Neurocysticercosis – A Window to Intraocular Involvement

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Abstract

Objective : To analyse the clinical characteristics of intraocular cysticercosis and the association of neurocysticercosis with intraocular involvement in these patients

Materials and methods
Settings and design: Retrospective case series of records of patients managed at a Tertiary Care Hospital in South India. Case records of patients managed at a Tertiary Care Hospital in South India over two years (October 2012 – October 2014) were reviewed and cases reported with intraocular cysticercosis were included in the study and results analyzed. Results: Five (5) patients of intraocular cysticercosis were diagnosed in a two year period at our centre, 60% being bilateral. Eight (8) eyes of five (5) patients had intraocular cysts documented by clinical examination and B mode ultrasonography (75% eyes with active cysts, 25% eyes inactive). Visual acuity at presentation of eyes with active cysts ranged from perception of light to counting finger 4m. The 75% eyes had tractional retinal detachment at presentation. Neurocysticercosis was documented in 80% of the cases on computed tomography / magnetic resonance imaging. Six (6) eyes with active cysts underwent parsplanavitrectomy with cyst removal using vitreous cutter with concurrent management of tractional retinal detachment in five (5) eyes. Postoperative visual recovery was poor in eyes with associated tractional retinal detachment while good anatomical outcome was achieved in all six (6) cases. Conclusion: Intraocular cysticercosis can be associated with cysts in other areas. High number of patients with neurocysticercosis (80%) in those with intraocular cysticercosis in our study may indicate positive association between the two which needs further investigation.

Keywords: Neurocysticercosis, Intraocular Cysticercosis, Vitrectomy

Introduction

Taenia solium is a common parasite of humans who form the intermediate hosts for the parasite. The larval form, Cysticercus cellulose can migrate from the small intestine to various parts of the body including the eye and the central nervous system leading to intraocular- and neuro-cysticercosis respectively, both of which can present variably with variable manifestations (Bouteille B, 2014). Numerous case reports of neurocysticercosis with intraocular cysticercosis has been reported
in literature which suggests some association between them (Sohoni CA, 2013). Bilateral intraocular cysticercosis has also been reported in literature and is believed to occur in less than 5% of cases (Wender JD et al, 2011). Disseminated cysticercosis with involvement of multiple organs including ocular structures, neural tissue, subcutaneous tissue, liver and other intra-abdominal tissues have also been reported (Goenka AH, 2013; Vishnu VY et al, 2013; Vaidya A et al, 2013).

We present a case series of clinical characteristics of Five (5) patients of intraocular cysticercosis. Four (4) of these patients had associated neurocysticercosis and three (3) of the four (4) with neuro-ocular cysticercosis had bilateral intraocular cysticercosis. This may indicate a possible correlation between neurocysticercosis and intraocular cysticercosis and warrants further investigation.

**Subjects and methods**

Our study is a retrospective analysis of medical records of all patients who were diagnosed to have intraocular cysticercosis and underwent treatment over 2 years from October 2012 to October 2014. Our study adheres to the tenets of the Declaration of Helsinki.

Data collected included demographics, history of any neurological or systemic involvement, detailed systemic and neurological evaluation, Clinical Ocular findings including slit lamp examination for anterior segment and vitreous inflammation, posterior segment evaluation by clinical examination using binocular indirect ophthalmoscopy, slit lamp biomicroscopy with condensing lenses and B mode ultrasonography.

All patients were subjected to neuroimaging (Computed tomography or Magnetic resonance imaging) to identify associated neurocysticercosis. Search for other extraocular cysticerci was also done. Oral antiepileptic and corticosteroids were started prior to surgery.

All patients had undergone surgical cyst removal by vitreous cutter with parsplanavitrectomy. Patients without associated retinal detachment underwent 23 G based vitrectomy while those with retinal detachment underwent 20 G based vitrectomy for better management.

Postoperative anatomical and functional outcome were defined by degree of restoration of normal anatomy and visual improvement attained respectively.

**Results**

Five (5) patients were identified, all being male, with ages ranging from 18-40 years. Two (n=2, 40%) patients had uniorcular and three (n=3, 60%) patients had binocular involvement. All patients resided in South Karnataka and had a mixed diet. All patients had history of diminution of vision (range 4 – 12 months), One patient (n=1, 20%) presented with associated pain, one (n=1, 20%) with history of floaters and flashes and one (n=1, 20%) with history of seizure at onset of symptoms treated with antiepileptics.

Eight (8) eyes of the five (5) patients were found to have intraocular cysticerci of which six eyes (n=6, 75% eyes) of five (5) patients had active disease and two eyes (n=2, 25% eyes) of two (2) patients had inactive disease (calcific cysts). One eye (n=1, 12.5% eyes) had multiple intravitreal cysts with additional membranous adhesions over the cysts.

At presentation, the visual acuity in the involved eyes ranged from perception of light to counting fingers 4m in those with active intraocular cysticercosis and 6/36 and 6/6 in two (2) eyes of two (2) patients with inactive disease. Six eyes (n=6, 75% eyes) of five (5) patients had vitritis of which two eyes (n=2, 25% eyes) of two (2) patients had additional spill over anterior uveitis. Four eyes (n=4, 50% eyes) of four (4) patients had Tractional Retinal Detachment. All patients were found to have Posterior Vitreous Detachment suggestive of Chronic Inflammation.
Image 1 shows the clinical photographs and the Computed Tomography of Brain of a patient included in our series demonstrating bilateral intravitreal cysticercus and neurocysticercosis respectively. Image 2 demonstrates the fundus photograph of a patient from our series having twin intravitreal cysticerci.

**Image 1**: Clinical photographs of a patient demonstrating
- Right Eye - Intravitreal cysticercus with associated tractional retinal detachment
- Left Eye - Intravitreal cysticercus with visible scolex
- Computed Tomography of brain - Neurocysticercosis

On B-mode ultrasonography, well defined cystic lesion with hyperechoic dot in the centre persisting on low gain suggestive of scolex were found in vitreous cavity, with or without adhesions to the retina in six eyes (n=6, 75% eyes) with active intraocular cysticercosis. Associated features such as vitritis, posterior vitreous detachment and tractional retinal detachment were also identified.

On neuroimaging (Computed Tomography or Magnetic Resonance Imaging), four of the five (5) patients (n=4, 80% patients) were found to have multiple pial based ring enhancing lesions (average 5×5 mm) in various areas of the brain parenchyma including cerebral cortex, thalamus, basal ganglia, midbrain, and cerebellum suggestive of neurocysticercosis. All four (4) patients had multiple cysts with peri-lesional edema and contrast enhancement. Two patient (n=2, 40% patients) had associated leptomeningitis.

No other extra-ocular and extra-cranial cysticerci were found in any of the patients.

Clinical characteristics of the cases are summarized in Table 1.

**Table 1: Clinical Characteristics**

<table>
<thead>
<tr>
<th>SL No.</th>
<th>Age</th>
<th>Sex</th>
<th>NCC</th>
<th>Ocular Disease</th>
<th>Laterality</th>
<th>Eye</th>
<th>VA (at presentation)</th>
<th>Duration Ocular Symptoms</th>
<th>Anterior Segment</th>
<th>Vitritis</th>
<th>CYST Type</th>
<th>RD</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>18Y</td>
<td>M</td>
<td>-</td>
<td>Unilateral</td>
<td>Left</td>
<td>Left</td>
<td>CF CF</td>
<td>12 months</td>
<td>AC reaction+</td>
<td>+</td>
<td>Degen +</td>
<td></td>
</tr>
<tr>
<td>2.</td>
<td>27Y</td>
<td>M</td>
<td>+</td>
<td>Unilateral</td>
<td>Right</td>
<td>Right</td>
<td>CF 4m</td>
<td>6 months</td>
<td>WNL</td>
<td>+</td>
<td>Degen +</td>
<td></td>
</tr>
<tr>
<td>3.</td>
<td>30Y</td>
<td>M</td>
<td>+</td>
<td>Bilateral</td>
<td>Right</td>
<td>Right</td>
<td>PL+</td>
<td>6 months</td>
<td>WNL</td>
<td>+</td>
<td>Degen +</td>
<td></td>
</tr>
<tr>
<td>4.</td>
<td>30Y</td>
<td>M</td>
<td>+</td>
<td>Bilateral</td>
<td>Right</td>
<td>Left</td>
<td>CF 3m</td>
<td>4 months</td>
<td>WNL</td>
<td>+</td>
<td>Degen +</td>
<td></td>
</tr>
<tr>
<td>5.</td>
<td>40Y</td>
<td>M</td>
<td>+</td>
<td>Bilateral</td>
<td>Right</td>
<td>Left</td>
<td>HM+</td>
<td>4.5 months</td>
<td>SIMC</td>
<td>-</td>
<td>Calcific -</td>
<td></td>
</tr>
</tbody>
</table>

One (1) patient was on treatment with anti-epileptics for seizure disorder and was put on oral corticosteroids perioperatively. All other patients were started on oral corticosteroids and antiepileptics perioperatively in consultation with a neurologist.

Six eyes (n=6, 75% eyes) of five (5) patients underwent surgical management with pars planavitrectomy and cyst removal using vitreous cutter. Those without retinal detachment underwent 23 G pars planavitrectomy (one (1) eye) and those with additional retinal detachment (three (3) eyes of three (3) patients) underwent 20 G pars planavitrectomy along with retinal detachment management in form of encirclage with 240 band with additional internal tamponade with silicone oil if deemed necessary (three (3) eyes of three (3) patients). Image 3 demonstrates the intraoperative appearance of the cysticerci in 2 of the patients in our series.

Image 3: Intraoperative photographs demonstrating:

- Brilliant transillumination of the cyst using endo-illuminator
- Cyst seen intraoperatively superior to macula

Anatomical success at discharge was noted in all six eyes (n=6, 100% eyes operated) of five (5) patients. Postoperative vision ranged from PL+ to 6/18 as described in Table 2.

Table 2: Visual Outcome

<table>
<thead>
<tr>
<th>SL. No.</th>
<th>Eye</th>
<th>Preop VA</th>
<th>Final Postop VA</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Left</td>
<td>CF, CF</td>
<td>CF, CF</td>
</tr>
<tr>
<td>2.</td>
<td>Right</td>
<td>CF, 4m</td>
<td>CF, 2m</td>
</tr>
<tr>
<td>3.</td>
<td>Right</td>
<td>PL+, HM+</td>
<td>HM+, 6/24</td>
</tr>
<tr>
<td>4.</td>
<td>Left</td>
<td>HM+, PL+</td>
<td>PR inaccurate</td>
</tr>
<tr>
<td>5.</td>
<td>Left</td>
<td>HM+</td>
<td>6/18</td>
</tr>
</tbody>
</table>


Image 4: Postoperative fundus photograph of patient demonstrated in Picture 1 showing anatomical resolution of tractional retinal detachment and removal of the intra-ocular cysticercus.

Postoperative visual recovery was poor in patients with additional retinal detachment. Two eyes (n=2, 33% eyes operated) of two (2) patients with cyst removal alone had good postoperative visual recovery from CF 3m and HM+ to 6/24 and 6/18 respectively. All eyes maintained the outcome achieved at 6 months follow up.

Discussion

Ocular manifestations of intraocular cysticercosis can be devastating due to gradual increase in size of the lesions which can ultimately lead to blindness over 3-5 years (Atul K et al, 1995). Even death of the ocular parasites induces liberation of toxins that can cause extensive ocular damage. Treatment of intraocular cysticercosis with anti-helminthic medication alone cannot aid in cure and an early surgical approach must be considered in patients for early cure and rehabilitation (Wender JD et al, 2013).

Neurocysticercosis is the most common parasitosis involving the central nervous system. Consumed proglottids release the eggs within the small intestine and the released larval forms (Cysticercosiscellulosae) can migrate to various tissues in the body including the eye and the central nervous system causing cysticercosis.
In our study, all of the patients were males and had a mixed diet. Consumption of raw pork has been a known risk factor in causation of intraocular cysticercosis along with other factors like inadequate cooking of pork and fecal contamination of water and other food sources (Bouteille B, 2014). Various studies have demonstrated variable gender distribution with slight male preponderance in different geographic areas (Bouteille B, 2014; Wender JD et al, 2011; Atul K et al, 1995). These variations have been attributed to the different cultural or social traditions in men being more exposed to infection and inequalities in health care access. Other studies have demonstrated an equal gender preponderance (Bouteille B, 2014; Wender JD et al, 2011; Malik SR, 1968).

Average age of presentation in our series was 29 ± 7.87 years. Intraocular cysticercosis has been reported to affect mainly young individuals and our finding is in accordance to published Literature (Bouteille B, 2014; Atul K et al, 1995).

Our series had patients with equal involvement of both eyes (four (4) eyes each for Right and Left). Literature has shown a slightly greater preponderance of involvement of the Left eye probably due to the more direct course of the left common carotid from the aorta (Atul K et al, 1995; Reddy PS et al, 1964; Sharma T, 2003). However occurrence of bilateral disease has been rare, with reports of incidence of bilateral disease ranging from <1% to as high as 5% in various studies (Wender JD et al, 2011; Atul K et al, 1995). Our series had three (3) of five (5) patients (60%) cases with bilateral disease. This may represent a Berksonian Bias as cases in our series were reviewed from records of patients who were surgically managed at the Retina Department of our hospital.

It is believed that the cysticerci enter the eye via the choroidal vessels due to extensive choroidal blood flow. Once in the choroidal circulation the cysticerci enter the sub-retinal space and thereafter through a retinal break (leaving behind a retinal scar) into the vitreous cavity (Wender JD et al, 2011; The Lancet Infectious D, 2014). In our series, one eye (n=1, 12.5% eyes) was found to have multiple cysticerci while the remainder of the cases had solitary intravitreal cysticerci (87.5% eyes). Entry into the vitreous cavity is associated with intense inflammation, which is responsible for the secondary effects like proliferative vitreoretinopathy, peripheral vitreous detachment and tractional retinal detachment in up to 50% cases and spill-over anterior uveitis in up to 20% cases. This is in concordance with the previous reports in literature (Wender JD et al, 2011; Atul K et al, 1995; The Lancet Infectious D, 2014).

Visual prognosis associated with intraocular cysticercosis in the past has been recorded to be poor. This is attributable to the delay in the diagnosis of the disease. Delayed diagnosis has been chalked down to lead to greater associated complications like severe vitritis, retinal detachment at presentation which in themselves are indicative of poor visual prognosis (Wender JD et al, 2011; Atul K et al, 1995; The Lancet Infectious D, 2014). In our series, cases with no additional complications achieved good visual outcome while those with additional retinal detachment and vitritis achieved good anatomical outcome but not good visual outcome. Our study is based in South India where high poverty levels and inadequate medical care access accounts for the delayed diagnosis and consequent greater incidence of associated complications and hence poorer visual outcome.
Surgical management is the norm for management of intraocular cysticercosis (Wender JD et al, 2011; The Lancet Infectious D, 2014).

Additional management of co-existent retinal detachment was done in the same sitting.

Extra-ocular sites of cysticercosis are rare and most common site is within the CNS (Del Brutto OH, 2014). Other extraocular and extraneural sites of cysticercosis are rare. Previous reports include a subdermal site and hepatobiliary site in addition to the intraocular site (Goenka AH, 2013; Vishnu VY et al, 2013). None of our patients had any other site of involvement.

Four (4) of our five (5) patients (80% patients) were found to have neurocysticercosis diagnosed by neuroimaging (CT or MRI). Neurologists have long termed Neurocysticercosis as “The Great Imitator” as it can present and mimic almost any neurological disease (Bouteille B, 2014; Del Brutto OH, 2014). Neurocysticercosis has been identified as the most common cause of acquired seizure disorder. Most common manifestation of neurocysticercosis has been found to be seizure disorder in up to 70% of cases which are related to the associated inflammation (Burneo JG, 2014). Seizures in such patients with ocular cysticercosis need rigorous preoperative management as these patients need surgery for management of intraocular cysticerci. Any episode of seizure during the perioperative period can be detrimental for the patient and hence such patients need perioperative prophylactic anti-epileptics for intra-operative and postoperative wellbeing of the patient.

Patients with neurocysticercosis can also present with focal neurological signs, intracranial hypertension and even psychiatric disturbances (Del Brutto OH, 2014). Cysticidal drugs like albendazole and praziquantel are the mainstay in management of neurocysticercosis. Patients need symptomatic support in the form of anti-epileptics and corticosteroids to prevent the secondary effects due to inflammation caused by death of the scolices during cysticidal therapy. Corticosteroids need to be started at least 2 days prior and continued for 14 days after cysticidal therapy in neurocysticercosis (Del Brutto OH, 2014; Sinha S, 2009; The Lancet Infectious D, 2014). Associated intraocular involvement may warrant use of oral corticosteroids for longer duration to treat the intraocular inflammation.

In our series four (4) of five (5) patients (80%) had neurocysticercosis. Very few case reports of such association between intraocular cysticercosis and neurocysticercosis are present in literature (Sohoni CA, 2013; Rani A, 2006; Karande S, 2007). In previous studies, not all cases of intraocular cysticercosis had intracranial involvement. In our study, we found that all patients with neurocysticercosis who had presented to us, had intraocular cysticercosis. Neurocysticercosis can be a predictor of intraocular cysticercosis and further investigation is needed to confirm this hypothesis. Complete ophthalmic examination is necessary in patients presenting with neurocysticerci to improve the visual outcome in such patients and ameliorate the complications.

**Conclusion**

Intraocular cysticercosis can be associated with a variable presentation and outcomes with numerous factors playing a role in the outcome. Ocular involvement can be associated with cysticercosis of other parts of the body. High incidence of neurocysticercosis (80%) in our case series supported by numerous case reports of similar association may suggest a positive correlation between the two.

Literature recommend screening of ocular fundus in all cases of neurocysticercosis prior to definitive management of neurocysticercosis. This practice may help us identify more cases of intraocular cysticercosis earlier and help reduce ocular morbidity.
Neurocysticercosis as a window or predictor for intra-ocular cysticercosis needs further investigation to establish any positive correlation between the two and the degree of correlation for holistic patient care.

References


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