

Extraocular Muscle Cysticercosis: Clinical Presentations and Outcome of Treatment

Kanwar Mohan, MS; Vandana Saroha, MS; Ashok Sharma, MS;
Surinder Pandav, MS; and Usha Singh, MS

ABSTRACT

Purpose: To report various clinical presentations and treatment outcomes in a series of patients with extraocular muscle cysticercosis.

Methods: This retrospective study reviewed the charts of 43 patients diagnosed with extraocular muscle cysticercosis with computed tomography and orbital B-scan ultrasonography between January 1991 and December 2002. Clinical presentation, results of investigations, treatment, and outcome were recorded.

Results: The superior rectus was the most commonly affected extraocular muscle. Restricted ocular motility was present in 88% of patients, and inflammatory signs were noted in the involved quadrant in 70% of patients. Eleven patients were treated with oral albendazole alone and 31 patients were treated with oral albenda-

zole and prednisolone. Four extraocular muscle cysts were excised surgically, and five extruded spontaneously. Inflammatory signs subsided in all patients, and residual restriction of ocular motility was seen in 16 (50%) of 32 patients at a mean follow-up of 5 months. Type of treatment made no significant difference in the ocular motility outcome.

Conclusions: Extraocular muscle cysticercosis should be considered in patients who present with restricted ocular motility and inflammatory signs. The direction of motility restriction does not indicate the muscle involved. Residual restriction of ocular motility is common despite the addition of corticosteroids to albendazole therapy.

J Pediatr Ophthalmol Strabismus 2005;42:28-33.

INTRODUCTION

Cysticercosis is infestation by *Cysticercus cellulosae*, the larval form of the cestode *Taenia solium*.

Drs. Mohan and Sharma are from the Squint Clinic, Grewal Eye Institute; and Drs. Saroha, Pandav, and Singh are from the Department of Ophthalmology, Postgraduate Institute of Medical Education and Research, Chandigarh, India.

Originally submitted August 26, 2003.

Accepted for publication January 20, 2004.

Address reprint requests to Kanwar Mohan, MS, Grewal Eye Institute, SCO: 168-169, Sector 9-C, Chandigarh - 160009, India.

This parasite is endemic in various parts of the world, including Africa, Mexico, Southeast Asia, Eastern Europe, Central and South America, and India.¹ Ocular cysticercosis can involve the anterior segment,² posterior segment,²⁻⁴ or adnexa.^{5,6} Infestation of the extraocular muscles by *C cellulosae* is rare. There are some isolated case reports⁷⁻¹¹ and a few small series of patients with extraocular muscle cysticercosis.¹²⁻¹⁴ Patients with extraocular muscle cysticercosis usually present with restricted ocular motility and recurrent inflammation.^{12,13} Several investigators^{7,12-14} have used oral albendazole in

Presentation	No. (%)
Restricted ocular motility	38 (88)
Inflammatory signs	30 (70)
Diplopia	23 (53)
Ptosis	15 (35)
Strabismus in the primary position	11 (26)
Proptosis	7 (16)

combination with corticosteroids to treat extraocular cysticercosis. To date, the clinical presentation and treatment results in a large series of patients with extraocular muscle cysticercosis have not been reported.

We report the clinical presentation and treatment outcomes for 43 patients with extraocular muscle cysticercosis.

MATERIALS AND METHODS

The medical records of 43 patients with extraocular muscle cysticercosis diagnosed at Grewal Eye Institute and Postgraduate Institute of Medical Education and Research, Chandigarh, India, between January 1991 and December 2002 were reviewed. Clinical presentation, the results of investigations, treatment, and outcome were recorded. The diagnosis of extraocular muscle cysticercosis was made by the observation of a cystic lesion with or without a hyperdense focus (scolex) within the affected muscle on computed tomography (CT) scanning (18 patients) and by the presence of a sonolucent area with well-defined margins with or without an echodense area (scolex) within the affected muscle on orbital B-scan ultrasonography (16 patients). Fusiform enlargement of the muscle without a cystic lesion was seen in four patients on CT scanning and in five patients on ultrasonography; none of these nine patients had any clinical sign of thyroid ophthalmopathy. In these nine patients, the diagnosis of extraocular muscle cysticercosis was presumptive, based on the knowledge that cysticercosis was endemic in our region and the clinical presentations were similar to those in other cases. Enzyme-linked immunosorbent assay (ELISA) for detection of anticysticercal antibodies was performed in nine patients.



Figure 1. Inflammatory signs in the eyelids and conjunctiva, and restricted abduction of the left eye in a patient with cysticercosis of the left medial rectus muscle.

Eleven patients were treated with oral albendazole (15 mg/kg body weight) alone for 4 weeks and 31 patients received oral albendazole and prednisolone (1.5 mg/kg body weight). One patient was not treated with any oral medication and underwent surgical removal of the cyst.

Thirty-two patients underwent follow-up for 2 to 24 months (mean, 5 months) and were evaluated to assess the outcome of treatment. At their most recent follow-up, six patients underwent CT scanning and eight patients underwent orbital B-scan ultrasonography to assess the status of the cyst and the condition of the muscle. Data were analyzed using the chi-square test.

RESULTS

There were 22 males and 21 females. Patient age ranged from 5 to 44 years (mean, 19 years). The right eye was involved in 25 (58%) patients and the left eye in 18 (42%) patients. All of the patients had a cyst within a single extraocular muscle. The muscles affected were the superior rectus in 16 patients, inferior rectus in 8 patients, lateral rectus in 8 patients, medial rectus in 7 patients, and superior oblique in 4 patients. The various clinical presentations are summarized in Table 1. Thirty-eight (88%) of 43 patients exhibited restricted ocular motility, which was present in the direction of the involved extraocular muscle in 10 patients, in the opposite direction of the involved extraocular muscle in 16 patients (Fig. 1), and in both directions in 12 patients (Fig. 2). Inflammatory signs consisting of pain and congestion in the involved quadrant of the eye were seen in 30 (70%) patients (Fig. 1). Twenty-three (53%) patients complained of diplopia in the direction of restricted movement.

Computed tomography scanning was per-

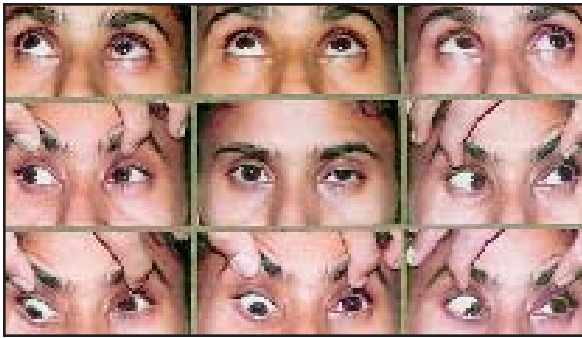


Figure 2. Ptosis and restricted elevation and depression of the left eye in a patient with cysticercosis of the left superior rectus muscle.

formed in 22 patients and showed a cystic lesion with a scolex in the affected extraocular muscle in 2 patients (Fig. 3A), a cystic lesion without a scolex in 16 patients (Figs. 3B-C), and a diffuse enlargement of the muscle but no cystic lesion in 4 patients (Fig. 3D). Orbital B-scan ultrasonography was performed in 21 patients and showed a cystic lesion with a scolex in the affected extraocular muscle in 6 patients (Fig. 4), a cystic lesion without a scolex in 10 patients, and a diffuse enlargement of the muscle but no cystic lesion in 5 patients.

Of the nine patients in whom ELISA was performed for detection of anticysticercal antibodies, a cyst was visible on CT scanning in seven and fusiform enlargement of the muscle without a cystic lesion (presumed extraocular muscle cysticercosis) was noted in two. ELISA was positive in only 2 (22%) patients, both of whom had a cyst visible on CT scanning. Of the 22 patients with CT scans, no patient manifested associated neurocysticercosis.

In 2 of the 11 patients who were treated with oral albendazole alone, the cyst spontaneously extruded 7 to 10 days after starting therapy; in 1 patient, the cyst migrated anteriorly and was removed surgically. In 3 of the 31 patients who received oral albendazole and prednisolone, the cyst spontaneously extruded 3 to 15 days after starting medication; 2 patients subsequently underwent surgical removal of the cyst. One patient who did not receive any oral medication underwent surgical removal of the cyst. Histopathologic examination of the four surgically removed cysts and two of the five spontaneously extruded cysts confirmed the diagnosis of cysticercosis (Fig. 5).

Table 2 presents treatment outcomes in 32 patients who underwent follow-up for a mean of 5

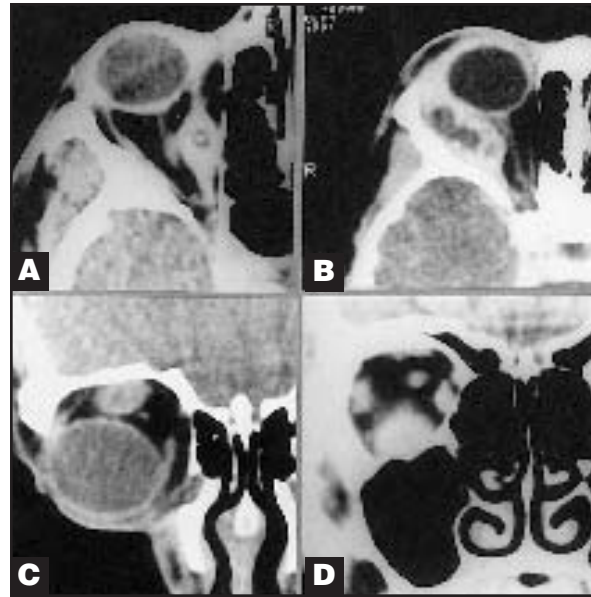


Figure 3. Computed tomography scans of patients with extraocular muscle cysticercosis. (A) Fusiform enlargement of the right medial rectus muscle and a cystic lesion with a hyperdense scolex. (B) Well-defined cyst without a scolex in the right lateral rectus muscle. (C) Cystic lesion without a scolex in the left superior rectus muscle. (D) Diffuse thickening of the left inferior rectus muscle.

months. Ocular and periorbital inflammation subsided in all 32 patients. Sixteen (50%) patients had residual restriction of ocular motility, and 8 (25%) patients had diplopia in the extreme position of the involved gaze. The effect of type of treatment on ocular motility outcome is presented in Table 3. The type of treatment made no statistically significant difference in the outcome of ocular motility ($P = .528$). Repeat CT scans in six patients and orbital B-scan ultrasonography in eight patients were normal at their most recent follow-up examination.

DISCUSSION

Cysticercus cellulosae is the larval form of the tapeworm *T solium* for which the pig is the intermediate host and the human is the definitive host. However, humans occasionally serve as the intermediate host by ingestion of pork, fecally contaminated vegetables or water, or autoinfection from reverse peristalsis. The eggs mature and the larvae penetrate the intestinal mucosa to enter the portal circulation and are carried to other organs where the cysts develop. Cysticercosis mainly affects the central nervous system, the skin, and the skeletal muscle.¹⁵

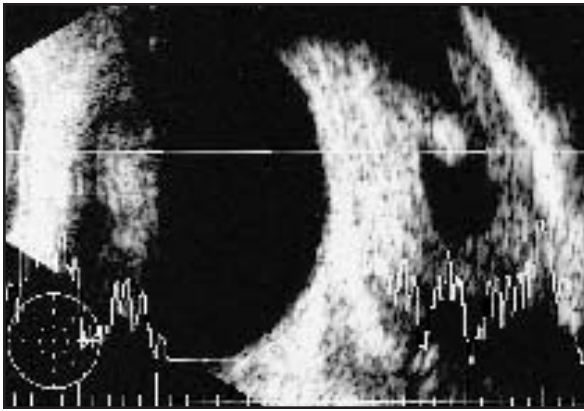


Figure 4. Orbital B-scan ultrasonogram showing a sonolucent cyst with an echodense scolex in the right medial rectus muscle in a patient with extraocular muscle cysticercosis.

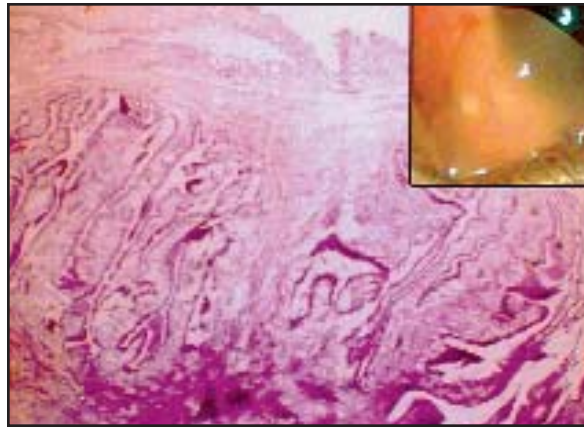


Figure 5. Photomicrograph of a cysticercus cyst (inset) shows the scolex (hematoxylin-eosin, original magnification $\times 4$).

Larvae may enter the eye through the choroidal circulation and migrate in the subretinal space or enter the vitreous. Ocular or adnexal involvement occurs in 13% to 46% of infected patients.¹⁶ Kruger-Leite et al.³ reviewed all documented cases of ocular and adnexal cysticercosis and found 35% of the cysts were in the subretinal space, 22% in the vitreous, 22% in the subconjunctival space, 5% in the anterior segment, and only 1% in the orbit.

Cysticercosis has been known to involve any of the extraocular muscles except the inferior oblique.¹²⁻¹⁴ In a few small series of patients with extraocular muscle cysticercosis, the most common muscle involved was the medial rectus^{12,14} or the inferior rectus.¹³ However, in our patients, the most common extraocular muscle involved was the superior rectus. This indicates that cysticercosis has no predilection for any particular extraocular muscle. We also did not have any patient with cysticercosis of the inferior oblique muscle.

Cysticercosis of the extraocular muscles presents with recurrent inflammation, restricted ocular motility, proptosis, and ptosis.¹² Restricted ocular motility may be secondary to a cyst within the muscle or inflammation of the extraocular muscles. A severe inflammatory reaction is believed to be caused by the liberation of toxins, particularly when the larva dies. Approximately 90% of our patients had restricted ocular motility and 70% had inflammatory signs in the involved quadrant of the orbit. Therefore, we suggest extraocular muscle cysticercosis should be considered in patients who present with restricted ocular motility and inflammatory signs. Ocular motility was restricted most common-

ly in the direction opposite to the involved muscle, as observed by Sekhar and Lemke.¹² Some patients exhibited restricted ocular motility in the direction of the involved muscle as well as in the opposite direction of the involved muscle. Therefore, the direction of restricted ocular motility does not indicate which muscle is involved and may even be misleading. Diplopia as a result of restricted motility is also a common presenting symptom.

Fusiform enlargement of the affected muscle and a well-defined cystic lesion with a scolex on either CT or ultrasonography are characteristic of cysticercosis.^{12,13,17-19} However, a scolex may not be visible in all patients, and a diagnosis of cysticercosis may be considered in such patients if other ocular signs are suggestive of cysticercosis. Even fusiform enlargement of the muscle without a cystic lesion should warrant suspicion of cysticercosis because a cystic configuration may be masked by diffuse thickening of the muscle and the cyst may become visible only after the inflammation has subsided, as observed by Sekhar and Lemke.¹² One also should consider the possibility of a thyroid ophthalmopathy in patients with diffuse enlargement of the inferior rectus muscle. We observed fusiform enlargement of the muscle but no cystic lesion in nine patients; none of these patients had any clinical sign of thyroid ophthalmopathy. Because cysticercosis is endemic in our region, we made a presumptive diagnosis of cysticercosis in these nine patients as their presentations were similar to those in other patients.

Enzyme-linked immunosorbent assay for detection of anticysticercal antibodies has been found to

TABLE 2
TREATMENT OUTCOME IN PATIENTS WITH
EXTRAOCULAR MUSCLE CYSTICERCOSIS AFTER
MEAN FOLLOW-UP OF 5 MONTHS (N = 32)

Clinical Sign	No. (%) Patients	
	Before Treatment	After Treatment
Restricted ocular motility	32 (100)	16 (50)
Inflammatory signs	25 (78)	0 (0)
Diplopia on eccentric gaze	22 (69)	8 (25)
Strabismus in the primary position	11 (34)	0 (0)
Ptosis	8 (24)	3 (9)
Proptosis	5 (15)	0 (0)

be positive in 61% to 79% of patients with neurocysticercosis²⁰ and in 8% to 20% of patients with extraocular muscle cysticercosis.^{12,13} ELISA was performed in 9 of our patients, but was positive in only 2 (22%) patients. As extraocular muscle cysticercosis is largely an isolated and relatively mild infection, it is possible circulating levels of antibodies are low, which may result in false-negative results. Therefore, a negative ELISA does not rule out the diagnosis.

Neurocysticercosis has been associated with both orbital¹² and intravitreal²¹ cysticercosis. The CT scan performed for extraocular muscle cysticercosis can show neurocysticercosis if present. None of our 22 patients with CT scans manifested associated neurocysticercosis. As we did not perform CT scanning in all of our patients and diagnosed extraocular muscle cysticercosis in almost half of our patients based on orbital ultrasonography only, it is possible neurocysticercosis might have been missed in some patients. Currently, we perform CT scanning in all patients with extraocular cysticercosis.

Extraocular muscle cysticercosis has been treated successfully with oral albendazole.^{7,12-14} The greater bioavailability of the drug due to better vascularity of extraocular muscles may be responsible for the excellent results seen in these patients. Because of the possibility of a severe inflammatory reaction to toxins, particularly when the larva dies, some investigators have advocated simultaneous albendazole and corticosteroid therapy^{7,12} or initial corticosteroid treatment administered a few days prior to treatment with albendazole.¹³

TABLE 3
RELATIONSHIP OF TREATMENT TYPE TO OCULAR
MOTILITY* IN PATIENTS WITH EXTRAOCULAR
MUSCLE CYSTICERCOSIS (N = 32)

Treatment	No. Patients	Ocular Motility	
		No. (%) Normal	No. (%) Residual Restriction
Albendazole alone	5	1 (20)	4 (80)
Albendazole and prednisolone	18	10 (56)	8 (44)
Cyst excision	4	2 (50)	2 (50)
Spontaneous extrusion	5	3 (60)	2 (40)

*At patients' most recent follow-up examination.

Corticosteroids serve to control the inflammation and prevent permanent muscle fibrosis. It also has been reported steroids increase the plasma levels of albendazole.²²

Some surgeons have performed surgical removal of the cyst in extraocular muscle cysticercosis.^{9,10,12} However, these cysts are surgically challenging because of their attachment to the muscle and their propensity to rupture intraoperatively. Surgery also is fraught with inadvertent muscle injury and postoperative fibrotic response, which may result in restricted ocular motility. Spontaneous extrusion of the extraocular muscle cysticercosis cyst probably as the result of inflammation and subsequent necrosis of the overlying conjunctiva also has been reported.^{12,23}

We treated our patients with albendazole alone, albendazole combined with corticosteroids, and surgical removal of the cyst. Spontaneous extrusion of the cyst occurred in a few patients after the start of therapy; it is unclear whether treatment played any role in this event. Following treatment, inflammatory signs resolved in all of our patients. Half of our patients had residual restriction of ocular motility and half demonstrated diplopia in the extreme position of gaze. We did not find a significant difference in the incidence of residual motility restriction in patients treated with albendazole alone compared to those treated with albendazole combined with corticosteroids. The addition of steroids to albendazole therapy helps in controlling inflammation. It is possible that in our patients, structural damage to the affected muscle had already occurred

before inflammation was controlled with steroids. Therefore, the possibility of residual restriction of ocular motility should be explained to patients. Residual diplopia in the extreme position of gaze usually is not troublesome for most patients and needs to be treated only in patients who find it bothersome.

This study represents the largest reported series of patients with extraocular muscle cysticercosis. The findings indicate a high index of suspicion, awareness of the various clinical presentations, and familiarity with radiographic and ultrasonographic signs are important in diagnosing extraocular muscle cysticercosis. Although the addition of corticosteroids to albendazole therapy did not help in improving restricted ocular motility in our patients, it did control the inflammation caused by the dying cyst. Therefore, we recommend adding corticosteroids to albendazole therapy for treating extraocular muscle cysticercosis.

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