A Man With Unilateral Ocular Pain and Blindness
(See page 418 for the Photo Quiz.)

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 phylogenetic 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 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

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