CASE STUDY

Baylisascariasis: A young boy with neural larva migrans due to the emerging raccoon round worm

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Abstract

A 17-month-old boy from Vancouver, Canada, presented with a 5-day history of progressive somnolence, ataxia, and torticollis. Additional investigations revealed eosinophilic encephalitis with deep white matter changes on MR imaging. On day 13, serology came back positive for Baylisascaris procyonis antibodies. While prophylaxis after ingestion of soil or materials potentially contaminated with raccoon feces can prevent baylisascariasis, timely treatment can sometimes alter a disastrous outcome. Populations of infected raccoons are propagating globally, but cases of Baylisascaris neural larva migrans have so far only been reported from North America.

Introduction

Human baylisascariasis is primarily caused by larvae from the raccoon roundworm Baylisascaris procyonis.¹,² Eggs that are excreted in raccoon feces become infective after 2–4 weeks and can remain so for years. When contaminated soil is ingested, Baylisascaris larvae can migrate through the brain (neural larva migrans; NLM), eye (ocular larva migrans; OLM), and other organs (visceral larva migrans; VLM). Baylisascaris procyonis is indigenous to North America but is emerging in Europe together with its host.³

Case Study

A previously well 17-month-old boy presented to the emergency department with a 5-day history of progressive somnolence, ataxia, bradykinesia and new onset torticollis and fluctuating visual inattention.

He lived in suburban Vancouver, British Columbia, Canada, with his family, who was well. He never travelled, had no pets, and had been playing mainly indoors during the winter months that preceded presentation. Vital signs were normal and he was afebrile. He was sleepy but could be roused. Both eyes were deviated maximally to the right with sluggish pupillary reactions. There was discrete left nasolabial fold flattening and correctable right-sided torticollis. Appendicular muscle tone was increased throughout with spastic catches at the knees, two beats of ankle clonus, brisk deep tendon reflexes, and positive Babinski sign bilaterally. There was bilateral coarse tremor of both arms accentuated by movement. Ophthalmologic examination demonstrated chorioretinitis and profound vision loss of the left eye.
Blood work revealed elevated erythrocyte sedimentation rate (22 mm/h), c-reactive protein (16 mg/L) and white blood cell count (22.9 x 10^9/L) with 42% eosinophils. Cerebrospinal fluid (CSF) analysis revealed an elevated nucleated cell count of 34 x 10^6/L with 60% lymphocytes, 28% monomacrophages, and 11% eosinophils. CSF opening pressure, glucose, and protein were within normal range. CSF PCR was negative for HSV 1 and 2, EBV, CMV, VZV, and enterovirus. CSF culture remained negative.

Magnetic resonance imaging (MRI) revealed multiple discrete T2/FLAIR hyperintensities and T1 hypointensities within the cerebral juxta-cortical white matter and cortex with gadolinium enhancement (Fig. 1Ai/Bi). Electroencephalography was compatible with moderate to severe encephalopathy without epileptiform activity.

Serologic tests did not detect evidence for EBV, HIV, Bartonella, Mycoplasma pneumoniae, syphilis or Toxoplasma infection.

Dilated fundus examination under anesthesia on day 6 showed vitritis and granulomatous deposits on the iris in the pupillary area (Koeppe nodules) and the anterior lens surface of the left eye.

In light of the MRI and eye findings in the setting of eosinophilic encephalitis, the patient was empirically treated for toxocariasis with a 5-day course of mebendazole 200 mg PO twice daily and intravenous methylprednisolone pulse.

On day 13, serology returned negative for *Toxocara*. However, *Baylisascaris procyonis* infection was diagnosed based on serum antibodies against the 37 Kd rBpRAG1 protein in the context of a compatible clinical picture. Raccoons are found widely throughout Vancouver, but presence around the house was denied by the family. The boy had however occasionally put driveway pebbles in his mouth.

Mebendazole was restarted while special access permission for albendazole was obtained. Consecutively he was switched to albendazole 400 mg PO daily for 28 days together with an oral prednisone weaning regimen.

Repeat MRI at 16 days demonstrated diffuse confluent white matter T2 hyper and T1 hypo-intensities without gadolinium enhancement and generalized atrophy (Fig. 1Aii/Bii). MRI at 63 days revealed improvement of the white matter signal abnormalities and progressive generalized atrophy (Fig. 1Aiii/Biii).

Initially there was only minor improvement of his clinical condition with significant developmental regression and multiple cognitive, visual, and mobility deficits. However, on day 19 he was transferred to a rehabilitation...
center where he continued to improve. And 3 years after presentation he remains developmentally delayed, but he is able to follow simple 2-step commands, say a few words, and, despite mild spasticity and bilateral optic nerve atrophy, runs unassisted without severe visual impairment (visual acuity 20/30). He has no seizures and does not receive regular medications. His MRI shows diffuse mild atrophy of cerebrum, cerebellum, pons and thalami with delayed myelination and left optic atrophy (Fig. 1Aiv).

Discussion

In 1984, the first human baylisascariasis case was recognized in a 10-month-old Pennsylvania boy with pica. He received no specific treatment. His condition was diagnosed post-mortem by the detection of Baylisascaris procyonis larvae on brain autopsy.

Less than 30 Baylisascaris NLM cases have been described in the literature. Most were children of which most were boys under the age of 2. Several children were reported with antecedent developmental delay and/or a history of pica/geophagia and many had been observed eating dirt or sucking on debris like wood chips from areas known to be frequented by raccoons. In most NLM cases there was no treatment effect with severe residual neurological deficits, often cortical blindness. Five of 23 children with NLM died.

In four children, different degrees of improvement were noted: A 7-year-old autistic boy from Toronto presented with a clinical picture that was very similar to our case, 2 weeks after his first symptoms. His parents reported a raccoon latrine in the backyard and the child often played in an open sandbox with a habit of putting his hands in his mouth. Empiric treatment for baylisascariasis with albendazole and methylprednisolone was initiated within 48 h of admission. Baylisascaris procyonis serology returned positive afterwards and they identified Baylisascaris eggs in his sandbox. He improved with treatment, becoming more alert with resolution of his gaze deviation. By 4 months he could not speak, had cortical visual impairment and a seizure disorder, but he was ambulatory, interactive and reportedly regained a relative good level of functioning. Gradual improvement after treatment was also reported in a 14-month-old Massachusetts toddler who was admitted 9 days after symptom onset and started treatment with albendazole and methylprednisolone 3 days later. Full recovery was reported in a 4-year-old Louisiana boy with sickle cell disease. He was admitted after 1 day of relatively mild symptoms, started on dexamethasone on day 5 and on albendazole on day 9. These cases with better improved outcome are noteworthy for the relatively early administration of steroids and albendazole, although early treatment does not guarantee success.

One Oregon teenager received only steroids and also improved over time. A 13-month-old from California with biopsy-proven baylisascariasis however continued to deteriorate neurologically despite an initial response to treatment with steroids only. Other children who received only steroids did not demonstrate a favorable treatment response.

When interpreting these results, one should know that positive Baylisascaris procyonis serology can also be encountered in healthy subjects from areas with infected raccoons. The presence of diffuse deep white matter changes on MRI has been suggested as characteristic of baylisascariasis. Except for the 4-year-old from Louisiana that fully recovered, the above cases that improved all had these typical MRI findings.

The American Academy of Pediatrics Redbook states that, on the basis of CSF penetration and in vitro activity, albendazole, in conjunction with high-dose corticosteroids, has been advocated most widely. Albendazole dosing and duration have been specified by the Centers for Disease Control and Prevention as 25–50 mg/kg/day for 10–20 days. If not immediately available, mebendazole or ivermectin may be used in the interim. Others, however, argue against the use of ivermectin because it does not penetrate well into the central nervous system. Associated OLM may be treated by direct killing of the larvae by photocoagulation. Post-exposure prophylaxis with albendazole should be considered for children who ingested soil or sucked on materials potentially contaminated with raccoon feces. Mice treated with oral albendazole on days 1–10 post infection were 100% protected from developing NLM, while all untreated mice died. Primary prevention consists of monitoring children at play, handwashing, and clearing of raccoon latrines (while wearing appropriate personal protective equipment).

Besides three Canadian cases (two children from Ontario and one asymptomatic elderly person from British Columbia), all the reported Baylisascaris NLM patients lived in the U.S., which has an estimated raccoon population of 5 million. Germany now has 1 million raccoons, the consequence of a steady propagation since their introduction in 1934 for hunting and fur. More than 70% of the German raccoons are infected with Baylisascaris procyonis. Raccoons spread to at least 20 other Eurasian countries. So far, the only human baylisascariasis cases reported from Europe concerned one German OLM patient who kept a raccoon pet and one Austrian OLM case where infected raccoons lived in the area. The importance of vigilance to timely recognize and prevent more baylisascariasis cases should be stressed. To the best of our knowledge, the only other...
human baylisascariasis cases reported from the rest of the world concerned two OLM cases from Brazil where the infective source was not clear but with links to dogs whose intestines can also harbor Baylisascaris worms and to non-raccoon procyonid hosts such as kinkajous or coatis. Over the last 4 decades more than 20,000 pet raccoons have been introduced in Japan, but so far only animal Baylisascaris NLM cases have been reported. China has also reported infected raccoons, but no human cases.

In conclusion, findings of encephalopathy with eosinophilia and/or chorioretinitis should prompt brain MRI and serologic testing and treatment for baylisascariasis, particularly in toddlers or patients with intellectual disability who may come in contact with soil contaminated with raccoon feces, as timely treatment may improve prognosis and survival.

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Conflict of Interest

The authors declare no conflicts of interest.

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