



OCULAR CYSTICERCOSIS



Eye Learn
All about the Eye

Dr. Krati Gupta
Dr. Saurabh Deshmukh

www.eyelearn.in

OCULAR CYSTICERCOSIS



Eye Learn
All about the Eye

1. Write in brief on ocular involvement in ocular Cysticercosis and briefly outline the management of ocular cysticercosis. D2012
2. Discuss clinical features, diagnosis and management of intraocular and extraocular cysticercosis 3+3+ 4 J2013
3. Clinical features, diagnosis and management of intra-ocular cysticercosis.3+3+4 D2016
4. Write down modern management of posterior segment intraocular cysticercosis. D2010
5. Clinical features and management of intraocular cysticercosis. (2008)
6. Cysticercosis and the eye. (2001) (2005) (2003)

Introduction

1. Cysticercosis is the most common ocular tapeworm infection.
2. It occurs especially in underdeveloped areas where hygiene is poor.

Epidemiology

1. Human infection is caused by *Cysticercus cellulosae*, the larval stage of the cestode *Taenia solium*, which is endemic to Mexico, Africa, Southeast Asia, Eastern Europe, Central and South America, and India.
2. Although the eye is more commonly affected than any other organ, neural cysticercosis is associated with significant morbidity and a mortality of 40%.
3. Ocular cysticercosis is a disorder of the young, occurring most frequently between the ages of 10 and 30 years, without sex predilection.

Life cycle

1. Human cysticercosis is caused by ingestion of water or foods contaminated by the pork tapeworm.
2. The eggs mature into larvae, penetrate the intestinal mucosa, and spread hematogenously to the eye via the posterior ciliary arteries into the subretinal space in the region of the posterior pole.
3. Larvae within the subretinal space may cause an exudative retinal detachment or may perforate the retina, causing a retinal break and gain access to the vitreous cavity.

Morphology of cyst

1. The characteristic clinical appearance is that of a globular or spherical, translucent, white cyst with a head, or scolex, that undulates in response to the examining light within the vitreous or subretinal space.
2. The cyst itself varies in size from 1.5 to 6 disc diameters.

Clinical presentation

1. Ocular involvement is usually unilateral but bilateral involvement may occur in cases of disseminated cysticercosis.
2. 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.
3. The medial side of the eye has been more commonly involved on account of the anatomic course of the ophthalmic artery, which after giving off the lacrimal branch runs on the medial side of the orbit before diving into the terminal branches.
4. Infestation of the ocular adnexae is probably through the anterior ciliary artery.
5. Parasite reaches the posterior segment through the posterior ciliary artery.
6. Localization of the cyst to optic disc may occur through the central retinal artery.

Symptoms

1. Depending on the location of the cyst, patients may present asymptotically with relatively good vision or may complain of floaters, moving sensations, ocular pain, photophobia, redness, and very poor visual acuity.
2. Cysticercosis may involve any structure of the eye and its adnexae.

A. Extra-ocular cysticercosis

1. Lid and subconjunctival cysticercosis

1. Lodgment of cysts in the subconjunctival space is another common site, followed by the eyelid, optic nerve and retro-orbital space
2. Involvement of the eyelids present as a subcutaneous, painless, mobile mass with varying degrees of mechanical ptosis.

3. Conjunctival involvement is usually in the form of a painless or painful yellowish, nodular subconjunctival mass with surrounding conjunctival congestion.
4. Rarely subconjunctival abscess or granuloma may occur.
5. Subconjunctival presentation could be due to spontaneous extrusion of cyst from extraocular muscle into the subconjunctival space.
6. Because of its constant motility it may erode through the conjunctiva and comes out leaving a rent in the conjunctiva which ultimately heals within a short period.

2. Extraocular myocysticercosis

1. The extraocular muscle form is the commonest type of orbital and adnexal cysticercosis.
2. Cysticercosis of extraocular muscle usually presents as recurrent pain, redness, proptosis, ocular motility restriction, diplopia and ptosis.
3. One or more extraocular muscles may be simultaneously involved, although a propensity for involvement of the superior muscle complex and the lateral rectus muscle has been reported.
4. Depending on the muscle involved, clinical findings of Duane Retraction Syndrome or Brown Syndrome may be observed.
5. Blepharoptosis may occur due to severe inflammation, anterior orbital cyst or involvement of superior muscle complex.

3. Lacrimal gland

1. Lacrimal gland cysticercus cyst may cause a chronic dacryoadenitis and enlargement of the gland.
2. Lacrimal canalicular obstruction due to adnexal cysticercus has also been reported.

B. Intraocular cysticercosis

1. Optic nerve

1. Optic nerve compression by the cyst may cause decreased vision, disc edema and painful ocular motility.
2. Enlarging cyst may lead to axial proptosis, restricted ocular motility or simply may present as atypical optic neuritis.
3. A cyst with hooklets on the optic disc has been reported in a case.

2. Posterior segment cysticercosis

1. Posterior segment is involved most often, with the subretinal space harboring the parasite more often than the vitreous.
2. The macular region being the thinnest and most vascularized, the larvae lodges itself in the subretinal space from where it perforates and enters into the vitreous cavity.
3. In this process, the parasite can cause a retinal detachment, macular hole or incite an inflammatory response
4. It may cause exudative retinal detachment and focal chorioretinitis.
5. Both anterior chamber and vitreous may demonstrate inflammatory activity; vitreous inflammation is more pronounced during the early stages of the disease.
6. Larvae death produces a severe inflammatory reaction characterized by zonal granulomatous inflammation surrounding necrotic larvae on histologic examination.
7. RPE atrophy may be observed surrounding the presumptive entry site of the cysticercus into the subretinal space

Complications of intraocular cysticercosis include

1. Severe inflammation (vitreous exudates, organized membranes in vitreous),	2. Severe AC reaction	5. Proliferative vitreoretinopathy,	7. Complicated cataract, hypotony
	3. Retinal haemorrhages	6. Secondary glaucoma	8. Phthisis
	4. Retinal detachment		

3. Anterior Segment Cysticercosis

1. Anterior chamber cysticercosis is an unusual presentation and the occurrence of a live free floating cyst in the anterior chamber is a rarer occurrence with very few sporadic case reports of intracameral cysticercosis in literature.
2. The route entry of the cyst in the anterior chamber is debatable.
3. It can enter the anterior chamber from posterior segment through the pupil in aphakes, through vessels supplying the ciliary body or through the anterior chamber angle.
4. Ocular cysticercosis is commonly seen in the younger age group of first or second decade with no definite gender predilection.
5. The cyst may be adherent to the adjacent structures like the iris, anterior lens capsule or corneal endothelium by a stalk, or rarely remains freely floating in the anterior chamber.
6. The patient remains asymptomatic if the cyst is small or may present with complaints of diminution of vision, floater or leukocoria.

7. There may be pain and redness with associated iridocyclitis or glaucoma.
8. Glaucoma may be inflammatory in the presence of iridocyclitis or due to pupillary block caused by the cyst.
9. Intracamer al cysticercosis has been confused with cataract or anteriorly dislocated lens

C. CNS- Epileptiform seizures may be the first sign of cerebral cysticercosis

Diagnosis

- The characteristic appearance of a motile cysticercus in the anterior chamber, vitreous, or subretinal space is pathognomonic
- The clinical diagnosis of live intraocular cysticercosis is based on the morphology of the parasite as visualized with the ophthalmoscope or slit-lamp biomicroscope.
- The cyst and the scolex show characteristic undulating movements.
- When the scolex is invaginated, a dense white spot called the receptaculum capitis indicates its location within the cyst
- High resolution Ultrasonography USG, CT, MRI helps in detection of the orbital cyst.

1. Serological tests	used for the specific diagnosis of cysticercosis are indirect hemagglutination, indirect immunofluorescence, and immunoelectrophoresis such as ELISA specific serology
2. ELISA	Anticysticercus antibodies are detected by ELISA in approximately 50% and 80% of patients with ocular and neural cysticercosis, respectively.
3. CBC	A complete blood count may reveal eosinophilia
4. Stool examination	If a patient is a definitive host, with an adult tapeworm in the gastrointestinal tract, stool examination may find the eggs of T.solium.
5. AC tap	Anterior chamber paracentesis may reveal a large number of eosinophils
6. USG B scan	It may also be helpful diagnostically in the presence of intraocular cysticerci, revealing a characteristic picture of a sonolucent zone with a well-defined anterior and posterior margin. A central echo-dense, curvilinear, highly reflective structure within the cyst is suggestive of a scolex.
7. A-scan USG	It shows high amplitude spikes corresponding to the cyst wall and scolex. The scolex shows a high amplitude spike due to presence of calcareous corpuscles
8. OCT	It can also help identify the cyst in subretinal space
9. CT	Ocular cysticercosis on CT shows characteristic feature of a hypodense mass with a central hyperdensity suggestive of the scolex. Adjacent soft-tissue inflammation may be present. The scolex may not be visible if the cyst is dead or ruptured and has surrounding inflammation. CT may reveal intracerebral calcification or hydrocephalus in patients with neural cysticercosis.
10. MRI	It reveals a hypointense cystic lesion and hyperintense scolex within the extraocular muscle.

Differential diagnosis of posterior segment cysticercosis

<p>I. Conditions associated with leukocoria</p> <ol style="list-style-type: none"> 1. Retinoblastoma 2. Coats disease 3. Retinopathy of prematurity 4. Toxocariasis 5. Persistent fetal vasculature 6. Retinal detachment 	<p>II. Diffuse unilateral subacute neuroretinitis</p> <p>III. Choroidal tumours</p> <p>IV. Serous retinal detachment</p> <p>V. Other parasitic infections like toxoplasmosis</p>
--	--

Differential diagnosis of orbital cysticercosis

1. Idiopathic myositis,	3. Muscle abscess or hematoma
2. Tumours or metastasis	4. Parasitic infections like hydatid cyst

Treatment

- Left untreated, intravitreal or subretinal cysticercosis can cause blindness, retinal atrophy, and phthisis within 3–5 years.

A. Management of Orbital Cysticercosis

1. **Subconjunctival and eyelid cysticercosis**
 - Surgical removal is advocated
 - In case of subconjunctival cyst-excision biopsy is done to confirm the diagnosis
2. **Subconjunctival abscess**
 - Formation may occur due to release of toxins, which require drainage and removal of the cyst.
3. **Extraocular muscle form and retro-orbital cysticercosis**
 - Medical therapy is the recommended treatment



- In cases with proptosis, restricted motility, inflammation or ptosis CT imaging must be performed to rule out any cystic intramuscular lesion with scolex.
- If such a lesion is present or ELISA is positive, oral albendazole (15 mg/kg) and oral steroid (prednisolone 1 mg/kg) are given.
- In the presence of a cystic lesion without scolex or when ELISA is negative, oral steroids must be prescribed.
- In case of recurrence, repeat CT scan is required, and if there is a cystic lesion, a repeat course of albendazole and steroid is to be given.
- When there is no evidence of a cystic lesion then biopsy is indicated.

4. **Lacrimal gland cysticercosis** -Treatment is controversial due to the limited number of cases involving lacrimal gland

B. Management of Intra-ocular Cysticercosis

- Untreated intraocular cysticercosis incites severe ocular inflammation, more so when the cyst dies.
- Hence, surgery is the treatment of choice.

1. Optic nerve cysticercosis

- In a case report, surgical removal of the cyst was attempted for the optic nerve cyst near the entrance of the optic canal with remarkable visual recovery

2. Intravitreal cysts

1. Various modalities have been described in the surgical management of intravitreal cysticercosis such as diathermy, photocoagulation and cryotherapy.
2. But these methods have now become obsolete as it results in the release of toxins from the cyst causing severe intraocular inflammation.
3. Surgical removal of the cyst can be through either the transretinal or transscleral route.
4. Earlier intoto removal of cyst was advocated as it would help prevent any rupture of the cyst and release of toxic cyst products into the ocular cavity that may induce severe vitritis.
5. In the era of microincision vitrectomy surgery (MIVS), removal of the cysts using the vitreous cutter is advocated.
6. The high speed cutting rates with the maximum suction ensures that the cyst contents barely come in contact with the ocular structures with minimum release of toxins.
7. Systemic corticosteroids are used before and after surgical removal of the cysticercosis cyst.
8. Medical therapy other than corticosteroids is not advocated

3. Subretinal cyst

1. Earlier, small subretinal cysticercus was treated with xenon or argon photocoagulation.
2. Subretinal cysts anterior to the equator may be removed transsclerally, whereas subretinal cysts posterior to the equator are best removed transvitreally.
3. The cyst has to be localized with indirect ophthalmoscopy, the exact site marked with diathermy.
4. A radial sclerotomy of adequate size is made at this site, and preplaced sutures are placed.
5. The choroid is exposed and obvious blood vessels cauterized.
6. Indirect ophthalmoscopy should be repeated to confirm that the parasite had not moved.
7. The cyst can be removed through the choroidal incision with gentle pressure on the globe.
8. Pars plana vitrectomy is the safest and effective technique to remove the cyst by creating a retinotomy and bringing the cyst into the vitreous cavity.
9. This method ensures complete removal of the toxin and the remnants of the parasite.
10. Also, it avoids extensive periorbital dissection ensuring adequate retinopathy and retinal reattachment with faster recovery.

4. Communicating cysticercosis

1. Kumar et al first described a viable intravitreal cysticercus cellulosa in communication with subretinal space.
2. The cyst was removed intoto via the direct scleral approach.
3. Multiple cysts in the same eye at different locations may be present.
4. There may be a rhegmatogenous retinal detachment associated with it.
5. The current modality of treatment is pars plana vitrectomy.
6. With timely pars plana vitrectomy, good visual acuity can be achieved in patients with intravitreal and sub retinal cysticercosis without macular involvement.



5. Anterior chamber cysticercosis

1. As anthelmintic therapy can lead to severe inflammation in the event of a live cyst degenerating, surgical removal of the parasite in toto is the mainstay of treatment.
2. Systemic cysticercosis should be ruled out especially neurocysticercosis with the adequate neurosurgical examination and management of the same, as it would require anthelmintic therapy with steroid cover after intracameral cyst removal.
3. The different surgical modalities of surgical removal of anterior chamber cysticercosis cyst include paracentesis, cryoextraction, erysiphake extraction, extraction with capsule forceps and viscoexpression.
4. Viscoexpression is the treatment of choice as it is a simple and safe technique with minimal surgical manipulation in the anterior chamber, minimal risk of cyst rupture and does not require any sophisticated instrumentation or machinery.
5. Beri et al first described this procedure through a single 3 mm supero-temporal incision.
6. Another modification of this technique is a double-incision viscoexpression method described by Kai et al for the removal of a large intracameral cyst.

C. Neurocysticercosis

1. Anthelmintic drugs such as praziquantel and albendazole have been used successfully in the medical management of active neural cysticercosis
2. However, these drugs are generally not effective for intraocular disease.
3. They are frequently used in combination with systemic corticosteroids because larvae death is accompanied by worsening of the ocular disease and panuveitis.

Prevention

- Cysticercosis can be prevented through practicing good hygiene measures,
 1. Washing hands frequently,
 2. Washing raw vegetables and fruits well before consumption to prevent fecal-oral transmission
 3. Avoiding consumption of raw or undercooked pork and other meat.