CHANGING TRENDS IN OCULAR CYSTICERCOSIS OVER TWO DECADES: AN ANALYSIS OF 118 SURGICALLY EXCISED CYSTS

*S Madigubba, K Vishwanath, GBKG Reddy, GK Vemuganti

Abstract

Purpose: To evaluate the frequency of ocular cysticercosis and to demonstrate the changing trends in localisation of ocular cysticercosis along with a brief review of literature. Methods: A retrospective analysis of histology proven ocular cysticercosis cases seen over a period of 20 years (1981 through 2000) was done. The pathology record forms were reviewed for demographics, clinical features with specific reference to the location of cysts in four subgroups: subconjunctival; intraocular orbit and eyelid. The distribution of cases in four five-year periods namely group A: 1981-1985, group B: 1986-1990, group C: 1991-1995 and group D: 1996-2000 and the changing trends in the location of cysts was evaluated.

Results: One hundred eighteen cysts from 118 patients aged 4-72 (mean 17.1) years were submitted to the pathology service of S D Eye Hospital, Hyderabad. Male to female ratio was 1:1.2. Total number of cases in groups A, B, C and D were 33, 41, 16 and 25 respectively. Location of cysts was subconjunctival – 74 (62.7%); intraocular-31 (26.3%); orbital-8 (7%) and lid-5 (4%). In last 20 years, significant decrease \( (P=0.0001) \) was noted in subconjunctival cases \( (85\% \text{ vs. } 28\%) \) with a significant rise \( (P=0.0001) \) in intraocular cysticercosis \( (6\% \text{ vs. } 60\%) \).

Conclusions: Frequency of surgically excised ocular cysticercosis remained constant over last two decades with an increasing manifestation of intravitreal cysticercosis in the recent years. This could imply either improved diagnostic modalities, available expertise in vitreo-retinal surgery or ineffective medical treatment for intraocular parasitic infection. The relative decrease in extraocular cysticercosis is probably due to the increased preference and success with medical management.

Key words: Subconjunctival, Inrativitreal cysticercosis, *T. solium*, ocular cysticercosis

Ocular cysticercosis is caused by the growth of the larvae of *Taenia solium* with in the ocular tissues. 1,2 Soemmering was the first to report the cyst in the eye, in 1830 and Schott extracted the cyst from the anterior chamber in 1836. 3,4 Since then there have been many reports of ocular cysticercosis published in the literature. The cysts may be located in descending order of frequency, subretinal (35%), vitreous (22%), conjunctiva (22%), anterior segment (5%) and orbit (1%). 5,6 There may be geographic difference in the incidence of location of ocular cysticercosis; 5,7 for example, involvement of conjunctiva is most commonly reported in India, 5,8 whereas involvement of posterior segment of the eye is most commonly seen in Western countries. 11-14 At our regional eye hospital, in the recent past, we observed an increase in the number of intraocular cysts that were being removed from the eye. We speculated that the reported variation in the location of the parasite reported in Indian and western literature may be due to factors other than the geographic difference. This prompted us to evaluate and document the changing patterns of this parasitic infection and to determine the frequency of ocular cysticercosis at a regional eye hospital over the last two decades. We also present a brief review of literature of ocular cysticercosis.

Materials and Methods

All tissues submitted to the pathology department of Sarojini Devi Eye hospital, with a diagnosis of cysticercosis, over a period of 20 years (January 1981 through December 2000) were included in this study. This regional eye hospital serves the population of Andhra Pradesh and a few neighbouring states and includes patients from all strata, but being a government hospital caters to more patients from low socio-economic status. The histopathology requisition forms and the medical records whenever available, were reviewed for age and sex of the patient, presenting symptoms and signs and location of the cyst in the eye. The clinical diagnosis was based on external eye examination, slit-lamp examination, fundus examination and B-scan whenever required. In general, as per the preferred practice, all accessible lesions suspected to be cysticercosis are excised at our centre. Haematoxylin and eosin stained sections of all biopsies were reviewed for confirmation of diagnosis.

To document the changing trends over twenty years, the study period was segregated into four five-year periods, namely: group A: 1981-85, group B: 1986-90, group C: 1991-95 and Group D: 1996-2000. The location of the cysts was divided into four subgroups-subconjunctival, intraocular (which included subretinal, vitreal and anterior chamber) orbit and eyelid. Location of cysts in each study period was noted.
The changing trends in frequency, mean age and location were calculated statistically using Chi-square test for trend analysis. The literature was reviewed (Medline search) for comparison with Indian and Western series of similar nature.

Results

The total number of ocular specimens received between 1981-2000 was 3204 of which cysts from patients diagnosed as ocular cysticercosis were 118, constituting 3.6% of total specimens. Male to female ratio of 118 patients was 1:1.2. The mean age of patients with ocular cysticercosis was 17 years (4-72 years) with 75% cases (88/118) below 20 years of age. Information about the clinical symptoms and signs of these patients were available in 16% of cases. The subconjunctival and eyelid cysts presented as swelling and pain; orbital cysts presented with proptosis or diplopia; and intravitreal and subretinal cysts presented with diminution of vision. The location of cysticercosis was subconjunctival in 63% (74/118) (Fig. 1), intraocular in 26% (31/118) (Figs. 2, 3) and orbito-adnexal in 11% (13/118). On gross examination the cyst appeared transparent with a pearly white spot corresponding to the scolex (Fig. 4).

On microscopic examination, the cysts showed evidence of degeneration in 97/118 (82%) of cases. The characteristic three-layered wall with a branching protoscolex (Fig. 5) was seen in all the cases, while an intact scolex with suckers and hooklets was seen in 11/118.

Figure 1: Clinical picture of a patient showing presence of a well circumscribed mass seen in subconjunctival location

Figure 2: A patient with a free floating cyst in the anterior chamber

Figure 3: Fundus photograph of a patient showing an intravitreal transparent cyst

Figure 4: Picture shows the gross appearance of the cyst with a transparent cyst wall and a pearly white dot corresponding to the scolex

Figure 5: Picture shows the cyst wall, branching body cavity and protoscolex of Cysticercus cellulose (H and E stain, x125)
Changing trends

The frequency of ocular cysticercosis in each five-year period was 3.4% (33/943) in group A; 4.3% (41/932) in group B; 3.1% (19/594) in group C; and 3.5% (25/735) in group D. There was no significant difference in the frequency at different time periods \( P=0.8236 \). There was no significant difference in the age and sex distribution of cases in all groups, as shown in Table 1. The change in location of the cyst observed over the 4 time periods is shown in Table 1 and (Fig. 6).

There was a significant drop in the frequency of subconjunctival lesions (85% in group A to 28% in group C, \( P=0.0001 \)). A reverse trend was noted with intraocular cysts, which increased from 6% (2/33) to 60% (15/25) in group D, which was also statistically significant \( P=0.0001 \). There was no difference in the frequency of orbito-adnexal lesions.

Discussion

Taeniasis, the condition caused by infection with adult worm, Taenia solium, is worldwide in distribution, but endemic in some parts of the world. While taeniasis is rarely seen in those who do not eat pork, cysticercosis occurs in all ethnic groups regardless of dietary habits.2 Cysticercosis in man is caused by the encystment of the larval form of T. solium and is the most important clinical manifestation of T. solium infection in man.2 In the normal life cycle of T. solium, man is the definitive host and pig is the intermediate host.1,2 Cysticercosis in man occurs when man accidentally becomes the intermediate host, by ingestion of eggs through contaminated water and food which in turn is related to poor hygiene and poverty, therefore, the disease is mainly seen among low socio-economic classes in China, Eastern Europe, India, Indonesia, Latin America and Pakistan.2 It can also occur due to reverse peristalsis in patient with taeniasis. Although subcutaneous cysticercosis is probably the most common form of the disease, clinical symptoms are usually manifested only in patients with cerebral or ocular involvement.2 Ocular involvement is seen in 13-46% of the infected patients,2,13 at times leading to impaired vision or blindness.4,15 Interestingly, reports from India show a variation in the frequency of cysticercosis at different locations within the eye. One of the recent reviews from India on ocular cysticercosis, reports the common site as vitreous cavity,16 while others quote the subconjunctival location as the commonest site.5-10 We present here the changing trends in frequency of surgically excised ocular cysticercosis at a regional eye centre over a period of two decades.

Diagnosis of cysticercosis is usually made on history, clinical examination and radiologic investigations and by histopathology, when it can be excised. The clinical manifestations of ocular cysticercosis, like elsewhere in the body are determined by the location, size and by the host’s immune status and inflammatory reactions.16 It is believed that the viable cysts, evoke little inflammatory response,17 while the degenerating cyst rapidly increases in size due to osmotic regulation and causes compression of the surrounding tissues and release of antigens into the surrounding tissue inducing inflammatory response.4,14,15 In the vitreous or aqueous cavity it is visualized by an ophthalmoscope as a translucent cyst with a characteristic undulating expansions and contractions, which has earned the name of “living mobile pearl”.2 Sometimes the parasite can shift its location from subretinal space into the vitreous through a retinal break which could seal spontaneously leaving behind a small chorioretinal scar.14 Ultrasonography is helpful in

| Table 1: Frequency, mean age and location in each of the four study periods |
|-----------------|-----------------|-----------------|-----------------|-----------------|-----------------|
| Frequency | 33/943 (3.4) | 41/932 (4.3) | 19/594 (3.1) | 25/735 (3.5) | 118/3204 (3.6) |
| Mean age (years) | 15.4 | 12.5 | 20.0 | 21.2 | ------- |
| Subconj. | 28/33 (84.8) | 28/41 (68.2) | 11/19 (57.9) | 7/25 (28) | 74/118 (62.7) |
| Intraocular | 2/33 (6.06) | 7/41 (17) | 7/19 (37) | 15/25 (60) | 31/118 (26.3) |
| Orbit | 1/33 (3) | 5/41 (12) | 1/41 (5) | 1/25 (4) | 8/118 (7) |
| Eyelid | 2/33 (6) | 1/41 (2) | 0 | 2/25 (8) | 5/118 (4) |
identifying the cysts in extraocular location or when the media (aqueous, lens, vitreous) is cloudy. They appear as curvilinear echoes corresponding to the cyst wall with a small bright solid area that represents the protoscolex. In the orbit and subconjunctival location, after degeneration, the cyst may provoke inflammation, thereby masking the classical sonographic feature.

Morphologically, the cyst is about 1 cm, the size depending upon the location and maturity of the cyst. It has a single invaginating protoscolex in the cavity of a fluid filled cyst, which is seen, grossly as a small pearly white spot within the cyst. Microscopically, protoscolex is recognised by its suckers, hooklets and the branching variety of the protoscolex. The cyst wall is three-layered namely the ciliated cuticle, muscle layer and a tegumental cell with scattered calcific corpuscles.

In this study, the ocular cysticercosis cyst specimens constituted 3.3% of all ocular specimens received in our regional eye hospital. The frequency of ocular cysticercosis remained constant over the last two decades, indicating that there is no decline in this disease in this part of the country. In contrast, ocular cysticercosis is uncommon in Western countries, although it was reported frequently in the later half of the nineteenth century. Public health laws eliminated the disease, except for an occasional case. Most of the case reports from the developed countries were from rural areas or in immigrants from India and those cases in US literature involve mainly Mexicans.

In this study there was no sex predilection, which agrees with most of the series reported in the literature. Male preponderance was reported by only a few studies. It is important to note that 75% of our patients were below 20 years of age. This is similar to that observed in other studies from India. Though literature points towards a higher age of presentation for intraocular cysticercosis, we observed a minimal difference in the mean age of IO cysticercosis as compared to other locations (Table 1). Overall, the subconjunctival location was the most common site, constituting 63% of all cases, followed by intraocular location, as reported in various other Indian series (Table 2). This is in contrast to the high frequency of involvement of intraocular tissues in the Western countries (Table 3). Though no definitive cause could be attributed to this difference in the site of ocular predilection, hot, dry and dusty environment in tropical countries was considered as the possible modifying factor. Surgical excision is the mode of treatment for subconjunctival cysts which do not need sophisticated equipment or expertise in the diagnosis or treatment and almost all the cysts suspected as cysticercosis in the subconjunctival location were excised and sent for histological confirmation, in our hospital.

Interestingly, we observed a clear shift in the location of cysts from conjunctiva to intraocular site during the last decade (Fig. 6). Subconjunctival location was seen in 85% of cases in group A, which gradually decreased to 28% in group D and intraocular location had been on the rise from 6% in group A to 60% in group D. Atul et al. observed a similar shift in location of cysts and suggested that this change in trend might be due to unknown alteration in the environmental or biologic factors of the parasite. Another

<table>
<thead>
<tr>
<th>Reported series</th>
<th>Total cases</th>
<th>Subconjunctival</th>
<th>Intraocular</th>
<th>Orbit</th>
<th>Eyelid</th>
</tr>
</thead>
<tbody>
<tr>
<td>Reddy et al., 1964*</td>
<td>10</td>
<td>6/10 (60)</td>
<td>2/10 (20)</td>
<td>1/10 (10)</td>
<td>1/10 (10)</td>
</tr>
<tr>
<td>Sen et al., 1965</td>
<td>11</td>
<td>9/11 (82)</td>
<td>2/11 (18)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Malik et al., 1968</td>
<td>14</td>
<td>10/14 (72)</td>
<td>1/14 (7)</td>
<td>3/14 (21)</td>
<td>0</td>
</tr>
<tr>
<td>Rao et al., 1972</td>
<td>15</td>
<td>13/15 (86)</td>
<td>1/15 (7)</td>
<td>1/15 (7)</td>
<td>0</td>
</tr>
<tr>
<td>Reddy et al., 1980</td>
<td>15</td>
<td>13/15 (86)</td>
<td>0</td>
<td>0</td>
<td>2/15(14)</td>
</tr>
<tr>
<td>Atul et al., 1995</td>
<td>33</td>
<td>1/33 (3)</td>
<td>26/33 (79)</td>
<td>6/33 (18)</td>
<td>0</td>
</tr>
</tbody>
</table>

Figures in parentheses indicate percentages

<table>
<thead>
<tr>
<th>Western series</th>
<th>Total cases</th>
<th>Subconjunctival</th>
<th>Intraocular</th>
<th>Orbital</th>
<th>Eyelid</th>
</tr>
</thead>
<tbody>
<tr>
<td>*Graefe,1866</td>
<td>90</td>
<td>5 (5.5)</td>
<td>84 (93.4)</td>
<td>1 (1.1)</td>
<td>0</td>
</tr>
<tr>
<td>*Vosgien,'11</td>
<td>266</td>
<td>84 (31.6)</td>
<td>182 (68.4)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>*Laingneer,'32</td>
<td>35</td>
<td>7 (20)</td>
<td>27 (77.2)</td>
<td>1 (2.8)</td>
<td>0</td>
</tr>
<tr>
<td>Lech,'49</td>
<td>115</td>
<td>7 (6.1)</td>
<td>107 (93)</td>
<td>1 (0.9)</td>
<td>0</td>
</tr>
<tr>
<td>Welsh et al., 2000</td>
<td>13</td>
<td>1 (7.7)</td>
<td>12 (92.3)</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

Figures in parentheses indicate percentages, *Abstracted from Duke-Elder and Perkins
possible explanation offered for this trend in location change of ocular cysticercosis was that more of subconjunctival and orbital cysts are treated medically, with Albendazole (15 mgs kg\(^{-1}\) day\(^{-1}\) along with oral prednisolone 1 to 1.5 mgs kg\(^{-1}\) day\(^{-1}\) for 4 weeks). The therapeutic efficacy of this drug for extraocular muscle cysticercosis has been reported to be good. Administration of this drug resulting in spontaneous extrusion of cyst from the subconjunctival location has been reported, but more data may be required to establish the efficacy of this drug in these cases.

More number of intraocular cysticercus cysts was removed during the last decade in our institute than before. The incidence has increased from 6% in group A to 60% in group D. This change in trend could be due to availability of sophisticated investigative modalities such as B-scan, which is simpler and economical than CT-scan, which resulted in more number of diagnosed cases than before. B-scan is particularly useful in the diagnosis of intraocular cases where fundus examination or slit-lamp examination are not feasible due to hazy media. With new advances in basic and supportive instrumentation and surgical techniques, there is better visualisation and intraocular surgeries are now being performed with less complications, thus contributing to increased number of intraocular cysts in the recent years. Intravitreal surgeries were discouraged previously because of frequent anterior segment complications, scleral collapse and compulsory removal of lens. Modern pars plana surgery is now the standard surgical technique for intravitreal cysticercosis, which is a closed vitrectomy procedure. This avoids anterior segment and retinal and scleral complications and causes minimal operative trauma. This study, to the best of our knowledge and literature search, is the largest series of cases from a single centre and therefore reflects the true changing trends in ocular cysticercosis. Being a retrospective study, complete clinical details of the patients were available only in a few cases, which is a common limitation of any study from a regional referral centre. Another limitation is the inclusion of the surgically excised cases of cysticercosis, which possibly represent only part of the cases seen in the outpatient clinic. Additional studies on the total number of cysticercosis seen in the outpatient clinics would have been more informative.

In summary, frequency of surgically excised ocular cysticercosis remained constant over last two decades with an increasing manifestation of intravitreal cysticercosis in the recent years. Being a retrospective case series, the reasons for such pattern could only be speculated. The possible reasons could be: lack of availability of non-invasive diagnostic modalities in the initial period of the study; use of modern instrumentation by the trained vitreo-retinal surgeons which improved the diagnostic skills, available expertise in vitreo-retinal surgery and ineffective medical treatment for intraocular parasite. The relative decrease in extraocular cysticercosis is probably due to the increased preference and success with medical management. Lack of demonstrable decline of ocular cysticercosis in last two decades especially in children and young adults warrants a high index of suspicion, early diagnosis and prompt treatment.

References

17. Atul K, Kumar TH, Mallika G, Sandip M. Socio-demographic


Source of Support: Nil, Conflict of Interest: None declared.

Author Help: Sending a revised article

1) Include the referees’ remarks and point to point clarification to those remarks at the beginning in the revised article file itself. In addition, mark the changes as underlined or coloured text in the article. Please include in a single file
   a. referees’ comments
   b. point to point clarifications on the comments
   c. revised article with text highlighting the changes done

2) Include the original comments of the reviewers/editor with point to point reply at the beginning of the article in the ‘Article File’. To ensure that the reviewer can assess the revised paper in timely fashion, please reply to the comments of the referees/editors in the following manner.
   • There is no data on follow-up of these patients.
     Authors’ Reply: The follow up of patients have been included in the results section [Page 3, para 2]
   • Authors should highlight the relation of complication to duration of diabetes.
     Authors’ Reply: The complications as seen in our study group has been included in the results section [Page 4, Table]