

CASE REPORT

LAGOCHILASCARIASIS LEADING TO SEVERE INVOLVEMENT OF OCULAR GLOBES, EARS AND MENINGES

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SUMMARY

A case report of a 31 year-old woman from Paraíba State (North-Eastern Brazil) that presented severe involvement of ocular globes, ears and meninges. Diagnosis was established after enucleation of her left eye, when adult worms were seen in the midst of a granulomatous inflammatory process. Her response to the initial treatment with levamisole and cambendazole was good, but there was a relapse after the fifth month of treatment even with maintenance doses of both medications. She later received ivermectin and albendazol and responded well.

KEYWORDS: *Lagochilascaris minor*; Lagochilascariasis; Ocular involvement.

INTRODUCTION

Lagochilascariasis is a quite rare human infectious disease, caused by a nematode described almost a century ago called *Lagochilascaris minor* Leiper, 1909. It is an emergent helminthiasis, often chronic and sometimes fatal^{8,13,17}. There are about 110 reported cases in the literature^{9,15} but only one, published in 1926, described an ocular globe involvement similar to this case¹⁶. In recent years, however, there has been an increase in the number of cases, but many of them were neither notified nor published. Most of the cases occurred in tropical regions of Central and South America. Brazil is the leading country in the world, accounting for 75% of reported cases in humans, mainly in the Amazon region^{6,14,21}, but there are cases in other parts of the country^{6,9,14,16,20}. Cases have been reported in Venezuela^{13,23}, Colombia^{4,8}, Mexico¹⁹, Bolívia¹⁰, Suriname^{11,12}, Trinidad Tobago¹⁶ and Costa Rica^{12,14}. The infection is prevalent in individuals from lower social classes, particularly in rural areas. Many infected individuals live in small deforested areas, where people eat game^{14,15}, like armadillo and guinea-pig, which has been pointed out experimentally as intermediate or paratenic host of *Lagochilascaris minor*¹⁴.

CASE REPORT

A 31 year-old Caucasian woman, who lives in Paraíba State in North-Eastern Brazil, came to São Paulo one year ago, looking for medical care for her ears and eyes affection.

Three years ago she presented with otalgia and otorrhagia in the left ear, followed by partial deafness. She was then submitted to a surgical

procedure, of which not much is known. A month later, she had itching and a purulent discharge in her left eye, leading to an inflammatory growth and deformity of the orbit and blindness. Biopsy specimens showed a nonspecific granulomatous process. Four months ago a similar involvement occurred in her right eye, and two months ago partial deafness was observed in her right ear. During this period, she also presented hoarseness, weight loss (15 kg in one-year period), headaches, nausea and vomiting.

She had diabetes diagnosed a year ago, and she has been taking metformin 500 mg a day. She had no other diseases, did not smoke or drink alcohol, denied having travelled anywhere else, and had a dog living in the house. She lives in a brick house, with drinkable water and sewage in a small town in a rural area, 500 m away from the forest. Alimentary habits included game, such as armadillos and guinea pigs.

Apart from hypertension and diabetes in her father's family, there were no other family diseases, and nobody else showed the above mentioned symptoms.

Cardiovascular and abdominal examination resulted unremarkable. Neck examination showed some indurations of the cervical region, but no nodes were depicted. The left eye was frozen and proptotic (Fig. 1). Ocular motility was normal in the right eye. Although there was some visual acuity in the right eye, there were no light reflexes, neither efferent nor afferent in both eyes. A slit lamp examination revealed no pupil reaction in either eye and stromal infiltrates in the inferior half of the left cornea. Intraocular pressure was 15 mmHg in both eyes and funduscopy revealed a prominent posterior pole and pale optic disc, mainly in the

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Fig. 1 - Paralytic midriasis in both eyes and superior temporal left eye proptosis.

left eye. Laboratory findings showed a mild ferropenic anemia, no eosinophilia, and mild hyperglycemia. Imaging findings are shown in Fig. 2 and 3. Diagnosis was only established after enucleation of her left eye, when the adult worms of *Lagochilascaris minor* were seen on histology in the midst of the granulomatous process (Fig. 4).

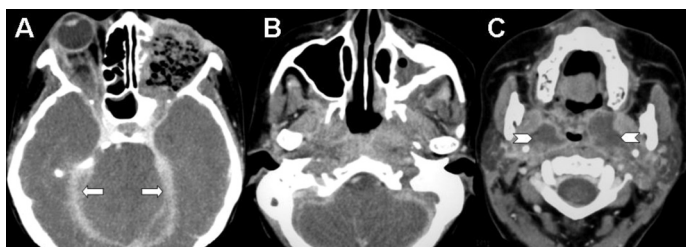


Fig. 2 - Axial post contrast CT images. (A) Inflammatory involvement of the orbits and left ethmoidal cavities (the left globe has been enucleated), as well as the marked infratentorial pachymeningeal thickening (arrows). (B) Chronic maxillary sinus involvement associated with bone thickening. There is also thickening of the nasopharynx walls. (C) Extensive fluid collections in the deep cervical compartments, mainly in parapharyngeal spaces (arrowheads).

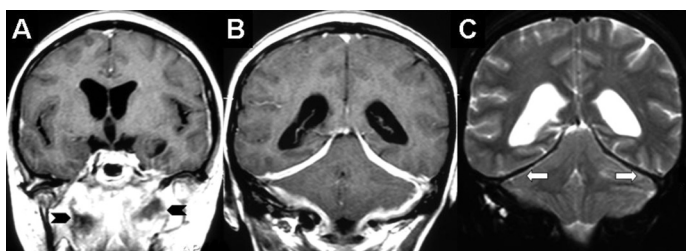


Fig. 3 - Coronal MR sequences. (A and B) Postcontrast T1-weighted images show bilateral fluid collections in the deep cervical spaces (arrowheads in A) and marked infratentorial pachymeningeal thickening (B). (C) Note the marked hypo intensity of the pachymeningeal thickening on T2-weighted image (arrows). There is also a ventricular enlargement depicted in all sequences.

She was treated with levamisole 150 mg/d every other day for six days for a total of three doses, followed by cambendazole 30 mg/Kg/d for five days. Her general condition slowly improved. Elimination of parasites was not observed.

Additional procedures included bilateral mastoid drainage, hearing devices and tarsorrhaphy of the right eye, which was necessary due to

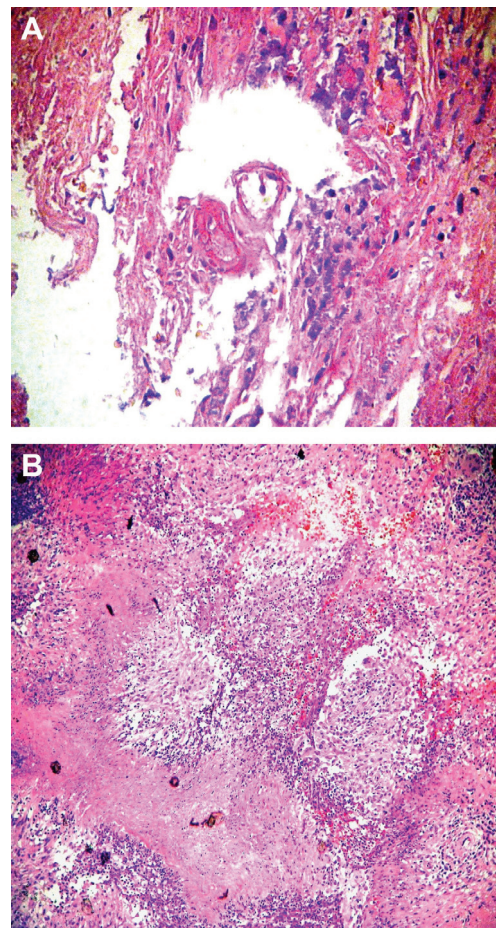


Fig. 4 - Light photomicrograph of hematoxylin and eosin-stained sections of the left orbit, showing in A: necrosis and fragments of adult *Lagochilascaris minor*; in B: necrosis, epithelioid granulomata and calcified *L. minor* eggs. (Original magnification 400 x).

conjunctival exposure and progressive proptosis. Doses of levamisole and cambendazole were given monthly, but on the fifth month her general condition and her right eye worsened. A purulent discharge was observed in the left orbit. The analysis of this material showed some third-stage (L3) larvae of *Lagochilascaris minor*. This finding strongly suggests the persistence of living adult worms shedding viable eggs, in spite of specific treatment, justifying the introduction of ivermectin. It was used in a dose of 300 µg/kg/weekly for 21 weeks, associated to albendazole (three cycles with 400 mg/d for 30 d, with 15 d intervals). Again, her general status slowly improved.

Currently, her general condition is good and she put on weight. We started a maintenance dose of ivermectin of 300 µg/kg every two weeks.

DISCUSSION

Infection with *Lagochilascaris minor* occurs mainly after the ingestion of third stage larvae encysted in different tissues of wild animals^{5,14,22}. The larvae get rid of the cysts in the host stomach, cross the digestive wall, and by an unknown tropism mechanism, migrate

to the commonly affected regions of head, neck, and thorax^{5,18,22}. The worms have a particular tropism for the cervical region, including oro- and nasopharynx, mastoid, and middle ear. They have an impressive migrating capacity through host tissues. Its pathways may become real tunnels. Proteolytic enzymes can facilitate invasion, migration, evasion from immune response, nutrition and parasite development in the host¹. A chronic granulomatous disease progressively develops in these regions^{15,18}. Osteolysis is a prominent feature that enables the parasite to migrate further. Intermittent elimination of the parasite through lesion orifices, ears, mouth, and nose is frequent, and in most cases is the main clue for the diagnosis^{15,21}. However, its absence must not rule out the possibility of lagochilascariasis, as seen in this case. Involvement of the central nervous system was reported in some cases^{17,20} but not the chronic and progressive orbital damage seen in this case. Weight loss, cellular and humoral immunosuppression can be observed. Diagnosis can be made if one can find eggs, larvae or adult worms in the lesion discharge, or by histopathological analysis of the affected tissue, which may reveal a granulomatous "foreign-body type" reaction, containing giant cells with parasite elements. The blood count is nonspecific, and leucocytosis or leucopenia may be present, as well as eosinophilia or, paradoxically, anaesinophilia. Imaging studies including computed tomography (CT) and magnetic resonance (MR) scans have been of great value to assess the extension and follow-up of the infection⁷.

Many drugs have been used to treat lagochilascariasis, and there is no ideal treatment^{2,3}.

Good results reported in literature were achieved with cambendazole and levamisole, alone or in association, given in attack and maintenance doses. Levamisole should be used, according to age, in a dose of 80 or 150 mg/d every other day for six days for a total of three doses. It should be followed by the administration of cambendazole 20/mg/kg/d (30 mg/kg/d if central nervous system is involved), for five consecutive days, once a month, in a total of four series. Maintenance therapy is justified by the high frequency of recurrence, and must include a course of cambendazole and levamisole every six months. Diethylcarbamazine, mebendazole, albendazole, praziquantel and ivermectin have also proven to be effective in different regimens^{2,3}, especially the association of albendazole and ivermectin that we used in this case after the recurrence. The combination was well tolerated and so far there have been no signs of disease activity. In many cases the specific treatment causes many worms to migrate to the skin. Surgical drainage of the lesions is a valuable resource to promote healing.

RESUMO

Infecção humana por *Lagochilascaris minor* com envolvimento ocular, auditivo e das meninges

Paciente do sexo feminino, com 31 anos, procedente da Paraíba, apresentava envolvimento severo de ambos os globos oculares, com perda da visão à esquerda, comprometimento da audição e das meninges. Após enucleação do olho esquerdo fragmento de helminto identificado como *Lagochilascaris minor* foi observado em processo inflamatório granulomatoso. Inicialmente a paciente foi tratada com levamisol e cambendazol, com bom resultado. Verificou-se, todavia, piora do quadro após cinco meses, com eliminação de larvas do ascarídeo em lesão presente na órbita esquerda, embora a medicação fosse mantida

com administração periódica. Houve boa resposta terapêutica, com regressão do quadro, após substituição dos anti-helmínticos anteriores pela associação ivermectina e albendazol.

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Received: 16 September 2008

Accepted: 13 October 2008