

Optic Nerve Cysticercosis at the Orbital Apex Presenting as Optic Neuritis

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Abstract

Purpose: To describe an unusual presentation of ocular cysticercosis and highlight the role of imaging in diagnosis.

Case Report: A 33-year-old female presented with loss of vision in her right eye and features suggesting optic neuritis. Magnetic resonance imaging (MRI) of the brain and orbits revealed a cystic lesion with peripheral rim enhancement in the optic nerve substance at the orbital apex. An enzyme-linked immunosorbent assay test for cysticercosis further established the diagnosis as optic nerve cysticercosis. She was treated with oral albendazole and steroids, resulting in remarkable improvement in visual acuity and resolution of the lesion.

Conclusion: A high index of clinical suspicion along with appropriate imaging methods can help diagnose rare presentations of ocular cysticercosis. With timely management, successful outcomes can be obtained.

Keywords: Magnetic Resonance Imaging; Ocular Cysticercosis; Optic Nerve; Orbital Apex

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INTRODUCTION

Cysticercosis is a parasitic disease caused by hematogenous spread and encystment of *Cysticercus cellulosae*, the larval form of the pork tapeworm, *Taenia solium*. It occurs when humans become the intermediate host of *Taenia solium*, as a result of ingesting its eggs from contaminated food. The cysts of *Cysticercus cellulosae* can lodge in the central nervous system, muscles, subcutaneous tissue, or eye resulting in varied clinical manifestations.^[1]

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CASE REPORT

A 33-year-old Indian female presented with loss of vision in the right eye for one month with associated headache and ocular pain, which increased with eye movement. The best corrected visual acuity (BCVA) was hand motions close to the face in the right eye, and 20/20 in the left eye. Extraocular movements were full but painful. Relative afferent pupillary defect was noted in the right eye, and remaining anterior segment examination was within normal limits. Examination of the right fundus revealed a hyperemic and swollen optic disc with blurring of disc margins [Figure 1a]. The macula and the retinal periphery were unremarkable. Examination of the left eye showed no abnormality. Systemic evaluation results, including the results of

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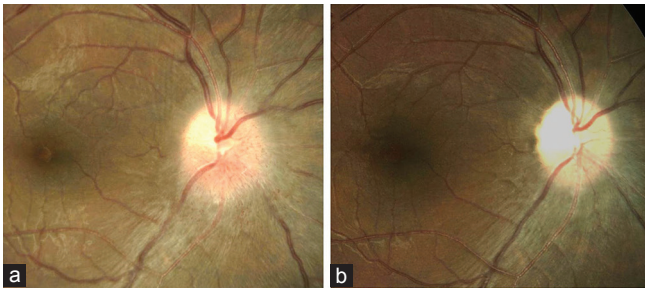


Figure 1. (a) Color photograph of the fundus of the right eye at presentation showing optic disc hyperemia and edema with blurred margins, especially inferiorly and nasally. (b) Color photograph of the fundus of the right eye following therapy with albendazole and steroids showing resolution of the disc edema.

detailed neurological examinations, were within normal limits.

B-scan ultrasonography of the right orbit showed elevation at the optic nerve head, the definitive nature of which could not be discerned. Contrast enhanced magnetic resonance imaging (MRI) of the brain and orbits demonstrated a swollen and tortuous right optic nerve with a well-defined T2 hyperintense cystic lesion with peripheral hypointensity in the optic nerve substance at the orbital apex [Figures 2a and b]. Post gadolinium contrast sequences revealed diffuse optic nerve enhancement with peripheral rim enhancement of the lesion. The brain and left orbit appeared to be normal. Based on the MRI findings, and the high prevalence and endemicity of cysticercosis in the region, a diagnosis of optic nerve cysticercosis was made. Pattern visual evoked potential (VEP) showed prolonged implicit time and decreased amplitude of the P100 wave. Hemogram revealed an elevated erythrocyte sedimentation rate, but no eosinophilia. An enzyme-linked immunosorbent assay (ELISA) test for cysticercosis was positive, which further confirmed the diagnosis.

The patient was started on a course of oral albendazole (15 mg/kg body weight in two divided doses) in combination with prednisolone (1 mg/kg body weight) for 4 weeks. BCVA showed a dramatic improvement to 20/30 in the right eye, which was associated with resolution of the disc edema [Figure 1b]. Oral steroids were tapered over the following month. An MRI scan taken 6 weeks after initiation of Albendazole revealed a complete resolution of the cyst [Figure 2c]. The clinical picture remained stable at 6-month follow up.

DISCUSSION

Clinical presentation of a patient infested with *Cysticercus* varies according to the size, location, and stage of evolution of the cyst, as well as its relation to neighboring structures. While a live cyst produces a mass effect, a dying parasite releases toxins that cause inflammation.^[2]

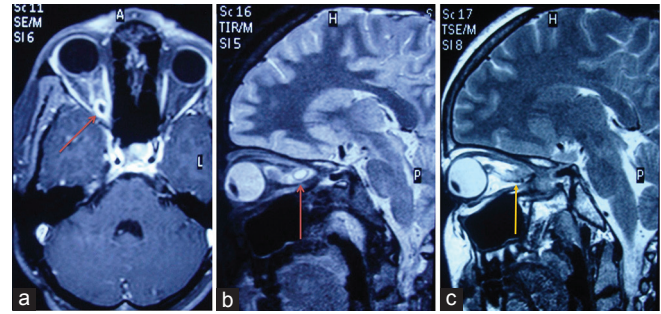


Figure 2. (a) T1 weighted axial magnetic resonance imaging (MRI) scan showing a well-defined cystic lesion near the orbital apex (red arrow). (b) T2 weighted sagittal MRI scan revealing that the lesion was in the substance of the optic nerve (red arrow). (c) Six weeks after initiation of treatment, T2 weighted sagittal MRI scan revealing complete resolution of the cyst (yellow arrow).

In the eye, cysticerci may lodge in the orbit, extraocular muscles, conjunctiva, intraocularly in the vitreous or subretinal space, or very rarely in the optic nerve.^[3]

Optic nerve cysticercosis is an unusual entity^[4-6] that results from hematogenous spread along the branches of the central retinal artery. The retrobulbar portion of the optic nerve is the commonest site for lodgement of the cyst.^[4] Clinical features include diminished vision, field loss, pupillary involvement, atypical optic neuritis, neuroretinitis, or optic atrophy.^[4] The presentation may mimic an optic nerve tumor such as a glioma^[5] or an inflammatory granuloma.^[6] Although ocular cysticercosis may be associated with neurocysticercosis in 18% of cases,^[3] optic nerve cysticercosis is usually isolated, and association with systemic cysticercosis is rare.^[5,6]

Intraocular cysticerci are usually easily recognizable due to their visibility; however orbital and optic nerve cysticerci may pose a diagnostic challenge. Serological testing for cysticercosis may be inconclusive as more than 50% of patients with neurocysticercosis have no quantifiable antibody response.^[7] Imaging studies are helpful in establishing the diagnosis of cysticercosis and may obviate the need for histopathological confirmation, which may be associated with significant morbidity. Ultrasound B scans have proven to be effective in detecting cysticercosis in the orbit and in eyes with hazy media.^[3] However, in the current case, MRI revealed the lesion in the optic nerve substance near the orbital apex. A similar location of cysticercosis, at the entrance of the optic canal, identified only with MRI and not with ultrasound has been described previously in only a single case.^[8] The cyst was present below the sheath of the optic nerve and was managed surgically.

Medical therapy consisting of a combination of Albendazole and steroids is safe and effective when instituted at the appropriate time.^[4,9] Albendazole causes death of the parasite and release of toxins, which may

lead to local inflammation. Because of this, prophylactic use of steroids is preferred, especially when the cyst is in proximity to the optic nerve.^[10] Steroids have also been reported to raise plasma levels of albendazole.^[11] A surgical approach has also been advocated for optic nerve cysticercosis,^[5,6,8] but it may result in complications and should be reserved for refractory cases.^[4]

To conclude, cysticercosis is a benign infection with a wide variety of clinical presentations. Optic nerve cysticercosis, especially when lodged in the intracanalicular portion of the optic nerve, is an exceedingly rare entity. Awareness of the varied manifestations of this infection, along with a high index of clinical suspicion, aids in early recognition and prevention of later sequelae in this condition. Neuroimaging plays a vital role in diagnosis. Medical therapy involving the combination of albendazole and steroids is effective in restoring visual acuity when instituted in a timely manner.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of Interest

There are no conflicts of interest.

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