Retrobulbar optic nerve cysticercosis

Case report

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The authors report a first case of intraoptic neurocysticercosis in a 12-year-old boy living on Reunion Island. Cysticercosis of the retrobulbar portion of the optic nerve is rare. Because of the patient’s age and disturbances in both visual acuity and visual field, it was initially believed to be an optic nerve tumor. Computerized tomography scans and surgical aspects were confirmed by pathological findings. A conservative removal using en bloc orbitotomy showed good functional and aesthetic results.

Key Words • cysticercosis • optic nerve tumor • orbitotomy • visual dysfunction • child

Cysticercosis is the most common parasitosis affecting the human nervous system. The disease is caused by infestation with the larval form (Cysticercus cellulosae) of the pork tapeworm (Taenia solium). It occurs when man becomes the intermediate host by ingesting Taenia eggs from contaminated food, water, or hands. Humans are the only known definitive hosts of the adult worm. The eggs hatch in the human intestinal tract, then oncospheres cross the intestinal wall, reach the circulatory system, and are spread into host tis-

Fig. 1. Chart showing Goldmann visual fields in a 12-year-old boy with intraoptic nerve cysticercosis and a very constricted left visual field.
sue, in which cysticerci develop. The main target organs of larvae are brain, skeletal muscles, heart, liver, subcutaneous tissue, and eyes. In the latter, cysticerci can lodge in the anterior chamber, vitreous, retina, and, rarely, exterior to the optic disc.1

An unusual location of cysticerci is the peripheral nervous system. Isolated cases of lumbar nerve root compression have been reported12 but there is no previous report of cranial nerve location except in a recent letter from Madan, et al.,9 in India.

Case Report

Examination. This 12-year-old boy living on the Pacific Reunion Island presented in September 1987 with a recent left-eye vision loss with no exophthalmos. There was no history of trauma or pain in the eye, headache, or vomiting. On initial ocular examination, uncorrected left eye acuity was 4/10 and right eye acuity was 10/10. The left Goldmann visual field was very constricted and the right was normal (Fig. 1). Left disc edema was confirmed by fluorescein test. Visually evoked potentials showed left disturbances as late prechiasmatic response (latency: 126 msec) and reduced amplitude. Heterogeneous enlargement of the left intraorbital optic nerve was detected on B-scan ultrasonography.

Because of the patient’s age and the frequency of the lesion, the initial diagnosis was of an optic nerve tumor, but general physical and systemic examinations were unrevealing. In particular, no café-au-lait spots or subcutaneous nodules were present.

Plain skull x-ray films of both orbits were normal. Routine laboratory evaluation was unrevealing.

Operation. A surgical approach for further exploration and histological diagnosis was finally chosen after anti-edema medication had been administered. A left frontotemporal incision made a one-piece frontal bone flap with a superior orbital roof possible via en bloc orbitotomy. Using a microscope, exposure of the orbital contents was achieved before optic nerve individualization. This nerve was found to have fusiform thickening 5 mm behind the optic globe with normal color aspect. Longitudinal incision of the vagina was made on the top of the bulge, then a punctiform neurotomy allowed a white gluey substance, very suggestive of cysticercosis, to leak out. Microdissection of the endonerve cystic cavity permitted the surgeon to see and remove the supposed tapeworm larva that was lodged between the nerve fascicles.

Postoperative Course. Histological findings confirmed the diagnosis of cysticercosis (Fig. 3). The postoperative course was uneventful and the patient’s vision recovered without nerve palsy. Praziquantel therapy was initiated as is usual.13 Upon questioning the family, we found that two sisters but not the patient had been treated for worms in the recent past. Pork from free-roaming pigs, which can sometimes be contaminated, was frequently consumed. It is the main risk factor for the disease in Reunion, which has endemic cysticercosis.10,13 However, no subcutaneous nodules could be palpated and there was no neurological defect. The cerebrospinal fluid (CSF) contained three lymphocytes per cubic millimeter, with normal chemistries. Immunological reactions to cysticercosis, both in blood and CSF, were negative according to enzyme-linked immunosorbent assay (ELISA). No pinhead-sized calcifi-

Axial computerized tomography (CT) of the head and orbit with contrast enhancement showed a spherical heterogeneous lesion inside the left retrobulbar optic nerve with spontaneous peripheral hyperdensity and noncontrast-enhancing central hypodensity (Fig. 2 left). Coronal views confirmed an intraorbital lesion with the retrobulbar segment thickened. The muscle cone and both optic foramina were preserved. No bone structure, parenchymal, or ventricular abnormalities were found. The right optic nerve was normal (Fig. 2 right).

FIG. 2. Left: Axial computerized tomography scan with contrast enhancement showing a spherical heterogeneous lesion inside the left retrobulbar optic nerve. Right: View in the coronal plane showing a noncontrast-enhancing central hypodensity.


FIG. 4. Postoperative computerized tomography scan showing the left retrobulbar optic nerve still enlarged but without cystic formation.

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cations of dead larvae were seen in the patient’s muscles on x-ray films of his hips and knees.

Six months postsurgery, CT showed a left retrobulbar optic nerve that was still thickened but without any cystic formation (Fig. 4). Uncorrected visual acuity in the left eye was 9/10, and the left Goldmann visual field had improved (Fig. 5). Five years later, the neuroophthalmological examination is still normal.

Discussion

To our knowledge there is only one recent case of intraoptic nerve cysticercosis described, although the optic disc is a well-known site of infection and figures in 3% to 5% of cases.\(^1\,^8\,^9\)

Clinical features of neurocysticercosis are so varied that recognition of a typical syndrome is not possible.\(^5\) The presenting symptoms and signs depend on the predominant location of the larvae. In most cases diagnosis can be confirmed with CT, magnetic resonance (MR) imaging, and CSF data.\(^2\,^6\,^10\) Unfortunately, MR imaging was not available in our case.

Some cases have an unusual clinical feature, CT or CSF findings that are nonspecific, and blood or CSF tests that are not always positive in the presence of the disease, as in our case.\(^11\) These cases are misdiagnosed until biopsy specimens are obtained for histological confirmation.\(^5\,^12\)

Intraoptic nerve cysticercosis is an example of such cases, because it presents in children like an optic nerve tumor such as glioma (retrobulbar pilocytic astrocytoma), cavernous angioma, hemangioblastoma, or meningioma.\(^17\) Neuroophthalmological signs of cysticercosis are not specific. Papilledema is the most common sign, with pupillary abnormalities and nystagmus.\(^8\) A prechiasmatic lesion can be confirmed with visually evoked potentials and neuroophthalmological examinations are very helpful to confirm beneficial treatment. General physical examination was unremarkable in this case probably because contamination was a recent and unique event. Children in endemic areas are at high risk because of poor personal hygiene, poor sanitation, and the presence of free-roaming pigs, yet there is often an extended interval between exposure and expression of disease.\(^4\,^10\,^13\)

Immunological reactions assessed by ELISA were normal in our case. This is already known to occur with neurocysticercosis.\(^6\) Computerized tomography can help with diagnosis by revealing intraoptic nerve hypodense cystic lesions with a peripheral spontaneous hyperdensity, which are not seen in other lesions primarily involving the optic nerve.\(^2\,^3\) However, cystic lesions can result from other nervous system parasitic diseases such as intraorbital hydatidosis or paragonimiasis.\(^11\)

A diagnosis of neurocysticercosis, given a contamination context in an endemic area and optic nerve lesion on morphological imaging, must be confirmed histologically. With regard to good visual function recovery, a conservative surgical treatment is indicated in such cases.

We hope that improvement in public health measures will decrease the incidence of cysticercosis in Reunion Island. Present knowledge can make this a reality.

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