CASE REPORT

Antonio Boschetti, M.D.[1] *and John Kasznica, M.D.*[2]

Visceral Larva Migrans Induced Eosinophilic Cardiac Pseudotumor: A Cause of Sudden Death in a Child

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ABSTRACT: A case of fatal cardiac larva migrans in a 10-year-old boy is described. The autopsy findings were quite dramatic, with a bosselated, sessile polypoid mass involving the left ventricular myocardium and protruding into the ventricular lumen. The precise morphologic characterization of the zoonotic ascarid larva was impaired by advanced resorption of the larva by an inflammatory infiltrate. Nonetheless, morphometry of the larval remnants strongly suggested the raccoon ascarid, Baylisascaris procyonis, as the causative agent.

KEYWORDS: pathology and biology, visceral larva migrans, heart, Baylisascaris procyonis, eosinophilic cardiac pseudotumor

Visceral larva migrans is a rather well known clinical entity in tropical as well as temperate zones. The basis for this parasitic disease entity is the presence of a larval parasite, (most often a helminth) in an unsuitable final host, in which the larva fails to mature. The larva is usually agitated, "dogged" by host immune defenses, and wanders through the viscera, to which it arrives from the blood or lymph. It often wreaks havoc in the involved organ or organs, before it dies and is resorbed. Sometimes the host organism is extremely ill, oftentimes with bizarre symptoms; fatalities may occur, especially in children if the brain or other vital organ is affected. In humans, probably the single most important clinical feature is a sometimes very high absolute and relative peripheral eosinophilia. Other features are malaise, fever, and accompanying allergic rashes. Local signs (including neurologic signs) are extremely variable.

This case is particularly interesting to medical examiners and pathologists because it discusses a rare case of cardiac larva migrans causing a sudden and unexpected death. The visceral larva migrans infections may cause death in children with no significant past medical history. In fact, some cases of larva migrans do not immediately shed light on human—animal contact, and only after careful questioning of parents or guardians, details of sand box play, pets, and/or geophagia are elicited.

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[1]Medical Examiner for Suffolk County, Commonwealth of Massachusetts, and instructor in pathology, Boston University School of Medicine, Boston, MA.

[2]Associate pathologist, Mallory Institute of Pathology, Boston, MA and assistant professor of pathology and obstetrics and gynecology, Boston University School of Medicine, Boston, MA.

Case Report

A ten-year-old boy complained of recurrent abdominal pain and in the evening was found to be unresponsive by his father. He was transported by ambulance to a central hospital, where in spite of continued resuscitation efforts he was pronounced dead. Because the sudden unexpected death occurred within 24 hours of admission, the autopsy was performed by the medical examiner.

On questioning the parents, it was discovered that the boy had several bouts of abdominal pain in the past weeks. During a recent clinic visit for the abdominal pain, peripheral blood eosinophilia with a differential of 15% was the only salient clinical laboratory finding. Stool examination for ova and parasites was negative.

Importantly, the past medical history included viral Hepatitis C, and hyperkinetic syndrome (minimal brain dysfunction syndrome) for which the boy was on therapeutic doses of methylphenidate HCl. Postmortem examination revealed a black male child with a weight of 80 pounds and length 58 inches. There were no external congenital anomalies. There was no evidence of trauma or violence.

Internal examination revealed the main pathologic changes to be concentrated in the thorax. The heart was found to contain an ovoid pale yellow-tan mass arising from the lower anterior wall of the left ventricle and neighboring interventricular septum (Fig. 1). The mass bulged well into the chamber of the left ventricle and its protruding surface was smooth and faintly bosselated. On gross cut section, it vaguely resembled a lymphoma, because of its nodularity and its fish-flesh-like character.

Indeed, the suspicion was raised even further, because there were some markedly enlarged abdominal para-aortic lymph nodes, measuring up to 5 cm in maximum dimension. On gross cut section, the nodularity of the lymph nodes was quite similar to that of the cardiac lesion.

Microscopic examination of the lymph nodes did not reveal any malignancy, but showed very severe reactive lymphadenopathy with marked follicular hyperplasia.

The microscopic changes in the heart, especially in the center of the lesion, were very dramatic. The nodules were found to be tumorlike collections composed entirely of eosinophilic granulocytes. The myocardium distant to the lesion was relatively unremarkable, but the myocardium between the eosiphilic nodules,

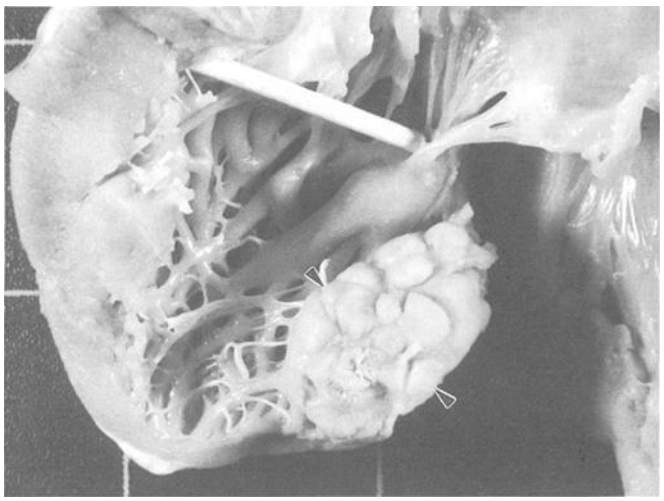

FIG. 1—*Gross photograph of the opened heart showing the prominent nodularity of the cut surface of the pseudotumor* (between arrows). *It is primarily located in the left ventricular myocardium, and bulges into the lumen.*

showed more eosinophilic cells in between the heart muscle fibers, with local disruption and focal necrosis of some fibres. Paracentrally, there was an area of the greatest concentration of eosinophilic granulocytes and foreign body type giant cells. These were seen surrounding partly resorbed ascarid larvae. Fragments of helminth cuticle were clearly evident within the mixed cellular and worm debris (Fig. 2). The diameter of the parasites' cross-sections ranged from 60 μm to 70 μm.

The large size of the larval remnants excluded consideration for larval Toxocara canis infection, which is the prototype of human visceral larva migrans [*1*], in favor of the larger larva of the raccoon ascaris Baylisascaris procyonis [*2,3*].

Neither larvae nor granulomata were seen in any of the other organs or tissues examined. The brain was essentially normal.

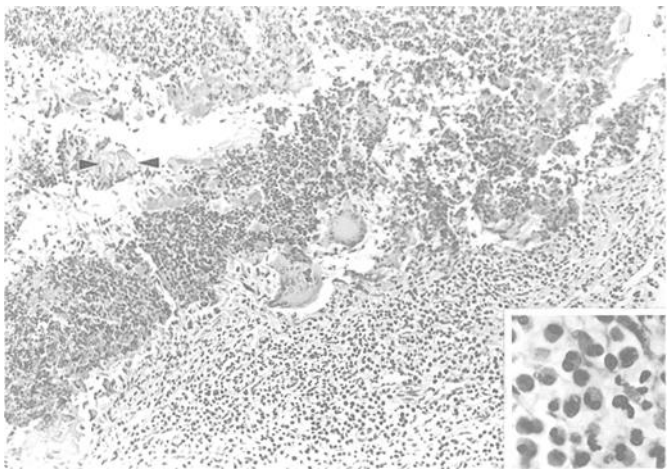

FIG. 2—*Low power microscopic view of the pseudotumor showing the marked eosinophilic infiltrate surrounding the necrotic worm and cellular debris. The residual poorly preserved worm cuticle is shown by* arrows. *Multinucleated giant cells are present in the center of the photograph. (Hematoxylin and eosin stain; magnification, ×70). The inset is a high power view showing the eosinophilic granulocytes, which comprise the host cellular component of the pseudotumor (inset magnification, ×400).*

The liver, which was somewhat enlarged (weighing 1050 g, as compared to an average of 850 grams for age), showed the histopathologic picture of mild chronic active hepatitis, consistent with Hepatitis C viral etiology.

Postmortem serum and blood were sent to a specialty laboratory for testing; however, the serological studies could not be performed due to sample deterioration.

Discussion

Larvae of the common raccoon round worm Baylisascaris procyonis are known to cause visceral larva migrans in animal and human paratenic hosts. Paratenesis is a concept that is important as it attempts to incorporate the biological relationships between the parasite and the host, and the extent of maturation of a larval parasite in a suboptimal host. The understanding of the visceral larva migrans syndrome as a host illness caused by a "frustrated ill-fated worm" is an over simplification. It entirely ignores the concept of transport hosts, which may be intermediate hosts, in which maturation of the helminth does not occur beyond a certain point; but, which may contribute to the successful dispersal and maintaining of the parasite, if and when a suitable intermediate host or final host becomes available [*1*].

Humans are a somewhat special case in that humans are often a "dead end host" for parasites in a paratenic relationship, (since when a human dies either because of the parasitic infestation or due to other cause or illness, the human's remains are rarely eaten by suitable final hosts). Also, ascarids, generally, have relatively simple life cycles, and this factor additionally complicates the precise understanding of the concept of paratenesis in animal hosts.

Baylisascaris procyonis has been associated causally with very serious, sometimes fatal, neurologic disease in free-ranging and captive species of wildlife and also in domestic animals. Veterinary public health officials discourage the adoption of orphaned raccoons for varied reasons including the risk of visceral larva migrans [*4*]. Raccoon pets must be dewormed regularly before the worms mature and lay eggs.

The first recognized human infection by Baylisascaris procyonis occurred in 1980 and involved a 10-month-old boy who died of eosinophilic meningoencephalitis; larvae were found in granulomas in the brain, lungs, mesentery, and myocardium [*5*].

A second human case of fatal eosinophilic meningoencephalitis and visceral larva migrans caused by the raccoon ascarid occurred in 1984, and involved an 18-month-old boy with Down Syndrome. Pathological findings in this second case showed multiple whitish nodules on the pleural surfaces, myocardium, epicardium, and were abundant in the mesentery. The major findings were in the brain with numerous larvae in the periventricular white matter, both with and without inflammatory reaction in the affected foci [*3*].

Baylisascaris larvae are considerably larger than the larvae of the two more common Toxocara species, T. canis and T. cati. Baylisascaris procyonis larvae measure 1.5–2.0 mm in length and have an average diameter of 60–75 μm; the corresponding measurements for Toxocara larvae are 0.4 mm and 12–20 μm [*2,3,5*].

There is also a strong suggestion that this parasite may be the cause of human "diffuse unilateral subacute neuroretinitis," comparable to experimentally produced ocular larva migrans in subhuman primates [*2*].

Interestingly, in neither previous case were large eosinophilic pseudotumors described although eosinophils did comprise a significant portion of the inflammatory infiltrate. Perhaps, this is an age related immune phenomenon, which is absent in infants and

very young children, or may be a manifestation of severe atopy or some other as yet unknown parasite-host immunological factor.

Transmission of the infection to the paratenic host is by ingestion of embryonated eggs, which seasonally abound in raccoon scats. The prevalence of Baylisascaris procyonis infection in raccoons is highest during the months of September, October, and November [6].

Raccoons are habitat generalists, and not surprisingly, there was little difference in the percentage of infected raccoon scats collected from urban and rural areas [6].

Geophagia (an important risk factor in the transmission of visceral larva migrans) in young children is known to be common, while persistence of this activity is seen in older children with the hyperkinetic syndrome.

The heart may be a target organ for various helminthiasis, some of these are echinococciasis, schistosomiasis, heterophyiasis, filariasis, ascaridiasis, trichinosis, and toxocariasis [7–9].

Pseudotumors caused by parasites are well known in the animal kingdom, for example, in fishes [10]. They may occur in man [11], and in experimental infections in New World monkeys [12]. The types and stages of the parasites inducing the pseudotumors, as well as the host cell composition of these masses varies greatly [10–12].

Pathologists and medical examiners must be prepared to encounter unusual cases of sudden death caused by parasites, some of which may be usual parasites of humans, others in which humans may act as a potential intermediate host, and finally those, in which humans serve as a paratenic host. If eye symptomatology is clinically documented, removal and study of the eyes at postmortem examination is mandatory to document possible neuro-ocular larva migrans.

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Address requests for reprints or additional information to
Dr. John Kasznica
Mallory Institute of Pathology
784 Massachusetts Avenue
Boston, MA 02118