

A Man With Unilateral Ocular Pain and Blindness

(See page 469–70 for the Answer to the Photo Quiz.)

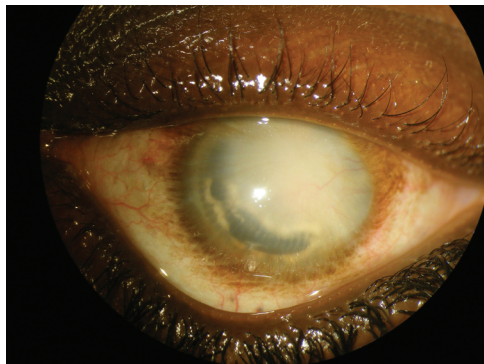


Figure 1. Slit lamp exam revealed the silhouette of a foreign body in the angle of the anterior chamber.

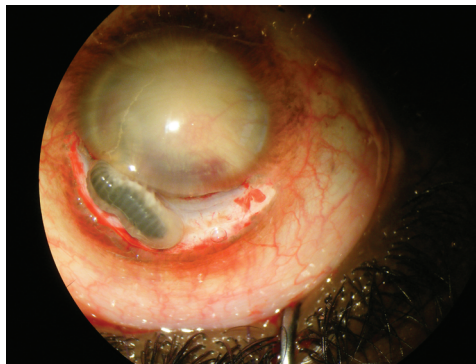


Figure 2. The parasite is shown crawling out of the eye following the incision.

A 25-year-old black man with unknown medical history presented at our ophthalmologic mobile outpatient clinic (District of Sankuru, East Kasai Province, Democratic Republic of Congo) with blindness and pain in his left eye. The examination showed a shrunken, nonfunctional left eye (phthisis bulbi), nonreactive to light, which, by slit lamp exam, revealed a large, blackish, crescent-shaped, worm-like foreign body wedged into the angle of the anterior chamber (Figure 1). Much to our surprise, the foreign body began a peristaltic motion upon physical stimulation of the eye. The patient was otherwise in good health and free of general symptoms. Physical findings were unremarkable; thus, no further diagnostic

tests were performed. The parasite was removed under local anesthesia, still alive and crawling (Figure 2). It was sent to the Hungarian Natural History Museum, Budapest, for further identification. The patient often consumed poorly cooked snakes. Despite the surgical procedure, the patient permanently lost vision in the left eye. What is your diagnosis?

Clinical Infectious Diseases 2013;57(3):418

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DOI: 10.1093/cid/cit309

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Diagnosis: Ocular pentastomiasis.

The parasite was identified as the larval form of an *Armillifer* species (Figure 1), a pentastomid. Brownish pigment in the gut is from consumed hemoglobin. It is clearly recognizable that the larva is in the middle of molting (parts of the molted “skin” have been torn off during handling of the animal). Also well visible is the mouth of the larva, surrounded by the 2 pairs of claws on each side of the mouth (arrows) used for attachment. In the first descriptions, these were also thought to function as separate mouths, hence the term Pentastomida, meaning “5 mouths.” The exact phylogenic position of these ancient parasites has long been debated until recent genetic evidences showed unequivocally that pentastomids are crustaceans [1].

The adult forms of *Armillifer* species live in the respiratory tract and paranasal sinuses of tropical reptiles. Human cases are almost exclusively caused by *Armillifer armillatus*. Of note, *Linguatula serrata* is a related pentastomid species that lives in the nasopharynx of temperate climate mammals [2]. Ingestion of the eggs with the nasal secretion of the definitive host results in visceral invasion of larval Pentastomida in the intermediate host (rat, sheep, goat, camel).

Pentastomid species can infect humans as either accidental definitive or accidental intermediate hosts. Ingesting undercooked viscera (liver, lungs, spleen) of the intermediate hosts may result in nasopharyngeal infestation called *halzoun* or *marrara* [3, 4], an illness caused by the adult form of *L. serrata* infecting the human paranasal sinuses where it feeds on blood and nasal secretions.

More commonly, in visceral pentastomiasis, humans serve as a dead-end intermediate host for the larvae. In most cases, it is caused by *Armillifer* species (eg, in our case). The ingested eggs hatch in the intestine; the larvae then penetrate the gut wall and migrate to parenchymal organs, surfaces of serous membranes, and soft tissues where they begin to molt and grow. They cause largely asymptomatic infestation of the liver, peritoneum, and lungs. The larvae usually die and calcify, leaving parts of their chitinous exoskeleton surrounded by a granuloma infiltrated with eosinophils. Larval pentastomiasis is usually a harmless condition accidentally found at autopsies. It is a rarity in developed countries, but still occurs in some parts of the world, for example, in Central Africa or in Malaysia [5, 6]. Infection of the



Figure 1. The molting larva of an *Armillifer* species. Note the 2 pairs of chitinous claws (arrows) around the mouth ($\times 10$ magnification).

eye is extremely rare. However, over a 3-year period, our ophthalmological examinations of 3000 patients in the Democratic Republic of Congo found 2 additional cases with macroscopically identical parasites, one of which was situated under the retina next to the papilla and the other one in the vitreous body, between a detached retina and the lens. Extended history revealed that all 3 patients came from the same region, where local eating habits include the consumption of various snakes, often raw. Due to the lack of any controlled data, the treatment of pentastomiasis is unclear, but surgical approach seems to be preferable in case of ocular localization. Differential diagnosis of ocular pentastomiasis includes myiasis and ocular larva migrans.

Notes

Acknowledgments. We are indebted to Zsolt Durkó for transporting the photographs.

Potential conflicts of interest. All authors: No reported conflicts.

All authors have submitted the ICMJE Form for Disclosure of Potential Conflicts of Interest. Conflicts that the editors consider relevant to the content of the manuscript have been disclosed.

Richárd Hardi,¹ Mihály Sulyok,² Lajos Rózsa,⁴ and Imre Bodo³

¹St Raphael Ophthalmological Center of Mbuji Mai, Democratic Republic of Congo; Departments of ²Infectious and Tropical Diseases, and ³Hematology and Stem Cell Transplantation, St István & St László Hospital, and ⁴Hungarian Academy of

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Correspondence: Mihály Sulyok, MD, Department of Infectious and Tropical Diseases, St István & St László Hospital, 5-7 Gyáli str, 1097, Budapest, Hungary (sulyok.mihaly@gmail.com).

Clinical Infectious Diseases 2013;57(3):469–70

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DOI: 10.1093/cid/cit316