Orbital cysticercosis: varied presentations with management plan

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Abstract

Introduction: Cysticercosis is a systemic parasitic disease caused by the larval form of the cestode pork tapeworm, Taenia solium. Along with subcutaneous tissue and skeletal muscles, eyes, brain, bladder wall, and heart are also the commonly involved sites of lodgment of the larva. The larval lodgment at these sites causes potentially harmful and variable clinical manifestations. Ocular cysticercosis can be extraocular or intraocular. Material and Methods: An immunologic reaction with fairly intense inflammatory signs and symptoms may be produced, and the surrounding structures may be compressed. Acquired strabismus, diplopia, recurrent redness, and painful proptosis are some of the clinical signs in patients with orbital cysticercosis. The present study report 8 cases of ocular cysticercosis with a variety of clinical presentations depending on the exact site of larval involvement. Result: Patient CT Scan was done to confirm the clinical diagnosis in most of the posterior segment and orbital lesions. Rest investigations were within normal limits. Conclusions: It becomes important to report these cases because of the relative rarity of the condition these days, myriads of presentation and age group, the relative concentration of cases in endemic areas, and CT Scan used as the definitive investigation to confirm orbital cysticercosis and rule out neurocysticercosis.

Keywords: Ocular cysticercosis, Myocysticercosis, Taenia solium, CT Scan

Introduction

Cysticercosis is the infestation by the larval form of the pork tapeworm Cysticercus cellulosae. It can be contracted by many modes such as ingestion of the infective cysticerci while consuming undercooked pork or ingestion of eggs of T. solium in contaminated water, food or vegetables, or by regurgitation of eggs from the small intestine called as autoinfection [1,2]. Therefore, Cysticercosis more commonly occur where sanitary conditions are poor and where raw, undercooked pork and beef are routinely consumed [2]. However, in endemic areas incidences are in the rise in vegetarians also. Appropriate sanitation and personal hygiene are important in the control of fecal contamination of water and food. Raw and improperly cooked food should be avoided, especially in endemic areas [3]. Ocular cysticercosis is common in the Indian subcontinent [4].

Materials and Methods

Study type and design: A progressive observational study design followed for patients diagnosed with ocular cysticercosis.

Duration of study: In the calendar year 2019 from Jan 2019 - Dec 2019, from the time of confirmation of diagnosis, patients were followed regularly for 6 months, starting from day 0, then weekly for 6 weeks then with every 2 weeks interval for next 6 weeks followed by 1-month interval for next 3 months.

Sampling method and sample size collection: All patients for whom the diagnosis of ocular cysticercosis was confirmed were included in the study.

Inclusion criteria: Confirmed diagnosis of ocular cysticercosis.

Exclusion Criteria

a. Immunocompromised patients
b. Cysticercosis involving any other system than ocular
c. Age < 5 years

Ethical consideration and permission: To carry out the study, necessary ethical clearance was taken from the institutional ethical committee. No financial gain or support was taken or offered to anyone involved in the study.
A total of 10 patients who presented to F.H. Medical College and hospital and were diagnosed to have ocular cysticercosis were included in the study. The demographic profile of the patients like age, sex, and address was recorded. A detailed general examination to determine cysticercosis elsewhere in the body and a thorough ophthalmic examination was performed. 2 patients were lost to the follow-up and were not included in the study.

A computerized tomography scan of the brain and orbit was carried out to rule out the possibility of neurocysticercosis and supplement the diagnosis of ocular cysticercosis. The diagnosis of ocular cysticercosis was based on CT evidence of cyst with or without scolex.

Medical management was given to all cases, which consisted of oral albendazole (15 mg/kg/day) in two divided doses along with prednisolone (1.5 mg/kg/day) in a single dose for four weeks. Prednisolone was tapered off over the subsequent four weeks. Patients were followed regularly starting from day 0, then weekly for 6 weeks, then for every 2 weeks interval for the next 6 weeks followed by 1-month interval for next 3 months All cases responded completely to the medical management.

Result

In our study out of the eight cases, five were males and three females. 3 cases were in the age group 9 to 13 and 5 in the age group 14 to 35, thus the Median age was 19 years. The mean follow up period was 5 months.

Table-1 shows out of 8 cases 2 were myocysticercosis with lateral rectus muscle involvement. 1 patient involved retrobulbar soft tissue and 2 patients had conjunctival involvement. There was 1 patient each involving preseptal orbit, submacular posterior segment, and optic nerve sheath.

On presentation, 3 patients had proptosis with diplopia. Out of these 2 patients also had extraocular movement restrictions. 2 patients presented with foreign body sensation, with redness in the eye and subconjunctival cystic swelling in each respectively. Lid swelling was presented in 1 patient and 2 had diminished vision.

Absolute eosinophil count in all 8 patients ranged between 200 to 300 cells/cm.

CT Scan Brain and orbit was done in 4 patients and only brain CT in 1 patient. In all the orbital CT Scan well defined hypodense cystic lesion was seen whereas brain CT was normal in all cases. In this study, all the patients were taken up for medical management.

Table-1: Clinical profile of orbital cysticercosis in 8 patients.

<table>
<thead>
<tr>
<th>Case</th>
<th>Type</th>
<th>Age/ Sex</th>
<th>Symptoms</th>
<th>AEC (cells/ cmm)</th>
<th>Elisa</th>
<th>CT Scan</th>
<th>Intervention</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Myocysticercosis (Lateral rectus muscle)</td>
<td>9/M</td>
<td>Proptosis, EOM restriction</td>
<td>220</td>
<td>Not done</td>
<td>Small ring-enhancing hypodense area measuring 9.5 * 8.4 mm was seen on the posterior aspect of the left lateral rectus muscle Brain Ct - Normal</td>
<td>Medical</td>
</tr>
<tr>
<td>2.</td>
<td>Myocysticercosis (Lateral rectus muscle)</td>
<td>12/F</td>
<td>Proptosis, EOM restriction, Diplopia</td>
<td>240</td>
<td>Not done</td>
<td>Well defined orbital hypodense lesion (2.6 * 1.5 * 1.3 cm) with peripheral isodense capsule and tiny eccentric hyperdense focus in left lateral rectus muscle Brain Ct – Normal</td>
<td>Medical</td>
</tr>
<tr>
<td>3.</td>
<td>Retrobulbar orbital soft tissue</td>
<td>22/F</td>
<td>Proptosis, Diplopia</td>
<td>220</td>
<td>Not done</td>
<td>Well defined hypodense lesion located behind the globe in orbital soft tissue</td>
<td>Medical</td>
</tr>
</tbody>
</table>
### Case 4: Conjunctival microabscess
- **Age/Sex:** 20/ M
- **Symptoms:** FB sensation, Irritation, Redness
- **AEC (cells/cm³):** 200
- **Elisa:** Not done
- **CT Scan:** Not done
- **Intervention:** Medical
- **Brain Ct - Normal**

### Case 5: Subconjunctival swelling
- **Age/Sex:** 30/ M
- **Symptoms:** Cystic swelling, FB sensation
- **AEC (cells/cm³):** 180
- **Elisa:** Not done
- **CT Scan:** Not done
- **Intervention:** Medical

### Case 6: Preseptal lid swelling
- **Age/Sex:** 13/ F
- **Symptoms:** Lid swelling
- **AEC (cells/cm³):** Not done
- **Elisa:** Not done
- **Intervention:** Medical

### Case 7: Submacular
- **Age/Sex:** 35/ F
- **Symptoms:** Diminished vision
- **AEC (cells/cm³):** Not done
- **CT Scan:** Brain CT - normal
- **Intervention:** Medical

### Case 8: Optic Nerve sheath
- **Age/Sex:** 18/ M
- **Symptoms:** Diminished vision
- **AEC (cells/cm³):** Not done
- **CT Scan:** Cystic lesion along optic nerve enclosed in O.N. sheath
- **Intervention:** Medical
- **Brain CT - normal**

### Discussion

**Demography:** In today’s scenario cysticercosis infestation is still a common entity in the developing country. Poor hygiene, low sanitation, consumption of undercooked pig meat, and history of travel to endemic areas specifically increases the incidences of the spread of the disease. Endemic areas generally lie over the tropical belt and include sub-Saharan Africa, India, and East Asia [5]. In India, 78% of the cases with ocular cysticercosis have been reported from states of Andhra Pradesh and Pondicherry [6,7]. In our study, 7 patients were local to Uttar Pradesh, while 1 patient was a resident of Bihar working as a daily wage worker in our area. Usually its more common in younger age groups though any age can be affected. There is no gender predilection [8]. In our study, the mean age was 19 years and 5 being males and 3 as females.

**Site of Lodgment:** The larval form of cysticercosis can lodge anywhere in the eye. Depending on that cysticercosis of the eye can be intraocular or extraocular.

Various literature shows that the intraocular sites are more commonly subconjunctival space, subretinal space, and intravitreal space whereas the anterior segment is very less common [8].

In the orbit, cysticercosis can lodge into any extraocular muscle or other adnexal structures. Studies suggest the most common predilection of orbital lodgment is the lateral rectus muscle and the superior muscle complex [9,10]. Similarly in our study to 5 cases were orbital with 2 of them involving lateral rectus muscle and 1 optic nerve sheath and rest 3 cases were intraocular.

Other various case reports and studies illustrate the involvement of Inferior Rectus alone, the combination of Levator palpebrae superioris and Superior rectus muscle, and unusual association of multiple brain neurocysticercosis with ocular cysticercosis [10,11].

**Clinical picture:** According to a study published in India, 12.8% of all cases of cysticercosis involve the eye or its adnexa [3]. Ocular cysticercosis can cause considerable loss of sight depending on its location and time-lapse in starting treatment.

Ocular involvement is usually unilateral but bilateral involvement may occur in cases of disseminated cysticercosis [12,13]. Left eye is more commonly involved in comparison to the right, possibly because larva may be preferentially routed to the left internal carotid artery which directly originates from the aorta; however, this has not been substantially proven [8]. In our study, all the 5 orbital cases and 1 intraocular case involved left eye whereas only 2 cases were in the right eye.

**Extraocular/orbital cysticercosis/Myocysticercosis:** It presents with a spectrum of signs and symptoms like repeated redness, pain, inflammation, proptosis, diplopia, and restricted ocular motility. This prompts to the differential diagnosis of Intraorbital Inflammatory diseases. Strong clinical suspicion required for conducting the necessary investigation and starting treatment.
Other rare presentations include Duane’s retraction syndrome, Brown syndrome, or blepharoptosis depending upon exact muscle involvement and the amount of inflammation [14].

Lid/Preseptal cysticercosis—In the eyelids it causes a mass or swelling that is subcutaneous, painless, and mobile and causing varying degrees of mechanical ptosis [5].

Subconjunctival cysticercosis: Conjunctival involvement is usually in the form of a painless or painful yellowish, nodular subconunctival mass with surrounding conjunctival congestion. Rarely subconjunctival abscess or granuloma can also occur [15].

Optic nerve: Optic nerve compression by the cyst may cause decreased vision, disc edema, and painful ocular motility [16]. The enlarging cyst may lead to axial proptosis, restricted ocular motility, or simply may present as atypical optic neuritis.

Diagnosis: The diagnosis of ocular cysticercosis is based on clinical, serologic, and radiological findings. Out of these clinical and serologic reports are often non-confirmatory [8]. Hence, radio imaging studies (High-Resolution USG B Scan and CT Scan of the orbit) becomes the mainstay of diagnosis by direct observation of cystic lesion suggestive of the scolex. In our study Absolute Eosinophil Count was done in all patients that were within the normal range.

Stool Examination: It is done for examining adult T. solium worm or its eggs in suspected cases of myocysticercosis. But the disadvantage of this test is that it is not essential that all patients with myocysticercosis have the adult worm in their intestines except in those cases, which are acquired by autoinfection [8].

B-scan ocular ultrasonography- It reveals a well-defined cystic lesion with clear contents and a hyperechoic area suggestive of a scolex [17].

Ocular ultrasonography is a useful tool not only for diagnosis but also for monitoring of the cyst during treatment.

CT scanning of the orbits is a reliable technique to help establish a diagnosis of ocular cysticercosis. The characteristic feature is a hypodense mass with a central hyperdensity suggestive of the scolex. Usually, a solitary cyst with wall enhancement is observed. Adjacent soft-tissue inflammation may be present. The scolex may not be visible if the cyst is dead or ruptured and has surrounding inflammation. Concurrent neurocysticercosis may be present and should be excluded [6].

In our study, a CT scan of the orbit showed well defined hypodense cystic mass with peripheral wall enhancement. This was used as the confirmatory marker for cysticercosis.

Other tests that can help are serum Enzyme-Linked Immunosorbent Assay (ELISA) for anticysticercal antibodies and complete blood count. Positive ELISA test results help confirm the diagnosis in cases where scolex is not visible but negative results do not exclude cysticercosis. A complete blood count may reveal eosinophilia [18].

Treatment: Albendazole and praziquantel are the larvicidal drugs used in the treatment of cysticercosis in humans. Once the diagnosis of orbital cysticercus is confirmed, it is of utmost importance to rule out intraocular and central nervous system involvement. Dying cysticercus releases its toxin and incites severe inflammatory reactions leading to vitreous and may lead to blindness. Hence it is mandatory to check for intra-ocular involvement of cysticercus cyst. Cure rates range from 60 to 85% in the usual dosing with most series showing albendazole yielding slightly higher cure rates [8].

It is usually given at 15 mg/kg per day with a maximum of 400 mg/bid (higher doses have been given) with repeated dosing as clinically warranted. In this study, we treated all the patients medically with Tab. Albendazole with the same dosing regimen along with corticosteroid (Tab. Prednisolone) 1 mg /kg body weight.

Treatment may increase inflammation as the cyst involutes, leading to worsening clinical status. Thus, concomitant administration of corticosteroids is recommended to avert an inflammatory response that usually occurs 2-5 days after initiation of therapy [17,19]. There is a general consensus that corticosteroids should be used prophylactically when patients have numerous, large, or critically located lesions. In our study, only 1 patient with lateral rectus involvement reported an inflammatory response at 1-week post-treatment. It resolved on its own by another 1 week.
Fig-1: CT Scan showing enhancement of lateral rectus muscle with mild enhancement. Small ring-enhancing hypodense area measuring 9.5*8.4mm seen in the posterior aspect of the left lateral rectus muscle.

Orbital cysts are best treated conservatively with a 4-week regimen of oral albendazole (15 mg/kg/d) in conjunction with oral steroids (1.5 mg/kg/d) in a tapering dose over a 1-month period. Albendazole is a well-tolerated broad-spectrum cytocidal drug and destroys approximately 85% of cysts with a single course.

Albendazole has largely supplanted praziquantel because of slightly greater cure rates, decreased cost and increased availability. Serial B-scan ocular ultrasonography or CT scanning of the orbit helps to follow the resolution of the cyst, which is recognized by the disappearance of the scolex. Cysticercosis can be prevented through practicing good hygiene measures, such as washing hands frequently, washing raw vegetables and fruits well before consumption to prevent fecal-oral transmission, and avoiding consumption of raw or under-cooked pork and other meat.

**Conclusion**

For patients from poor socioeconomic status and who could not afford all the serologic and radiologic tests, CT Scan Head and orbit along with strong clinical suspicion becomes the mainstay of diagnosis.

**What does the study add to the existing knowledge?**

Medical management in orbital cysticercosis is extremely effective and achieves a clinical resolution in most patients.

**Author’s Contribution**

Dr. Noorjahan Sajid: Study design
Dr. Priyangee Sen: Manuscript preparation
Dr. Shoeb M.: Manuscript preparation

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**Conflict of interest:** None declared

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**References**


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