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Ocular Cysticercosis-A Review

Dr Amandeep singh[1], Dr Unni krishnnan[2]

[1]Assistant Professor, Department of Shalakya tantra, Jammu Institute of Ayurveda and Research,Nardani Jammu.[2] Chief Consultant, Dr Krishnnan Eye clinic ,Kerala.

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Abstract

Cysticercosis is a preventable and eradicable cause of blindness endemic in the Indian subcontinent, South-East Asia and other developing countries. Ocular and orbital cysticercosis has varied presentations depending upon the site of involvement, number of lesion and the host immune response. In this article we present a review of the various clinical manifestations, diagnosis and management protocol for orbital and ocular cysticercosis. Owing to its varied presentation, cysticercosis may pose a diagnostic challenge to the health professionals. Early diagnosis and management can prevent the vision loss and optimize visual outcomes.

Keywords: cysticercosis, orbital cysticercosis, ocular cysticercosis, intracameral cysticercosis

INTRODUCTION

Cysticercosis is a rare infectious disease caused by the presence and accumulation of the larval cysts of a tapeworm (cestode) within tissues of the body. The scientific name for the tapeworm that causes cysticercosis is Taenia solium (T. solium), which is also known as the pork tapeworm. Cysticercosis is a preventable cause of blindness endemic in India[1]. It is a parasitic infestation caused by Cysticercus cellulosae, which is the larval form of Taenia solium. World Health Organization recognized neural cysticercosis as an international public health issue and major cause of epilepsy.

PATHOPHYSIOLOGY

Taenia solium belongs to the class of cestodes. Human beings are definitive hosts that harbor the adult parasite in the intestine and pigs are the definite hosts harboring the larvae. Human cysticercosis occurs when they act as intermediate host by ingesting the

eggs via following modes of infestation: 1) contaminated food and water with the Taenia solium eggs (hetero-infection); 2) reinfection by ingestion ova of the existing parasite (external auto-infection) 3) retrograde peristalsis causing the transport of mature proglottids bearing eggs from bowel to stomach (internal auto-infection).

After ingestion, the eggs of Taenia solium hatch into larvae, which pierce the gut to reach the blood stream and enter various tissues (particularly the central nervous system, eyes and striated muscle)[2] where they develop into cysts producing the clinical syndrome of cysticercosis.

Cysticercus celluosae mainly has three stages of evolution. The live or vesicular cyst is the living cyst with a well-defined scolex. It causes minimal or no inflammation in the tissue. As larva begins to die the cyst wall becomes leaky, releasing toxins and causing varying degrees of inflammation. This is the colloidal vesicular stage. Eventually, the larvae die and are either totally resorbed or calcified. This is the calcified nodular stage[3].

Cysticercosis is found in the areas with poor sanitation and is endemic in South East Asia, Indian subcontinent, Mexico, South America and sub-Saharan Africa[2]. Ocular cysticercosis has a varied presentation depending upon the site of involvement, number of lesion and the host immune response. In contrast to Western literature, Indian studies have reported ocular adnexa as the most common site of involvement. While the most common site of localization reported in Western studies is the posterior segment, in the Indian literature the ocular adnexa is the most common site[8]–[10].

ORBITAL CYSTICERCOSIS

Orbital and adnexal cysticercosis have varied clinical presentation. The extraocular muscle form is the commonest type of orbital and adnexal cysticercosis. Accumulation of cysts in the subconjunctival region is ofetnly seen, followed by the eyelid, optic nerve and retro- orbital space. Lacrimal sac cysticercosis has also been reported [6].

The common clinical complains reported are periocular swelling, proptosis, ptosis, pain, diplopia, restriction of ocular motility,

strabismus, blurred vision, lid edema and orbital cellulitis.In cases of extraocular muscle involvement, superior rectus muscle commonly involved[7]. Subconjunctival involvement could be a secondary stage in those cases in which the cyst may have extruded from the primary extra ocular muscle site[8].

Optic nerve involvement is rare. Optic nerve compression by the cyst may be associated with decrease in vision and disc edema [9]. A large cyst may cause axial proptosis and restricted ocular motility.

Diagnosis of Orbital Cysticercosis

Serological tests used for the specific diagnosis are indirect hemagglutination, indirect immunofluorescence, and immune electrophoresis such as enzyme-linked immunosorbent assay (ELISA)[10]. The serology may show false positive reports.

High resolution ultrasonography (USG), computed tomography (CT), B-scan ocular ultrasonography and magnetic resonance imaging (MRI) help in detection of the orbital cyst. Stool examination for the adult worm may be performed in cases of suspected cysticercosis.

ELISA for Follow-up

In a recent study [11], ELISA using larval somatic and excretory secretory (ES) antigens was positive in 32.5% and 45% cases respectively. Anti-ES antibodies were detected more frequently in cases having extra ocular cysts compared to intraocular location.

Management of Orbital Cysticercosis

Orally Albendazole[12] (15 mg/kg) and oral steroid (prednisolone 1 mg/kg) are given.

OCULAR CYSTICERCOSIS

Intraocular cysticercosis can involve either the anterior or the posterior segment. While anterior segment cysticercosis is rarely seen, posterior segment involvement is common.

Posterior Segment Cysticercosis

In the posterior segment of the eye, vitreous cysts are more common than retinal or subretinal cysts and the inferotemporal subretinal cyst is most frequently encountered[13].

It is hypothesized that the parasite reaches the posterior segment of the eye via the high flow choroidal circulation through the short ciliary arteries. The macular region being the thinnest and most vascularized, the larvae hosts itself in the subretinal space from where it perforates and

enters into the vitreous cavity. In this process, the parasite can cause a retinal detachment, macular hole or shows an inflammatory response. On development it causes atrophic changes of the overlying retinal pigment epithelium. Sometimes, it may cause exudative retinal detachment and focal chorioretinitis. The central retinal artery is the most likely route for cysticercosis involving the optic nerve head. Very few cases of optic nerve cysticercosis have been reported in literature[14]. 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 [15].

A dying cysticercosis cyst can incite a severe inflammatory response, due to the leakage of the toxins from the micro perforations present in the cyst wall[16]. Inflammatory reaction can be present even with living parasite, and more so with vitreous cysts than subretinal cysts.

Complications

Complications of intraocular cysticercosis include severe inflammation (vitreous exudates, organized membranes in vitreous), severe anterior chamber reaction, retinal haemorrhages, retinal detachment, proliferative vitreoretinopathy, secondary glaucoma, complicated cataract, hypotony and phthisis.

Diagnosis of Posterior Segment Cysticercosis

The diagnosis of cysticercosis is made by (ELISA), USG (A and B scan), CT scan and MRI scan.On indirect ophthalmoscopic examination, a live cyst can be seen as a translucent white cyst with dense white spot formed by the invaginated scolex with typical undulating movements

Treatment of Posterior Segment Cysticercosis

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

Intravitreal cysts

Diathermy, photocoagulation and cryotherapy, Surgical removal of the cyst, Systemic corticosteroids are used before and after surgical removal of the cysticercosis cyst..

Subretinal cyst

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. This

method ensures complete removal of the toxin and the remnants of the parasite.

Anterior Segment Cysticercosis

Anterior chamber cysticercosis is an unusual presentation and the occurrence of a live free floating cyst in the anterior chamber is a rarer occurrence. The route entry of the cyst in the anterior chamber is debatable. It can enter the anterior chamber from posterior segment through the pupil in aphakes, through vessels supplying the ciliary body[17]–[18] 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 [19].

The cyst may be adherent to the adjacent structures like the iris, anterior lens capsule or corneal endothelium by a stalk[20],or rarely remains freely floating in the anterior chamber[21]. The patient remains asymptomatic if the cyst is small or may present with complaints of diminution of vision, floater or leukocoria. There may be pain and redness with associated iridocyclitis[22]-[23] or glaucoma.

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 different surgical modalities of surgical removal of anterior chamber cysticercosis cyst include paracentesis, cryoextraction, erysiphake extraction, extraction with capsule forceps and viscoexpression[24].

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.

Conclusion

Cysticercosis is a disease closely related to improper hygiene and sanitary conditions. Therefore prevention by health education of the population is an important aspect of disease control. Prevention is possible by avoiding the consumption of undercooked or raw pork, proper washing of hands after using toilets and before food handling and by washing and peeling of raw vegetables and fruits before eating. Ocular and orbital cysticercosis has varied clinical

manifestations depending upon the site of involvement, stage of the cyst and the host-immune responses. With the advent of the new imaging techniques, ocular and orbital cysticercosis is now increasingly diagnosed even in non-endemic zones. A high index of suspicion along with characteristic features on imaging helps us to establish an accurate diagnosis and initiate appropriate treatment depending upon the site of involvement

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Corresponding author:
Dr Amandeep singh
Assistant Professor, Department of Shalakya tantra, Jammu Institute of Ayurveda and Research, Nardani Jammu.
Email: mannu1777@gmail.com

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