NEOTROPICAL ECHINOCOCCOSIS IN SURINAME: ECHINOCOCCUS Oligarthrus IN THE ORBIT AND ECHINOCOCCUS Vogeli IN THE ABDOMEN

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Abstract. This paper reports two cases of neotropical echinococcosis caused by Echinococcus oligarthrus and E. vogeli, neither of which has been reported from Suriname. Case 1, a six-year-old boy, presented a 15 × 25 mm retroocular cystic tumor (observed by ultrasound, computed tomography scan, and magnetic resonance imaging) causing exophthalmia, chemosis, palpebral ptosis, and blindness of the left eye. Of two tentative diagnoses, Echinococcus cyst or dermoid tumor, the former was shown to be correct at surgery when a clear liquid and detached protoscoleces were aspirated. Rostellar hooks of the protoscolex were characteristic of E. oligarthrus. Case 2, a 41-year-old man, had polycystic masses excised from the liver and abdomen. A presurgery diagnosis of E. vogeli infection was made due to calcifications seen in the lesions, positive serology, residence of the patient in the tropical forest, and later by the size and shape of rostellar hooks. The presence of these two parasites in one of the former Guianas is not surprising; both species are endemic in tropical forest in Central and South America wherever people have not exterminated wild canids, especially the bush dog, Speothos venaticus, and felids (wild cats of several species), along with pacas, agoutis, and other rodents that serve as intermediate hosts of these two cestodes. Eighty-six cases of polycystic echinococcosis are known in people from 11 countries from Nicaragua to Argentina: 32 due to E. vogeli, three to E. oligarthrus, and 51 for which determination of the species was not possible because the hooks of the protoscolex were not found or described. Research to elucidate aspects of transmission of E. vogeli and E. oligarthrus is of practical importance for defining measures for preventing the severe and frequently fatal illnesses caused by these two cestodes.

Humans may become accidental intermediate hosts of the larval stage of each of the four species of the genus Echinococcus. Echinococcus granulosus is the most common and cosmopolitan, and E. multilocularis is present only in the holarctic region and never has been found in Central or South America. The other two species, E. vogeli and E. oligarthrus, have so far been found in the tropical forest of Central and South America. Their life cycles depend on the predator-prey relationship existing between the final and intermediate hosts: for E. vogeli, between the wild dog Speothos venaticus and the paca Cuniculus paca; for E. oligarthrus, between wild cats and several species of rodents, especially agoutis Dasyprocta spp.). The larval stage of these two cestodes seems to be typically polycystic. This paper reports the occurrence of a larval cestode in the orbit of a child, representing the third proven infection by E. oligarthrus, and a case of polycystic echinococcosis of the abdomen caused by E. vogeli in an adult. Neither of these two parasites have been previously reported from Suriname or from French Guiana, where these cases were clinically studied.1–3

CASE REPORTS

Case 1. Infection with E. oligarthrus. A six-year-old boy, born in Suriname but presently a refugee in French Guiana, was treated for an eye condition at a St. Laurent du Maroni health center. After three weeks of conjunctivitis, he was studied at Hôpital Andre Bouton in St. Laurent du Maroni; at this time he showed an unilateral exophthalmos, blepharoptosis, and poor light perception-vision of the left eye. An ultrasound examination showed a cystic, ovoid structure, 15 × 25 mm, at the posterior-temporal portion of the orbit. The preliminary differential diagnoses were a cyst, a hematoma, or a cold abscess. He returned to Suriname, where the lesion was considered to be a hemangioma and surgery was not recommended. Because the lesion worsened, a year later, the patient returned to Cayenne, French Guiana and was referred to the Hôtel Dieu Hospital in Paris, France for further diagnosis and treatment.

The patient presented with lesions in the left eye: irredible exophthalmos, extensive chemosis, only light-perception vision, mydriasis, and a pale optic nerve head (Figure 1). Ultrasound, computed tomography (CT) scan, and magnetic resonance imaging (MRI, not available for inclusion) showed a large, liquid-filled cyst, 27 × 32 mm, surrounded by a 3 mm-thick, fibrotic capsule at the apex of the orbit. The optic nerve was pushed toward the inner orbital wall (Figure 2). An indirect hemagglutination test (IHT) result for echinococcosis was negative and the white blood cell and differential counts were normal.

Surgery was carried out at the Lariboisiere Hospital in Paris, France using a fronto-temporal incision. No periorbital fat tissue was seen. A fibrous capsule, observed through the damaged ocular motor muscles, was opened and 10 ml of clear liquid was aspirated from the cyst. Because echinococcosis was a possibility, hypertonic saline solution was injected, and was replaced after 5 min by isotonic fluid. Only a small portion of the cyst wall could be removed because the capsule was firmly attached to the orbital tissues.

Due to the risks to the eye that might accompany the complete removal of the cyst, the surgeon opted to prescribe treatment post-surgery with albendazole. The post-operative follow-up was uneventful. The severe proptosis and che-
mosis were corrected, but a slight blepharoptosis remained, as well as blindness of the left eye. A CT scan did not demonstrate additional cystic lesions in other organs. A parasitologic diagnosis of echinococcosis was made and treatment with albendazole (8 mg/kg/day for 15 days each for three consecutive months) was initiated before the child returned to French Guiana. A year later it was learned that the patient was in good health and that the light perception of the eye had apparently improved.

A squash preparation of the few protoscoleces obtained from the cyst fluid showed *Echinococcus* hooks and several calcareous corpuscles. It is now recognized that the measurements and shape of the hooks are consistent with those of *E. oligarthrus*. The mean length (range) of 30 hooks were 31.5 μm (28–37 μm). The hooks had a rather straight back and a guard located at about their center; the handle of the hook comprised 39% of its total length (Figure 3). The tissue sections (periodic acid–Schiff stained) showed a thin, dead, laminated membrane of *Echinococcus*.

**Case 2. Infection with *E. vogeli***. A 41-year-old man, born in Suriname, was studied radiologically because of the recent appearance of an abdominal mass. A CT scan and ultrasound demonstrated lesions in the liver and the abdominal cavity. Because the studies carried out to establish the etiology were inconclusive, the patient traveled to France to be assessed further.

Due to the cystic aspect of the tumors and the presence of calcification in some of the masses, the diagnosis of polycystic echinococcosis was suggested. The IHT titer (1:640) and the immunoelectrophoresis arc were positive for echinococcosis. At surgery, polycystic lesions were excised from liver, diaphragm, abdominal wall, mesentery, omentum, and pouch of Douglas. Two partial hepatectomies and an ileum-colectomy were required to eliminate all visible cysts. The patient left the hospital 10 days later and the post-operative follow-up was uneventful. No remaining cysts were observed in a post-surgical CT scan. Treatment with albendazole (800 mg/day) was prescribed. The patient did not present signs of recurrence of the cyst three years later.

Pathologic study demonstrated larval stages of *Echinococcus*, probably *E. vogeli*, based on the clinical and surgical findings. The laminated membrane was folded and due to the size and shape of the rostellar hooks of the protoscoleces, it is now recognized that the larval cestode was *E. vogeli* (Figure 4).

**DISCUSSION**

*Echinococcus granulosus* is the most important and frequent etiologic agent of echinococcosis in the world. In the Americas, this species in endemic in the countries of the southern cone, mainly present in domestic settings of sheep-raising areas. The life cycle involves the domestic dog and domestic ungulates. In the rest of the Americas, infection is rare and is observed mostly in immigrants from endemic areas. However, infections in dogs and ungulates and few human autochthonous infections are also known to occur.
The former three Guianas are vast areas of tropical forest crossed by several rivers, regions not suitable for raising sheep. There is no record of *E. granulosus* infections in domestic animals.

Although in areas endemic for *E. granulosus* the orbital location of cystic echinococcosis is rare (less than 500 cases have been reported), the clinical presentation of our case with orbital involvement was similar to other reported patients infected with *E. granulosus*. *Echinococcus granulosus* (and perhaps *E. oligarthrus*) is the most common agent of orbital cysts in areas of echinococcosis distribution. In areas where echinococcosis is absent, dermoid tumors are the most frequent etiology of orbital cysts, particularly in children.4–6

Due to the increasing global travel of patients looking for specialized treatment, or to the arrival of transient or refugee populations, it is important to differentiate the etiology of cystic orbital tumors. Fortunately, the use of modern imaging techniques has improved the diagnosis of cystic lesions. Our patient demonstrated an unilateral cystic structure by ultrasound testing, and the CT scan showed the typical round, well-circumscribed lesion with a sharply defined, hyperdense border. The orbital MRI also confirmed that the cyst contained fluid that was equal in density to that of water or cerebrospinal fluid. On the other hand, orbital dermoid tumors contain a more heterogeneous liquid, frequently including fat, which is more easily detected by either CT or MRI scanning.5,6 The treatment for dermoid tumors is surgical removal.

As observed in the present case, the cysts were unilateral and usually fertile, vision was impaired, and the serologic test results for echinococcosis were negative. It was the clear appearance of the cystic fluid aspirated during surgery that confirmed the diagnosis of echinococcosis; the size and shape of the rostellar hooks of the protoscolex confirmed the diagnosis of *E. oligarthrus*, rather than of *E. granulosus* infection. The sectioned portion of the cyst wall showed only a small fragment of a thin laminar membrane of a dead *Echinococcus*. Thin, nonconvoluted, laminated membrane and a thick germline membranes are characteristic of *E. oligarthrus* in animals but it is not known if the same occurs in human infections.7–11

Interestingly, the first human infection with *E. oligarthrus*, reported from a Venezuelan child, also involved a single cyst in the orbit;12 the second case, a Brazilian man, had two *E. oligarthrus* cysts in the heart.10 Cysts of *E. oligarthrus* in the animal host are often extrahepatic and single, and may be septated. The lesions are in the musculature of the extremities, and in the psoas and diaphragm. On the other hand, those in the spleen, liver, or lungs are frequently polycystic.7,13 In contrast, larvae of *E. vogeli* are mostly polycystic and located in the liver of the paca;2 in humans, the liver is involved in 78% and the lung in 14% of the cases. Other organs (but not the orbit), have also been affected.11

The second case in this report is a good example of an abdominal-visceral presentation of a polycystic echinococcosis due to *E. vogeli*.11 Whether *E. oligarthrus* may produce similar lesions is not known at the present time.

Except for an *E. granulosus* cyst seen in a Peruvian immigrant (Fournie B, unpublished data), and the two cases presented in this report, there is no record of echinococcosis in humans or animals in the former three Guianas. However, the presence in Suriname of the two species of neotropical *Echinococcus* is not unexpected. It has become clear that the polycystic echinococcosis present in humans and wild animals in Central and South America is caused by the metacestodes of *E. vogeli* or *E. oligarthrus*. Their presence should be expected in human populations living in tropical forested areas of the New World, in regions where people have not exterminated wild canids and felids, along with rodents that serve as intermediate hosts of these two cestodes.

We now know of at least 86 cases of human polycystic echinococcosis from 11 countries (Nicaragua, Costa Rica, Panama, Colombia, Ecuador, Venezuela, Brazil, Suriname, Uruguay, Argentina, and Chile), a remarkable increase since 1979 (12 cases in four countries). *Echinococcus vogeli* has been found more frequently than *E. oligarthrus*; of the 86 cases, 32 were due to *E. vogeli*, three to *E. oligarthrus*, and in 51 the species could not be determined because rostellar hooks of the protoscolecites were not described or found.11

A survey of Colombian wild animals demonstrated that among 325 pacas (*Cuniculus paca*), 30% were infected with *E. vogeli*.14 On the other hand, *E. oligarthrus* was found in 1% of the 325 pacas, in less than 1% of 1,168 spiny rats (*Proechimys spp.*), and in none of 118 agoutis (*Dasyprocta spp.*).15 Hunters, however, provided an agouti heart infected with this cestode. Infected agoutis have also been found in Panama, Brazil, and Venezuela.11–14 Another nominal species of *Echinococcus*, *E. crazi* Brumpt and Joyeux, 1924, described in an agouti in Brazil, has been found to be synonymous with *E. oligarthrus*.15

Transmission of *E. vogeli* to humans is probably attributable to the ingestion of eggs eliminated in the feces of domestic dogs or possibly of bush dogs (*S. venaticus*), although people rarely have contact with them. *Echinococcus vogeli* does not develop in cats. Transmission of *E. oligarthrus* is due to the ingestion of eggs eliminated with the feces of wild cats or domestic cats (but not dogs). Wild bush dogs and *Felis* spp. can infect humans when kept as pets or in zoos.11,16

The documented cats naturally infected with *E. oligarthrus* are the yaguarundi (*Felis yagouaroundi*), the puma (*F.
concolor), the jaguar (F. onca), the ocelot (F. pardalis), the pampa cat (F. colocolo), and the Geoffroyi cat (F. geoffroyi). Their distribution is generally wider than that of the bush dog, which extends from Panama to the tropical forested areas of Peru, Bolivia, Paraguay, and Brazil. For this reason it has been suggested that the polycystic echinococcosis reports from humans outside these countries (Nicaragua, Costa Rica, Chile, Argentina and Uruguay) may have been due to *E. oligarthrus* rather than to *E. vogeli*. However, another factor may be the existence of other canine (other than bush dog) final hosts in these regions. That possibility is being investigated.

Domestic dogs are probably unable to capture pacas, but they are infected when fed the viscera of infected pacas at a hunting site or, apparently less often, when the hunter returns home. It is also possible that domestic and small wild cats may be infected with *E. oligarthrus* during hunting and ingestion of small infected rodents. This may be the way the small pampa and Geoffroyi wild cats are infected with *E. oligarthrus* in Argentina. The small rodents of the genera *Microcavia* and *Octodon* were reported to be infected with *E. granulosus* before *E. oligarthrus* had been found in southern Argentina.

Research to elucidate aspects of transmission of *E. vogeli* and *E. oligarthrus* are of practical importance for suggesting control measures in the severe and frequently fatal illnesses caused by them.

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REFERENCES