaranya et al also recommended immobilization of the nematode by cryopexy before removal.[22] Complications, such as hemorrhage or post-operative retinal detachment, can be reduced by careful cauterization of bleeding sites, and laser around the retinotomy site. In this case, the risk of recurrent retinal detachment was high due to significant proliferative vitreoretinopathy, which portends a poor visual outcome due to the associated inflammation and scarring. Despite the measures taken to reattach the retina, the inflammation and scarring thereafter contributed to recurrent retinal detachment, leading to complete functional blindness in this right eye.

Surgical outcomes after removal of the nematode depend on the extent of inflammation, location of the nematode, involvement of the retina, and damage to the retina or optic nerve due to prior inflammation. In one of the largest case series on ocular angiostrongyliasis (N = 18), surgical removal was successfully performed in 10 cases (56%), but none of the cases presented with rhegmatogenous retinal detachment as in this current case report.[23] Posterior segment cases most commonly involved the vitreous, the subretinal space, or optic neuritis.[19] Other treatment modalities included focal laser (78%), anthelminthic drugs (61%), and steroids (89%). Unfortunately, visual acuity did not change dramatically in 67% of cases regardless of treatment type. In another report, parasites were successfully removed from the anterior chamber in 14 cases and the vitreous fluid in 15 cases.[20] However, there was only slight improvement in visual acuity. Anterior segment involvement is more favorable with a better chance of recovering good vision than posterior segment involvement of the retina, vitreous, or optic nerve.[2]

Practical measures to prevent angiostrongyliasis include proper hygiene by washing vegetables thoroughly and avoiding eating raw snails and other immediate hosts (eg, crabs and shrimp).[11] Education of the general public regarding the dangers of raw mollusk consumption can be particularly helpful in endemic areas where food is home grown or collected locally. Prevention of ocular angiostrongyliasis includes timely diagnosis and treatment of systemic angiostrongyliasis, and immediate ophthalmic evaluation for any patient presenting with ocular symptoms of decreased vision and floaters.

This is the first reported case in the US of ocular angiostrongyliasis with vision loss due to retinal detachment associated with a subretinal nematode. This patient developed vision loss in Chicago, but he acquired the disease on the Island of

Hawai‘i, which is a known endemic area. This highlights the importance of obtaining recent travel history from an endemic area for angiostrongyliasis. He also first developed eosinophilic meningitis, which has been noted in half of cases with angiostrongyliasis and should be a warning sign for possible infection. The definitive treatment for ocular angiostrongyliasis is surgical removal of the nematode, often after initial treatment with laser to immobilize or kill it. This case highlights the risk of severe vision loss in ocular angiostrongyliasis, especially when the posterior segment of the eye is involved.

Conflict of Interest

None of the authors identify a conflict of interest.

Acknowledgments

The authors acknowledge the editorial assistance of Maria Won, research coordinator at Hawai‘i Macula and Retina Institute.

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