

Case Report

Baylisascaris Procyonis Induced Diffuse Unilateral Subacute Neuroretinitis in New York City

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Abstract Diffuse unilateral subacute neuroretinitis (DUSN) secondary to raccoon roundworm (*Baylisascaris procyonis*) infection has been reported in rural and suburban areas of North America and Europe with extant raccoon populations. Here, we present a case of *Baylisascaris*-induced DUSN from the densely populated borough of Brooklyn in New York City and alert urban ophthalmologists to consider this etiology even in areas not typically thought to be associated with endemic risk factors. Infected raccoons also occur in urban settings, and urban patients may be exposed in surrounding areas. Most patients with *Baylisascaris* ocular larva migrans-DUSN will not have concomitant neurologic disease; this fact and larval neurotropism are both misconceptions regarding this infection.

Keywords *Baylisascaris procyonis*, raccoon roundworm, ocular larva migrans, DUSN, diffuse unilateral subacute neuroretinitis

1 Introduction

Diffuse unilateral subacute neuroretinitis (DUSN) is a progressive inflammatory and degenerative disease affecting the retina, retinal vessels, and optic nerve head, secondary to infection with nematode and possibly other helminth larvae [10, 11]. Thus, DUSN is a clinical or pathologic form of ocular larva migrans (OLM). The raccoon roundworm, *Baylisascaris procyonis*, is considered the primary cause of the large nematode variant of DUSN, with intraocular larvae in the 1500–2000 μm size range [13, 15]. Cases

of *Baylisascaris* DUSN are usually observed in regions where raccoons, the definitive host, are found such as rural and suburban communities in the northern and Midwestern United States [10, 13, 15]. This entity has been less often reported within densely urban communities, even though raccoons are also known to occur in this setting [13]. Unfortunately, this environmental bias may cloud consideration of this etiology and lead to delayed diagnosis and poor visual outcomes. Nevertheless, as raccoons have migrated from rural to urban areas, more city dwellers will be exposed to the raccoon roundworm and may suffer grave consequences [22]. Here, we present a notable case in order to alert ophthalmologists in urban settings to consider DUSN as well as *B. procyonis*. Recently, a New York City Department of Health public health advisory was released, in response to this case as well as another of our urban patients, an infant with *B. procyonis*-induced neural larva migrans (NLM), who suffered permanent severe neurological impairment [21].

2 Case report

A 15-year-old female from Brooklyn, New York, presented with severe painless vision loss in the right eye discovered incidentally on a routine eye exam. The onset of the visual loss was not clear. The patient had no significant ocular or medical history, including direct contact with animals. However, there had been documented sightings of raccoons rummaging through garbage cans near her home in the borough of Brooklyn, New York City, and a raccoon was also seen by family members on their front porch. The patient's travel

history was significant for travel to a rural area in Sullivan County in lower New York State the previous year, and she denied any history of pica.

The patient's best-corrected visual acuity was finger counting at two feet in the right eye and 20/20 in the left eye. Examination of the right eye was significant for an afferent papillary defect, no stigmata of anterior uveitis, diffuse retinal pigment epithelium mottling, arteriole attenuation, and optic nerve pallor. The initial differential diagnoses included inflammatory, infectious, or degenerative etiologies, although the ocular findings were also consistent with diffuse unilateral subacute neuroretinitis (DUSN) [10,11]. Laboratory tests were negative for eosinophilia, serologic evidence of *Toxocara canis* and *T. cati*, toxoplasmosis, Herpes zoster, *Treponema pallidum* antibody, angiotensin converting enzyme, human leukocyte antigen B-27, rheumatoid factor, anti-nuclear antibody, C-reactive protein, erythrocyte sedimentation rate, and lysozyme activity. A full-field electroretinogram revealed a normal left eye, but a diffuse reduction in rod and cone responses in the right eye, corroborated in the multifocal electroretinogram with extensive reduction throughout the macula.



Figure 1: Color fundus photograph of the right eye, demonstrating a coiled subretinal nematode in the inferior macula (arrow), arteriole attenuation, pigment epithelium mottling, and optic nerve pallor.

Fluorescein angiography confirmed diffuse abnormality of the retinal pigment epithelium. While not appreciated ophthalmoscopically, fundus photography documented a motile worm in the subretinal space (Figures 1 and 2). Upon morphometric evaluation of the parasite, its length was determined to be approximately 1850 μm , consistent with a *Baylisascaris* larva. A diagnosis of DUSN secondary to

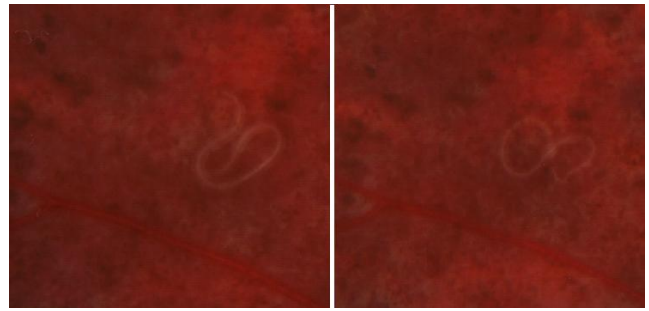


Figure 2: Color fundus photograph of the right eye, after one (left) and two (right) weeks later, demonstrating a magnified view of the alive, uncoiled, motile nematode.

B. procyonis infection was considered in this case. The worm was treated directly with photocoagulation using the green diode laser (at settings of 200 mW, 0.2 seconds, 200 μm spot size) without incident or use of systemic steroids (Figure 3).



Figure 3: The worm subsequently moved superiorly, and was photocoagulated.

There was no return of vision despite successful eradication of the worm. Serology for *Baylisascaris* infection was performed at Purdue University, and the patient gave a weakly positive reaction to *B. procyonis* larval excretory-secretory antigens by enzyme-linked immunosorbent assay. The New York City Department of Health collected raccoon feces and soil samples from both the patient's home and a small city playground she frequented. These were examined for parasite eggs at Purdue University, and although the three samples were found to be negative for *B. procyonis* eggs, this did not preclude her infection from another local source.

3 Discussion

As first proposed in the mid-1980s, DUSN is a multi-etiological syndrome, being caused by several possible species of nematodes whose larvae fall into different size ranges [10,13,15]. *B. procyonis* is the primary cause of the large nematode variant (1500–2000 μm) and is the most common cause of DUSN in the northern and midwestern United States and Canada, with cases also reported from Europe [16]. The small nematode variant of DUSN may be caused by *Toxocara* spp. (400 μm) or hookworm larvae (*Ancylostoma caninum* or others; 500–700 μm), both parasites implicated in DUSN cases in the southeastern United States, Caribbean, and South America [4,6,7,10]. At least two cases of DUSN have also been caused by trematode larvae (*Alaria* spp.) indicating a probable common pathogenesis of ocular lesions following helminth larval invasion [18]. In our case, the larva size and morphology allowed for identification of the parasite as *Baylisascaris* spp. rather than the smaller *Toxocara* spp. or *Ancylostoma* spp., and based on the patient's exposure history *B. procyonis* was considered the likely cause.

B. procyonis is the most common cause of clinical larva migrans in animals, having produced NLM in over 130 species of birds and mammals, including humans [14, and K. R. Kazacos, unpublished]. The parasite is being recognized with increasing frequency as a cause of human NLM and OLM, although covert or asymptomatic infections are more common [12,14,20]. Human infection risk with *B. procyonis* is greatest for those exposed to areas or objects contaminated with raccoon feces. *B. procyonis* eggs are extremely hardy, infective for years in the soil, and are resistant to destruction by most chemicals and environmental conditions [14].

Humans, accidental hosts, are affected in three clinical states: visceral larva migrans (VLM), NLM, and OLM, which includes DUSN [14]. In contrast to NLM, which almost exclusively affects infants and toddlers who engage in geophagia of soil contaminated with raccoon feces and *B. procyonis* eggs, isolated OLM and DUSN usually occur in otherwise healthy adults where there is no obvious exposure to raccoons or their feces, suggesting that DUSN follows ingestion of relatively few *B. procyonis* eggs, lowering the chance of CNS involvement [20]. Moreover, given that raccoon feces and contaminated soil may contain thousands of eggs, eosinophilia is more frequent in geophagia-associated NLM, and usually is absent in the incidental raccoon exposure in cases of OLM-DUSN [12,20]. Thus, the epidemiology and clinical presentation of OLM due to *B. procyonis* parallels that of the more common *Toxocara* spp.

Having entered the human alimentary tract and migrated systemically and then intraocularly, *B. procyonis* may

present with early DUSN changes of vitritis, retinitis with migration tracks, retinal vasculitis, optic neuritis, and choroidal infiltrates [10,11,14,19]. Later DUSN changes include optic atrophy, narrowing of retinal vessels, and multifocal to diffuse retinal pigment epithelial degeneration [10,11]. Chronic endophthalmitis with retinal detachment and posterior pole granuloma may also be seen [1,19]. Despite its broad ophthalmic manifestations, the subretinal nematode itself can actually be difficult to identify depending on where it is located. In our case, biomicroscopy failed to identify the parasite, and it was only later identified on the fundus photograph from the initial visit (Figure 1), clearly attesting to the “photographer’s diagnosis” lesson of retinal pathology [2].

When the larva is visualized intraocularly, DUSN has been treated successfully with thermal laser photocoagulation, possibly combined with a short course of systemic steroids to blunt the inflammatory response to the necrotic helminth [13,18,23]. Care has to be taken not to lose sight of the worm, since it may disappear from view or escape into the vitreous, necessitating surgical removal [3,7,18]. Systemic anthelmintic therapy does not appear to be universally effective, but may be considered if no worm can be found, or if there is suspicion of bilateral involvement or presence of another worm, and obviously in the case of concomitant systemic involvement [4,6].

Over the years, several misconceptions concerning *B. procyonis* and DUSN have been perpetuated and the present case should help dispel them. When *B. procyonis* was first proposed as an etiology of DUSN, it was downplayed by others, who stated that “several patients with DUSN lived in highly urban centers in the northeast and Midwest, making exposure to raccoons highly unlikely” [17]. This was and remains untrue, based on the present case, other cases from urban localities, and the well-known occurrence of raccoons in some of our largest cities. Indeed, urban raccoon populations carrying *Baylisascaris* have been documented recently in metropolitan Atlanta, Georgia; Portland, Oregon; Chicago, Illinois; the Twin Cities metroplex in Minnesota; Orange County, California; and Toronto, Canada [8,9,14,22,24]. This parasite must be considered a possible infection and cause of DUSN in urban settings. Secondly, it has been repeatedly stated by DUSN authorities that *B. procyonis* is less likely involved since none of the patients also had severe or fatal neurologic disease [5,17]. However, this is as it should be and is a major ongoing misconception about this infection, which clearly demonstrates dose-related differences in both animals and humans.

Since relatively low percentages of larvae (est. 5–7%) actually enter the CNS following oral infection [14], in low dose infections humans will not normally develop clinical CNS disease. In high dose infections (e.g., in infants with geophagia) OLM-DUSN may be seen in combination with

NLM due to widespread systemic invasion [12,19,20], but for most cases of OLM and DUSN involving this parasite, only ocular disease will be manifest. It is important to note that *B. procyonis* is not a neurotropic parasite, but an accidental invader of both the CNS and the eye. In the majority of human infections, OLM-DUSN will occur without any evidence of CNS involvement, based on low systemic infection levels and chance migration into the eye. Finally, in cases such as these, if DUSN is suspected then repeated examinations should be performed in an effort to locate the worm, including a thorough evaluation of all fundus photographs and other imaging modalities. The larva can exist in the eye and migrate for an extended time period, usually weeks to months but in some cases apparently years [10]. Thus, early consideration, diagnosis and treatment are both imperative for the best hope of maintaining visual acuity in the affected patient.

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